New York Chapter
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Annual Scientific Meeting

Poster Presentations

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New York Chapter
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Medical Student Clinical Vignette
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A Series of Unfortunate Events - A Rare Presentation of Thoracic Outlet Syndrome

Venous thoracic outlet syndrome, formerly known as Paget-Schroetter Syndrome, is a condition characterized by spontaneous deep vein thrombosis of the upper extremity. It is a very rare syndrome resulting from anatomical abnormalities of the thoracic outlet, causing thrombosis of the deep veins draining the upper extremity. This disease is also called “effort thrombosis” because of increased association with vigorous and repetitive upper extremity activities. Symptoms include severe upper extremity pain and swelling after strenuous activity. A 31-year-old female with a history of vascular thoracic outlet syndrome, two prior thrombectomies, and right first rib resection presented with symptoms of loss of blood sensation, dull pain in the area, and sharp pain when coughing/sneezing. When the patient had her first blood clot, physical exam was notable for swelling, venous distension, and skin discoloration. The patient had her first thrombectomy in her right upper extremity a couple weeks after the first clot was discovered. Thrombolysis with TPA was initiated, and percutaneous mechanical thrombectomy with angioplasty of the axillary and subclavian veins was performed. Almost immediately after the thrombectomy, the patient had a rethrombosis which was confirmed by ultrasound. She was treated with a second thrombectomy and then the patient underwent surgery to have a transaxillary first rib resection. Following this procedure, a doppler ultrasound revealed an occlusive thrombus in the subclavian, axillary, brachial, and basilic veins. Repeat ultrasound one month later showed no change in the thrombus. She underwent a third thrombectomy, and finally a repeat ultrasound three weeks later showed no thrombus but decreased blood flow. The patient had overall improvement in symptoms. The incidence of Paget-Schroetter disease is estimated at around 3000-6000 cases per year in the US. Although it is a rare cause of thoracic outlet syndrome, it is essential for providers to recognize and appropriately treat it. Management includes removal of the thrombus as well as treating the underlying anatomical problem. Thrombolysis will not prevent recurrence of clots, which is why our patient underwent a rib resection. Despite undergoing appropriate treatment, she still had recurrent thrombosis. This case illustrates the need for further research regarding management and prevention of recurrent thrombotic events in Paget-Schroetter disease.
Karina Doucet MD Student

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A COLOSSAL PSYCHOTIC CONUNDRUM

This study explores the case of a young male with multiple risk factors for psychiatric illness who developed THC-induced psychosis in a time where mainstream use of marijuana has grown and education on its link with psychotic disorders has not kept pace. This is noted with regards to CBD’s therapeutic potential, in contrast with high-potency THC’s unfavorable effects, particularly in patients with a family history of schizophrenia.

A 19-year-old Caucasian male with no medical or psychiatric history was admitted to the hospital involuntarily from home due to suicidal ideation. He was agitated and out of touch with reality, claiming ‘I’m from the future.’ He said he would rather kill himself than be admitted again and that his ‘mind is clear.’ The patient had recently been hospitalized seven days for psychosis. He reported a three-year history of marijuana use for anxiety. Two months prior, he began smoking Colossal (99.9% THC oil) via electronic cigarette. As per the family, the patient socially isolated himself, stopped going to college, expressed paranoia that his friend would steal his ideas, and exhibited bizarre behavior. The patient also used mind-altering drugs like psilocybin and LSD several times in the past year. His maternal grandmother was diagnosed with schizophrenia, his father had substance abuse, and there were two completed suicides on the paternal side. This patient’s clinical presentation was consistent with a diagnosis of Cannabis-Induced Psychosis (CIP). Marijuana is made of multiple chemical compounds, the two most important being CBD and THC, which can be consumed in varying ratios. The two chemicals, CBD and THC are non-psychoactive and psychoactive, respectively. They have opposing effects on mental health. A recent case-control study with 901 patients showed that marijuana users smoking high-potency THC had a 1-6 times higher risk of having a psychotic disorder compared to never users. Among people who have a psychotic response to marijuana, 50% of them develop schizophrenia or bipolar disorder. Having a family history of schizophrenia poses a genetic predisposition to developing the disease. Although there is a correlation with THC use and psychosis, a 30-year translational investigation supports that CBD can be used as a therapeutic agent in psychotic disorders like schizophrenia. Further research must be done to understand the mechanism of THC-induced psychosis so that we can clarify the contraindications for medical marijuana and utilize these drugs to the best of their therapeutic potential.
MILLER FISHER VARIANT OF GUILLAIN-BARRÉ SYNDROME: A GREAT MASQUERADER

Introduction: Guillain-Barré Syndrome (GBS) is an acquired degenerative, demyelinating neurological disorder classically characterized by progressive, symmetrical ascending paralysis. Often associated to occur after a viral illness, most commonly an upper respiratory infection (URI), followed by gastrointestinal illnesses. Miller Fisher syndrome (MFS) is a rare variant of GBS, observed in only about 1% to 5% of all cases of GBS in Western countries. MFS presents with a triad of ataxia, areflexia, and ophthalmoplegia.

Case Summary: A 44-year-old Hispanic woman presented with a 2-day history of right-sided ptosis. She also reported having diplopia and blurry vision during the initial onset but had since subsided. She denied any headaches, trauma, seizures, eye discharge, changes in speech or weakness in any extremities or recent viral illness. Her past medical history was significant for diabetes mellitus, hypertension, obstructive sleep apnea, and obesity. No significant social or family history was noted. On physical examination, she was afebrile with blood pressure of 130/85 mm of Hg, pulse of 66/min, respiratory rate of 17/min and had O2 saturation of 99% on room air. Laboratory examination revealed normal electrolytes and white count. Computed tomography (CT) of the head showed no acute intracranial pathology and mild mucosal thickening if the sinuses. Initial ophthalmic examination was unremarkable. Based on initial impression of myasthenia gravis, cerebral vascular accident and cavernous sinus thrombosis, she was hospitalized for further work up. Magnetic resonance imaging (MRI) of the head without contrast showed no evidence of infarction. A trial of 30mg pyridostigmine was given every 6 hours and showed worsening of ptosis over the next 48 hours. Later she developed ataxic gait and ophthalmoplegia. GBS was considered and cerebrospinal fluid testing did not show any significant finding. Negativity inspiratory force and vital capacity were normal. Pyridostigmine was discontinued and the patient was started on intravenous immunoglobulin (IVIG) for 7-10 days. Acetylcholine receptor antibody levels were < 0.30, which confirmed that the patient did not have MG. Anti-GQ1b antibodies came back positive with a titer value of 1:3200, confirming the diagnosis along with sensory defect on nerve conduction studies. By day 3 of IVIG, the patient endorsed significant symptomatic improvement.

Discussion: MFS is a clinical diagnosis but can be confirmed serologically with positive anti-GQ1b antibodies. Our patient first presented with ophthalmoplegia. Later, serology showed positive anti-GQ1b antibodies with a titer value of 1:3200, confirming the diagnosis of MFS. These antibodies are found in high concentrations in cranial nerves 3, 4, and 6 and may be the reason that our patient initially presented with ophthalmoplegia. Neuroimaging is typically normal for patients with MFS. However, neuroimaging can be an important tool to exclude other potentially fatal diseases. The treatment of MFS includes, corticosteroids, plasmapheresis or intravenous immunoglobulin.
A CATASTROPHIC PRESENTATION OF AN UNCOMMON HEMOGLOBINOPATHY: HbSC DISEASE DEBUTS AS ACUTE CHEST SYNDROME AND PULMONARY EMBOLISM

35-year-old female from Togo with a self-reported history of hypertension and sickle cell trait presents to the ED with a one week history of progressive dyspnea associated with pleuritic chest pain and dry cough, that acutely worsened on the 2 days prior to presentation. She was noted to be desaturating to 60% on room air, with improvement to 99% on a non-rebreather mask. The patient denied any similar episodes in the past.

Physical exam remarkable for tachycardia, hypertension, tachypnea 45-50 breaths/min with accessory muscle use. Lung exam remarkable for bilateral crackles.

Relevant Labs: leukocytosis 20,600/uL with neutrophilia, normocytic anemia hemoglobin 10.8 g/L. LDH elevated 304 IU/L. Reticulocytes increased 9.93%. CMP notable for total bilirubin of 2.1mg/dL. Peripheral smear: anisocytosis, poikilocytosis, target cells and occasional schistocytes with few sickle cells.

Imaging: chest X-ray showed bilateral lower lung infiltrates and humeral head sclerosis. Due to high suspicion for pulmonary embolism (PE), the ED team decided to obtain a CT angiogram (CTA) of the chest, which revealed an acute non-occlusive PE and bilateral ground-glass opacities and consolidations. Sequelae of end-plate infarcts were seen in the thoracic spine and an atrophied and calcified spleen were seen as incidental, but critical, findings.

Taking these findings into consideration, acute chest syndrome complicated by PE was suspected. Critical Care and Hematology were consulted. They recommended hemoglobin electrophoresis to appropriately diagnose the patient and exchange transfusion was initiated emergently. Broad spectrum antibiotics and unfractionated heparin drip were started. Patient was admitted to the medical ICU for close monitoring. Initial electrophoresis showed HbS:46.0% HbC:43.1% HbA2:4.3%, consistent with hemoglobin SC disease.

Antibiotics deescalated to ceftriaxone only and heparin drip was discontinued on day 2, when she was switched to apixaban. Patient was downgraded to the medical floors on hospital day 5. The last electrophoresis showed HbS:13.9% HbA:68.4% HbA2:3.1% HbF:1.5% HbC:13.1%. She was discharged from the hospital on day 8 on apixaban.

Discussion

HbSC disease has an incidence of 1:833 live births in African-Americans and in some West African nations it can be as high as 1:25. HbC molecules induce leakage of potassium cations and water from erythrocytes, causing intracellular dehydration and crystal formation. This is mediated by exaggerated activation of the potassium-chloride-cotransporter (KCC). This, in turn, increases the mean corpuscular hemoglobin concentration of HbS to critical levels, precipitating vaso-occlusive crises. HbSC is also associated with hyperviscosity, which can lead to PE and stroke. Higher hematocrits in HbSC are associated with osteonecrosis, as was the case in our patient. Autosplenectomy is exceedingly rare, and it is less uncommon to see adult patients presenting with splenic sequestration. There is no specific treatment for HbSC disease, but exchange transfusions can be used for the same indications than in sickle cell disease.
LYMPHANGIOLEIOMYOMATOSIS: A RARE CAUSE FOR DYSPNEA

Introduction:
Lymphangioleiomyomatosis (LAM) is a rare condition of multifactorial etiology. Patients commonly present with fatigue, dyspnea, pneumothorax and pleural effusion. We report a case of this unique condition observed at our teaching institution.

Case Report:
A 43 year old female patient with no significant medical history presented to the emergency department (ED) with progressive dyspnea. While traveling to the caribbean islands for several days, she noticed dyspnea with exertion and developed new onset large volume hemoptysis. At the time, she was admitted to a local hospital ICU where Computer Tomography (CT) was reported to show “honeycomb lung”. She was diagnosed with interstitial lung disease and discharged after clinical improvement. Upon returning home from vacation, she became progressively fatigued with substernal chest pain and visual changes, prompting repeat hospitalization at our institution. CT revealed extensive cystic changes in the lungs suggesting the diagnosis of LAM (Figure 1). After clinical improvement on corticosteroid taper, she was instructed to follow up outpatient with a pulmonologist. Serum VEGF-D obtained at the time was 1051 pg/mL, confirming the diagnosis.

Discussion:
Lymphangioleiomyomatosis (LAM) is a unique pulmonary condition, with an incidence of 1:1,000,000, associated with tuberous sclerosis occurring primarily in pre-menopausal women. This condition causes smooth muscle proliferation, destruction of pulmonary parenchyma and diffuse cystic lesions with emphysematous changes resulting in respiratory failure and death. Patients commonly present with fatigue, dyspnea, pneumothorax and pleural effusion (typically chylothorax). High Resolution CT (HRCT) is the imaging study of choice for LAM with pulmonary cysts as a characteristic hallmark. LAM should be suspected in patients with high pulmonary cyst burden but further evaluation for non-pulmonary manifestations (including renal angiomyolipoma, chylothorax, peritoneal lymphangiomycytoma), serum testing (VEGF-D), or biopsy may be required. An elevated VEGF-D (above 800 pg/mL) in conjunction with cystic lesions on HRCT is highly specific and virtually diagnostic for LAM. Management focuses on early specialist involvement as an accurate diagnosis is essential in dictating management strategy. In addition to smoking cessation, vaccinations and supportive measures for airflow obstruction and pneumothorax, early manifestation with localized lung involvement can be managed with segmental dissection or lobectomy. First-line medication used is sirolimus (or other mTOR inhibitors). For patients with advanced disease refractory to mTOR inhibitors, lung transplantation can improve quality of life.

Conclusion:
We report a case of this unique condition observed at our institution. Early diagnosis and aggressive management play a critical role in this rare condition.
LATE-ONSET SYSTEMIC LUPUS ERYTHEMATOSUS PRESENTING WITH PNEUMONITIS AND CLASS IV LUPUS NEPHRITIS

Background

Systemic lupus erythematosus (SLE) is a multisystem chronic disease that predominantly affects women of childbearing age with a decline after menopause. Here we report a patient with late-onset SLE presenting with pneumonitis and class IV lupus nephritis.

Case Report

A 57-year-old woman with diabetes mellitus, chronic kidney disease (CKD), normocytic anemia and obesity presented with dyspnea, cough, and bilateral hand and wrist pain and swelling. She was initially febrile at 100.3°F and tachycardic to 128 beats/min. Her physical examination was remarkable for a malar rash, rales, and synovitis. Hemoglobin was 9 g/dl and serum creatinine was 1.9 mg/dL. There was no hemolysis and no lymphopenia. She had a urine protein creatinine ratio 0.8 g/d with few red blood cells. Chest x-ray showed pleural effusions and a retrocardiac patchy density. Computed tomography revealed pleural effusions, pericardial effusion, large areas of consolidation within the bilateral lower lobes, and bilateral axillary lymphadenopathy. She received azithromycin and ceftriaxone empirically for pneumonia. Antinuclear antibodies were present at 1:320 (speckled) and anti-double-stranded (ds) DNA antibody (ab) levels were elevated to >300 IU/ml. Antibodies to Smith were positive (>8). C3 and C4 were diminished. She was diagnosed with SLE, and started hydroxychloroquine 400 mg/d. Despite adequate antibiotic therapy and negative cultures, high-flow supplemental oxygen was required. Thoracentesis was performed, and the pleural effusion was exudative. Bacterial cultures were negative. Renal biopsy showed a proliferative and sclerosing glomerulonephritis with focal crescents, consistent with lupus nephritis class IV. Treatment with intravenous methylprednisolone 1 g for 3 days followed by prednisone 60 mg/d and mycophenolate mofetil resulted in improvement of respiratory status and arthritis as well as a decrease of anti-dsDNA ab to 56 IU/ml and normal C3 and C4 levels.

Discussion

This case demonstrates that late-onset SLE should be considered in the differential diagnosis of anemia, CKD, and arthritis in a postmenopausal woman. Our patient presented with serositis, which is more common in late-onset than early-onset SLE1. Fortunately, as was demonstrated in our patient thus far, prognosis in late-onset SLE can be quite favorable2.

References

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SEVERE HYPERTRIGLYCERIDEMIA, A RAPIDLY REVERSIBLE SEQUELA OF HYPEROSMOLAR HYPERGLYCEMIC NONKETOTIC SYNDROME (HHNS)

Introduction:
Hyperosmolar Hyperglycemic Nonketotic Syndrome (HHNS) is a complication of diabetes mellitus characterized by severe hyperglycemia, hyperosmolality, glycosuria, and a normal blood pH, with a mortality as high as 20%. Diabetes mellitus is associated with chronic abnormalities in lipid metabolism. However, there are few published reports addressing the acute interplay of HHNS and triglyceride profiles, as well as the optimal management thereof.

Case Presentation:
A 66-yr man with a history significant for prediabetes per a HgbA1c of 6.1% one year prior, dyslipidemia and obesity presented to the ED with polydipsia, polyuria, dizziness, and blurry vision for 5 days. He denied family history of triglyceride disorders and recent alcohol use. In the ED, vital signs were normal. Initial labs were notable for a blood glucose of 920 mg/dl, triglycerides of 761 mg/dl, cholesterol of 201 mg/dl, direct LDL of 65 mg/dl, HDL of 34.30 mg/dl, Na of 122 mEq/L, pH of 7.384, serum osmolality of 326 mOsm/kg, large glucosuria, HgbA1c of 12.1%, lipase of 43 U/L, and TSH of 1.841 mIU/L. All other laboratory values were unremarkable. One year ago, he had triglycerides of 230, calculated LDL of 190, HDL of 46, and cholesterol of 284. He was started on an IV insulin infusion, intravenous fluids, and admitted to the ICU for management of HHNS. Over the next four days, blood glucose down-trended to 187, and triglycerides decreased to 438, 279, 299, and 165, respectively. He was restarted on his home medications of atorvastatin 40mg and fish oil 1000mg after 24hrs. He was discharged home on NPH and metformin 500mg, with no changes to his previous lipid-lowering regimen.

Discussion:
In states of marked hyperglycemia, such as HHNS, circulating free fatty acids are converted into triglycerides (TGs) at an increased rate. In addition, hyperglycemia accelerates hepatic TG secretion, independent of both insulin and FFA levels. In settings of insulin resistance, insulin’s anti-lipolytic effects are diminished. Hypertriglyceridemia itself can further diminish these effects. This patient did not receive any TG-lowering therapy acutely, but experienced a rapid decline in TGs. Although atorvastatin and fish oil both have a TG-lowering effect, this effect is expected after two weeks and is thus not sufficient to explain the acute decrease in serum TGs observed. The cause is likely correction of the hyperglycemia. Aggressive hydration and IV insulin therefore may be the primary interventions necessary to address the elevated serum TGs observed in HHNS.

Conclusion:
In patients presenting with HHNS, the initial impression of hypertriglyceridemia may be misleading. Niacin, fibrates, and other treatments may not be necessary to control serum TGs in the acute setting. Instead, insulin infusion and hydration may be the most effective initial management for hypertriglyceridemia in HHNS, with statin therapy for continued metabolic control.
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Resident / Fellow/Medical Student
Research
**Medha Biswas BA**

Syed Zaid, MD, Hasan Ahmad, MD, Ryan Kaple, MD, Cenap Undemir, MD, Martin Cohen, MD, Steven L. Lansman, MD, PhD, Gilbert H. L. Tang, MD, MSc, MBA

**New York Medical College**

**Changes in Mitral Annular Geometry after Transcatheter Aortic Valve Replacement: Implications on Transcatheter Mitral Valve Implementation**

**AIMS**

Patients undergoing TAVR often have concomitant severe mitral regurgitation (MR) that does not improve, and MitraClip may not be feasible in some of these patients to improve their MR. Transcatheter mitral valve replacement (TMVR) is a potential option in these patients, but TAVR may affect mitral annular geometry impacting their candidacy for TMVR. We evaluated mitral annular characteristics before and after TAVR and the effect of TAVR on MR.

**METHODS**

From February 2013 to July 2017, 70/359 patients with symptomatic severe aortic stenosis who were undergoing TAVR with the Sapien (ES), Sapien XT (XT), Sapien3 (S3), CoreValve (CV) or EvolutR (ER) had pre- and post-TAVR multidetector computed tomography (MDCT). Mitral annular geometry measurements were taken at end-systole (30% R-R interval) and end-diastole (70% R-R interval) in each patient before and after TAVR. MDCT performed on average at 3.0 +/- 3.8 months (range 0.6 – 20.6 months) after TAVR for various clinical reasons. Septolateral (SL), trigone-trigone (TT) and intercommissural (IC) distances of mitral annulus were compared using 3Mensio Valves software. MR was determined by transthoracic echocardiography performed pre-TAVR, post-TAVR, and at 30-day follow-up.

**RESULTS**

At end-systole, no differences were found in TT and SL distances of mitral annulus after TAVR among all valve types while a significant decrease in IC was found overall (p<0.01). At end-systole, a significant decrease in IC distance in XT (p=0.027) and S3 (p=0.003) groups was observed. The SL distance decreased only in CV group (p=0.029) at end-systole. At end-diastole, no differences were found in TT, IC and SL distances of mitral annulus after TAVR among all valve types, except for a significant decrease in TT distance in ES (p=0.025).

Most patients undergoing TAVR had MR (N=59) with 38 patients exhibiting trace to mild MR and 21 patients exhibiting moderate to severe MR. After TAVR, 18/21 (85.7%) patients with moderate or greater MR improved at discharge and a total of 15/21 (71.4%) improved at 30 days.

**CONCLUSION**

In this study, we used MDCT to examine the impact of TAVR on the mitral valve and found that TAVR directly affects mitral annular dimensions. There was a decrease in several mitral annular dimensions after TAVR with both balloon-expandable and self-expanding valves. Additionally, we found an improvement in MR severity in patients with baseline moderate to severe MR after TAVR. This proof of concept study suggests that as TAVR indications expand to lower risk patients, similar analyses will be needed to evaluate patients with combined aortic and mitral disease for TAVR and TMVR.
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TEMPORAL TRENDS AND ASSOCIATION OF TYPE 1 VS TYPE 2 DIABETES MELLITUS WITH IN-HOSPITAL OUTCOMES IN PATIENTS WITH ACUTE MYOCARDIAL INFARCTION

Introduction: Patients with type 1 diabetes mellitus face an increased risk of cardiovascular disease. However, studies comparing the clinical outcomes after acute myocardial infarction (AMI) in type 1 diabetes mellitus face vs type 2 diabetes mellitus are limited.

Aim: We aimed to study the temporal trends and clinical outcomes after AMI in type 1 diabetes mellitus vs type 2 diabetes mellitus patients using a national database.

Methods: All patients > 18 years with a primary diagnosis of AMI were extracted from the National Inpatient Sample (NIS) from 2005-2010 using International Classification of Diseases (ICD-9) codes 410.x. Patients with type 1 diabetes mellitus and type 2 diabetes mellitus were identified using ICD-9 codes 250.x1, 250.x3 and 250.x0, 250.x2 respectively. Temporal trends and differences in in-hospital mortality, sudden cardiac death, and length of stay (LOS) were analyzed.

Results: A total of 1,344,095 patients were identified with AMI, of which 0.7% (n=9,812) had type 1 diabetes mellitus and 27% (n=363,174) had type 2 diabetes mellitus. Patients with type 2 diabetes mellitus were older (650.1 vs 57.30.3 years), more likely to be male (61% vs 51%) and had higher prevalence of hypertension (77% vs 66%), obesity (18% vs 11%), smoking (31% vs 23%) and hyperlipidemia (62% vs 50%) (p<0.0001 for all). In-hospital mortality (10% vs 7%), sudden cardiac death (5.2% vs 3.9%) and LOS (4.90.1 vs 3.90.1 days) were significantly higher in type 1 diabetes mellitus (p<0.0001 for all) with similar trends from 2005-2010 (Ptrend<0.0001 for all). Type 1 diabetes mellitus was associated with increased in-hospital mortality even after adjusting for demographic and risk factors {OR (1.75 [1.49-2.06]; p<0.001)}.

Conclusion: Type 1 diabetes mellitus was independently associated with increased in-hospital mortality and sudden cardiac death after AMI despite decreased prevalence of risk factors when compared to type 2 diabetes mellitus.
Unusual Complication of Acute Pyelonephritis: Transient Nephrotic syndrome

Introduction

Acute Pyelonephritis (AP) is a very common disease in which inflammation of the kidney is a complication of an ascending urinary tract infection. Symptoms are variable but most commonly include fever and flank pain. Early diagnosis with proper management can significantly impact the outcome and decrease the possibility of complications including recurrent infection leading to chronic pyelonephritis, abscess and acute renal failure [1]. Nephrotic syndrome is an unusual complication of AP in adults, with some reported cases in the pediatric patient population [2].

Case

A 37-year Asian female with a past medical history of hepatitis B which has been monitored as an outpatient by her PCP with a recent viral load of 172 IU/ml presented with vomiting, lower back pain, and malaise for three days. Vital signs were remarkable for blood pressure 92/50 mmHg, Heart rate: 102 beat/Minute, Temperature: 102F (oral), physical examination demonstrated right costophrenic angle tenderness. She was initially diagnosed with sepsis secondary to pyelonephritis, urinalysis demonstrated positive nitrite, large leukocyte esterase, and many WBCs. She was also found to have a leukocytosis with WBC of 17.4 K/uL and with Serum Creatinine of 1.8 mg/dl. Baseline Serum Creatinine was 0.5 mg/dl three months prior to admission leading to the diagnosis of acute renal failure has been made. Urine culture grew Enterococcus faecalis >100,000 cfu/ml. The renal Ultrasound showed a small stone with mild hydronephrosis of the right kidney. During the patient's hospitalization, her renal function declined, albumin level trended down to 2.1mg/dl (normal 3.4-5.4 mg/dl) with hypoproteinemia, and she subsequently developed anasarca with bilateral pleural effusion. Twenty-four-hour urine collection showed 3.5 g/d of proteinuria and her protein/creatinine ratio was 8.43.

Based on these findings, the diagnosis of nephrotic syndrome (NS) was considered at that time, Additional testing including ANA, ANCA, C3/C4, and Hep C Ab all yielded normal values. However, CRP was elevated at 9mg/dl. Renal biopsy was planned, but the patientâ€™s general condition improved with a few days with the appropriate antibiotics. The renal function normalized and edema resolved with an improvement in albumin level.

Conclusion

Acute renal failure is a possible complication of acute pyelonephritis, but nephrotic syndrome should also be considered during the assessment [5]. Nephrotic syndrome has been documented as a complication of chronic pyelonephritis in adults due to kidney scarring but never in the setting of acute pyelonephritis [6]. Our patient had transient nephrotic syndrome as a complication of acute pyelonephritis with negative workup for other causes of proteinuria and improved with the antibiotics treatment without steroids use. Renal biopsy may be considered to confirm the diagnosis of such cases, but would not change the outcome in this case.
Broken-Heart Syndrome; A fatal complication.

Introduction:

Takotsubo cardiomyopathy (TCM) or stress-induced cardiomyopathy (SICM) is a syndrome characterized by left ventricular dysfunction in the absence of significant coronary artery disease. This disease is usually benign, transient, and is usually treated conservatively with good prognosis. However, we are reporting a case of TCM with an uncommon presentation with a severe complication of ventricular rupture leading to death.

Case presentation:

A 77-year-old female with a history of hypertension and diabetes, presented to the ER with sudden syncope. She reported severe retrosternal chest pain, associated with nausea and vomiting. She stated she has been under extreme emotional stress due to sudden death of a family member. Examination was remarkable for hypotension (70/40 mmhg), cold clammy extremities, faint peripheral pulses, and diaphoresis. The ECG revealed ST-segment elevation in anterior-septal leads. Troponin level was elevated (3.390 ng/ml). Chest x-ray revealed enlarged cardiac silhouette. She was started on vasopressor for cardiogenic shock and underwent emergent cardiac catheterization which revealed non-occlusive coronary artery disease, severe anterolateral hypokinesis, apical dyskinesis, and apical ballooning with estimate LVEF of 20%. Takotsubo cardiomyopathy was speculated.

Over the course of the admission, her chest pain persisted which accompanied with severe back pain. The CT chest angiogram showed complex pericardial effusion compatible with hemopericardium. There was extravasation of contrast from the left ventricle into the pericardial space that was suspicious for ventricular wall rupture. The patient underwent emergent cardiothoracic surgery and was found to have left ventricular wall rupture which was repaired. The patient’s condition rapidly deteriorated and went into cardiac arrest. After extensive trials of cardiopulmonary resuscitation, the patient unfortunately expired.

Discussion:

TCM has been established as an entity in the past 30 years. Typically, it occurs in middle-aged female as a response to a stressful event, such as bad news, death, or natural disaster. TCM mimics acute myocardial infarction with electrocardiogram changes and elevated troponin.

This disorder may be caused by diffuse catecholamine-induced microvascular spasm or dysfunction, resulting in myocardial stunning, or by direct catecholamine-associated myocardial toxicity.

Ventricular wall rupture is a rare but significant complication. The risk factors for ventricular wall rupture in TCM include female gender, older age, and persistent ST segment elevation. Initial EKG findings of persistently elevated ST-segment in patients with TCM can predict a more serious course of the disease progression and provide a valuable parameter for risk stratification.
Resident/Fellow Clinical Vignette

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My Body Smells Fruity (Canagliflozin-Associated DKA in Type 2 Diabetes)

INVOKANA (canagliflozin) is a sodium-glucose cotransporter 2 (SGLT2) inhibitor causing osmotic diuresis, thus, promoting urinary glucose excretion reducing hyperglycemia. Diabetic ketoacidosis is a rare complication in diabetic patients treated with INVOKANA, more commonly diagnosed in type I than type II.

A 68-year-old male with a significant past medical history significant for type 2 diabetes mellitus presents to ED with complaints of polyuria, polydipsia, changes in urine odor, and a fruity smell in the body for 2 weeks. A clinical diagnosis of diabetic ketoacidosis was made. Patients Home meds include Canagliflozin. Pertinent labs showed BMP sodium 137 potassium 4.5 chloride 102 Bicarb 17 BUN 29 creatinine 1.59 glucose 288 LFTs normal.

Anion gap 18 serum ketones 4.8.Venous blood gas PH 7.27 Co2 38.7 PO2 62.1.

The patient was admitted to the ICU and diabetic ketoacidosis protocol was initiated as per hospital policy. After close monitoring of clinical status and blood chemistry, his anion gap closed and he was transitioned to subcutaneous insulin with remarkable improvement in his symptoms. The patient was started on basal insulin and metformin was continued. Canagliflozin was been discontinued. The patient was discharged to his home with follow-ups to the primary care physician.


DKA occurs more commonly in patients with type 1 diabetes, however, patients with type 2 diabetes are also at risk for developing DKA. Several factors can lead to DKA such as stress, surgery, medication noncompliance, infection and iatrogenic.

Euglycemic DKA is a rare, but reported side effect of canagliflozin in type 2 diabetics. Euglycemic DKA, is a DKA without hyperglycemia, defined as glucose levels <250 mg/dl. (7) This imposes a challenge to physicians in recognizing DKA in type 2 diabetics. In our patient, careful history taking, physical examination and blood chemistry aided in the prompt diagnosis and management of euglycemic DKA. SGLT-2 inhibitors documented benefits in decreasing mortality from cardiovascular causes, providing renal protection and improved blood glucose control. This side-effect should not deter a physician from prescribing those medications given the overall mortality and morbidity benefits. However, it is important to keep in mind the rare presentation of DKA in patients taking SGLT2 inhibitors. Finally, we recommend monitoring type 2 diabetes patients for signs and symptoms of DKA while taking SGLT-2 inhibitors.
Resident/Fellow Clinical Vignette

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THE SIMULTANEITY OF CORONARY ARTERY ECTASIA AND INTRACRANIAL DOLICHOECTASIA

INTRODUCTION

There have been a few documented cases in the literature that have reported the association between arterial ectasia in the two most vital organs of the body - the brain and the heart. In this study, we report a case of multiple hospital admissions presenting on account of transient ischemic attack (TIA) and angina-like symptoms. After extensive workup, the patient was found to have multiple coronary arterial ectasias and aneurysms together with multiple intracranial arterial dolichoectasias and aneurysms. The complexity of this case and the associated co-morbidities complicate the management protocols.

CASE

60-year-old female presented with a mild left chest pain which radiated to the left arm, EKG showed T-wave inversion in V4 to V5 and mild elevation of Troponin, Echocardiology showed no left ventricular wall motion abnormalities. Nuclear Stress Test showed moderate inferior and inferolateral ischemia with partial scarring. Coronary Angiography showed severe ectasia and stenosis of some segment of LM, LAD, OM1 and distal LCX and bilateral subclavian arteries. No stent was placed. Two weeks later, she presented with dizziness with no acute neurological deficit. Head imaging showed severe intracranial dolichoectasia involving the vertebrobasilar system, intracranial internal carotid arteries, and bilateral middle cerebral arteries. The patient was evaluated by Neurosurgery and Neuroendovascular teams and both endorsed the patient is not good candidate for any neurovascular intervention because the complexity of the finding.

DISCUSSION

Coronary artery ectasia is an abnormal dilatation of a coronary arterial segment of at least 1.5 times of a normal coronary artery. CAE is found in 3±8% of patients undergoing coronary angiography. The slow flow phenomenon may lead to ischemia and thrombosis. These may then lead to ACS. Dolichoectasia composes a dilatation and elongation of the arteries which affects both the anterior and posterior cerebral circulation. It may cause neurological complications like ischemic stroke, intracranial hemorrhage, or compression of surrounding neural structures. A rarefaction of elastic tissue of the media with degeneration of the internal elastic lamina, as well as matrix metalloproteinases dysfunction are a common pathological explanation for this condition. The Management of CAE and IADE is mostly conservative, in both conditions, it is essential to treat the risk factors and administrating of antiplatelet and anticoagulant agents. Angioplasty and stent could be used for some indicated patient of CAE while surgical treatment could be applied for some patient of IADE like; ventriculoperitoneal shunt, microsurgery, (STA-SCA) bypass, proximal balloon occlusion, or thrombectomy.

CONCLUSION

The concurrence between CAE and IADE is rare entity and should be studied more prospectively to establish specific guideline in the management of these cases.
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The deadly gas lurking within: a case of emphysematous pyelonephritis.

Emphysematous pyelonephritis is a life-threatening condition defined as severe necrotizing infection and presence of gas in the renal parenchyma or its surrounding tissue.

We present a case of a 63-year-old male with a medical history of type 2 diabetes mellitus on insulin (HA1C 12.3), ischemic stroke with residual left sided weakness, and chronic kidney disease who presented following a fall in the setting of fever and lethargy for 2 days prior to presentation. He also described urgency and frequency of urination however denied dysuria or flank pain.

Upon presentation his BP 86/54, heart rate 122, temperature 38.4°C, respiratory rate 18. Exam was notable for lethargic, obese middle-aged man, tachycardic but regular rhythm, lungs were clear to auscultation, abdomen was soft, non-tender with normal bowel sounds, negative renal percussion pain, and left upper and lower extremities with strength of 2/5.

Labs with leukocytosis of 16.2 (neutrophils 93%), hemoglobin 13.8, platelets 159, lactic acid 4, creatinine 2.5 (baseline 1.1), BUN 38, glucose 450, bicarb 25. Urine analysis: 3+ blood, 2+ protein, 3+ leuk esterase, and pyuria.

A CT abdomen and pelvis was performed and revealed diffuse left kidney hypodensity representing pyelonephritis with foci of air on the cortex possibly representing emphysematous infection without hydronephrosis.

He initially was treated with fluids resuscitation, vancomycin and piperacillin/tazobactam with improvement in his blood pressure however his kidney functions continued to worsen (creatinine 4.5) and patient became anuric. The decision was made to initiate hemodialysis.

Urine and blood cultures subsequently grew pan sensitive Klebsiella pneumonia and despite all supportive measures and IV antibiotics he continued to spike fevers with persistent leukocytosis and bacteremia. Repeated CT abdomen and pelvis showed frank emphysematous pyelonephritis of the left kidney with significant worsening and perinephric fat stranding with scattered gas. Urology was contacted and he underwent an urgent left nephrectomy. Biopsy showed acute pyelonephritis with extensive necrosis, marked acute inflammation and abscess formation.

Kidney function gradually improved post-operatively and he was discharged after 21 days off renal replacement therapy.

Discussion: Emphysematous pyelonephritis predominantly affects female diabetics. Urinary tract obstruction and immunosuppression are other predisposing factors.

Escherichia coli is the most common pathogen, followed by Klebsiella pneumonia and Proteus mirabilis.

CT scan is the modality of choice in the diagnosis and classification. Most can be managed with antibiotics and percutaneous drainage however patients who fail medical management or with extensive disease should be managed with nephrectomy.

We add to the literature a new case of uncontrolled type 2 diabetic male who presented with complicated urinary tract infection in the setting of emphysematous pyelonephritis due to Klebsiella pneumonia that was managed by nephrectomy and required temporary hemodialysis who showed a remarkable recovery.
Marked elevation of liver enzymes secondary to ceftriaxone use

Background: Acute hepatitis with marked elevation of liver enzymes is usually secondary to acetaminophen, hepatotoxins, acute viral hepatitis, autoimmune disease or vascular occlusion. Because of the infrequency of ceftriaxone usage causing acute liver injury, it is important to be cognizant of this possible adverse reaction.

Case presentation: 77 year old male with a past medical history of childhood polio, atrial fibrillation on dronedarone, heart failure, coronary artery disease status post coronary artery bypass grafting, recently diagnosed infectious endocarditis of bio prosthetic aortic valve with severe aortic stenosis was sent to the hospital from the cardiologist office for evaluation of elevated liver enzymes a week after starting a 6-week course of ceftriaxone 2g for Streptococcus endocarditis. During that visit, the patient reported anorexia with weight loss and mild epigastric discomfort. Physical exam was significant for jaundice, but no abdominal tenderness or signs of hepatic encephalopathy. Subsequent labs were significant ALT 1523, AST 1726, ALP 1160, bilirubin total 3.2, direct 2.7, indirect 0.5, INR 5.06, PT 56.01. Inpatient workup was negative for viral hepatitis and autoimmune hepatitis. CT scan showed cholelithiasis without biliary duct obstruction, hepatic necrosis or hepatic vein thrombosis. Ceftriaxone was stopped and the patient was started on penicillin G for endocarditis. Patient’s liver enzymes started to trend down. The INR and bilirubin, on the other hand, continued to uptrend, peaked on hospital day 5, and then down trended afterwards. AST and ALT normalized with mild elevation of ALP and bilirubin one month after discontinuing ceftriaxone.

Discussion: Parenteral ceftriaxone usage has been associated with biliary involvement with the biliary sludge in 3% to 46% of patients, of which only 5% to 10% of patients develop biliary colic. Even with symptoms, it is rare to have elevated serum enzymes or bilirubin levels. In our patient, he has been using dronedarone for 7 months with normal liver function tests, was not in hypotensive shock. He denied recurrent fevers, usage of acetaminophen or herbal medicine, or mushroom ingestion. Viral serology, ANA and ASMA were negative. A liver biopsy is not necessary unless there are diagnostic uncertainties. In this case, patient was on anticoagulation for left atrial thrombus, atrial fibrillation and a recent splenic infarct. Hence, the cause of acute liver injury was a diagnosis of exclusion and presumed to be due to ceftriaxone usage.

Conclusion: Acetaminophen usage is the most common cause of acute liver injury in the United States. However, medications such as ceftriaxone should be in the differential diagnosis of acute hepatitis in patients with no other apparent causes.
A NEGLECTED SYNDROME WITH GRAVE PROGNOSIS: NEPHROGENIC ASCITES

Introduction

Nephrogenic ascites is a clinical condition of refractory ascites seen in patients with ESRD on Renal Replacement Therapy. This entity was first described in 1970. The underlying etiology could be multifactorial including a combination of poor nutrition, inadequate dialysis and ultrafiltration, increased peritoneal membrane permeability and overall uremic state. Nephrogenic ascites is a rare syndrome and is often associated with a grave prognosis especially if it is not diagnosed early. Herein we describe a patient who presented to the emergency department with refractory ascites of nephrogenic origin.

Case

Patient is a 66 year old female with past medical history of DM I (diagnosed at age 11), ESRD secondary to diabetic nephropathy on hemodialysis (diagnosis 12/2017), bilateral diabetic retinopathy, ovarian cyst, hypertension, and anxiety who presented to the emergency department for evaluation of intractable abdominal pain, nausea and vomiting started 2 days. She was found to have large ascites. Diagnostic paracentesis was done and found to be exudative with Serum Ascites Albumin Gradient of 0.7. After detailed work-up, hepatic, cardiac, infectious and malignant causes for ascites were rule out. The diagnosis ascites of nephrogenic origin was made.

Discussion

This case represents a medical condition known as dialysis-related ascites, a problem that has been seen in ESRD patients on HD. Pathogenesis includes elevated Hepatic vein hydrostatic pressure, changes in peritoneal membrane permeability secondary to uremia, Obstruction of lymphatic channels caused by inflammatory infiltrate, Other predisposing factors could be hypoalbuminemia, hyperparathyroidism, congestive heart failure, constrictive pericarditis, pancreatitis, cirrhosis with portal hypertension. Low Serum Ascites Albumin Gradient is the hallmark of this syndrome. Detailed evaluation is required to rule out hepatic, cardiac, infectious and malignant causes of ascites. Management of nephrogenic ascites is complicated and includes Salt and fluid restriction, intensive hemodialysis with ultrafiltration and intravenous albumin infusions with a high protein diet, CAPD, Peritoneovenous shunt. Renal transplant is the only definitive treatment for nephrogenic ascites

Conclusion:

Nephrogenic ascites is a rare condition of grave prognosis. The etiology is unknown but it thought to be multifactorial. The diagnosis is made and this syndrome is differentiated from other causes of ascites by low Serum Ascites Albumin Gradient (<1.1). Aggressive renal replacement therapy along with nutritional support could be helpful, but renal transplant is the only definitive treatment option with complete resolution of the ascites.
Ana B. Arevalo M.D

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LUPUS NEVER FAILS TO DECEIVE US: A CASE OF ROWELL’s SYNDROME

Introduction: Rowell’s syndrome is comprised of the presentation of erythema multiforme-like lesions (EM) in association with lupus erythematosus (LE), along with serologies of speckled antinuclear antibodies (ANA), positive rheumatoid factor (RF), positive anti-La/anti-Ro, and the clinical finding of chilblains. As per the redefined criteria by Zeitouni et al, three major criteria in addition to at least 1 minor criteria are necessary for diagnosis. This reports the case of a previously healthy 20-year-old male who presented with EM-like skin lesions, as well as positive RF, speckled ANA, and positive anti-Ro.

Case presentation: A 20-year-old male presented with a one-week history of worsening non-pruritic erythematous maculopapular skin rash which appeared on the face and subsequently spread to the trunk, arms, legs, palms and soles. At the onset of rash, the patient reported headaches, associated with photosensitivity and intermittent fevers. Workup for viral meningitis yielded negative results. He recovered spontaneously with slight resolution of the rash and as a result refused skin biopsy. One month later, he presented once again with worsening of the rash as well as intermittent fevers. Examination revealed generalized well-defined, non-blanching confluent papules and plaques (resembling EM) in various stages of healing. Lesions were located on the face, trunk, arms, legs, as well as the palms and soles. Small areas of fine scaling were noted on the forearms and face. There was no mucosal involvement. Laboratory investigation revealed mild anemia (Hb 12.6 g/dL), elevated Erythrocyte Sedimentation Rate (ESR) of 75 mm/h, C-Reactive Protein (CRP) of 11.05 mg/L, a positive ANA titer of >1:1280 with speckled pattern, a positive anti-Ro/SSA, anti-La/SSB titers of >8.0, and a positive RF of 34 IU/ml. Lupus anticoagulant antibody was positive (>44.4 seconds), along with a low positive anticardiolipin IgM antibody (>13 U/ml). Anti-dsDNA, anti-Jo-1, anti-centromere, and anti-Scl-70 antibodies were negative. Serum chemistry and urinalysis were within normal limits. Hepatitis B virus (HBV), Hepatitis C virus (HCV), Human Immunodeficiency Virus (HIV), and Parvovirus B19 were negative. The patient underwent a left arm skin biopsy which demonstrated vacuolar interface dermatitis and positive colloidal iron stain suggestive of dermal mucin deposition, favoring the diagnosis of cutaneous collagen vascular disease. Cutaneous lesions improved with administration of oral prednisolone.

Discussion: This case highlights the importance of maintaining a diagnostic suspicion for Rowell’s syndrome in LE patients who present with EM-like lesions. Lupus anticoagulant antibody was positive in our patient but he did not meet criteria for Antiphospholipid syndrome (APS). It should be noted, however, that a rare association between APS and Rowell’s syndrome has been described.

Conclusion: Patients with cutaneous LE may develop coincidental EM. However, if characteristic serological abnormalities are present and there is no obvious precipitation factor, the association is known as Rowell’s syndrome.
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POWASSAN VIRUS ENCEPHALITIS, SEVERE BABESIOSIS AND LYME CARDITIS IN A SINGLE PATIENT

INTRODUCTION: Ixodes scapularis is responsible for the transmission of Borrelia burgdorferi, Babesia microti, Anaplasma phagocytophilum and Powassan virus to humans. The coexistence of dual pathogens occurs in 28% of ticks in North America. However, infection with Powassan virus is extremely rare. To the best of our research, this is the first case describing co-infection with Powassan virus, Babesia and Borrelia together.

CASE: 87 years old male presented with one-day history of high-grade fever with rigors and chills, in the background of two weeks of worsening lethargy, lightheadedness, and mild abdominal pain. On exam, the patient was somnolent but responsive to pain. Labs revealed hemolytic anemia (Hb 10.9 g/dl, hematocrit 30%, reticulocyte percentage 11.1%, LDH 1200 units/L, haptoglobin <10 mg/dl, indirect bilirubin 2.6 mg/dl and total bilirubin 3.4 mg/dl), thrombocytopenia (44,000/µl), leukocytosis (13,000/µl) with raised ESR 70 mm/hr and CRP 210 mg/L, lactic acidosis (5.6 mmol/L), hepatitis (ALT 134, AST 309), and acute kidney injury (Cr 1.8 mg/dl). The patient was empirically started on Doxycycline for Lyme and Atovaquone and Azithromycin for Babesiosis. In the next few days, Babesia serology and PCR returned positive, parasitemia was 11.9%, Lyme IgM and IgG were positive. The patient underwent exchange transfusion twice. His clinical condition improved significantly. However, on Day 9, he developed rapid deterioration in mental status with prominent myoclonus, and respiratory failure warranting intubation. CT and MRI brain was unremarkable, EEG showed generalized slowing. LP done for concern of neuroborreliosis showed WBCs 20/µl with 96% lymphocytes, 0% neutrophils, glucose 60 mg/dl and protein 48 mg/dl. Doxycycline was switched to IV Ceftriaxone. CSF PCR for Lyme returned negative but Arbovirus serology was positive for â€œPowassan Virusâ€€ (Reactive Powassan E Polyvalent Microsphere Immunofluorescence Assay). On Day 10, the patient developed AV block likely from Lyme carditis, so temporary transvenous pacemaker was placed. He was continued on supportive management, IV Ceftriaxone, and completed Azithromycin and Atovaquone course. On day 18, patient was extubated and the pacemaker was retrieved. However, patient demonstrated persistent vocal cord dysfunction and dysphagia with high aspiration risk. On day 28 of admission, the patient was discharged to a nursing facility with an NG tube and would be re-evaluated for gastrostomy tube placement.

DISCUSSION: In endemic areas, patients who fail to respond to treatment for common tick-borne illnesses, with the presence of neurological findings, testing for arboviruses like Powassan virus should be considered. Powassan virus has a high risk of permanent neurological deficits and long term rehabilitation is necessary. For severe Babesiosis, more studies are needed to elucidate whether there is a difference between the efficacy of Azithromycin plus Atovaquone and Quinine plus Clindamycin. Identification of arrhythmias in Lyme carditis and prompt treatment with medications or pacemakers could be life-saving.
Hypercalcemia in a patient diagnosed with a Vasoactive intestinal peptide tumor.

Abstract

Hypercalcemia is a relatively common clinical problem and is most commonly seen in patients with primary hyperparathyroidism and malignancy. Hypercalcemia, if seen in patients with neuroendocrine tumors (NETs) is typically attributed to hyperparathyroidism seen in multiple endocrine neoplasia type 1. This case will examine an unusual presentation of hypercalcemia in a patient diagnosed with a vasoactive intestinal peptide tumor without hyperparathyroidism.

Case

A 22-year-old female with celiac disease, osteopenia, and depression presented with a two-month history of worsening diffuse abdominal pain, diarrhea, and arthralgia. The patient denied fever, chills, unintentional weight loss or a history of recent travel. The patient was admitted for evaluation and treatment of severe hypercalcemia. Patient was treated with intravenous normal saline and correction of electrolytes. An abdominal ultrasound revealed two heterogeneous masses in the right hepatic lobe of the liver measuring 7.4 cm and 8.5 cm. MRI confirmed a large, solid, pancreatic tail lesion with liver metastases. iPTH (intact parathyroid hormone) and PTHrP (parathyroid hormone related peptide) were normal. VIP level > 900 pg/ml (normal 0-60 pg/ml). The diagnosis of hypercalcemia attributed to malignant pancreatic VIPoma with liver metastasis was established. Shared decision making led to conservative treatment with calcitonin and a bisphosphonate, surveillance, and outpatient follow up.

Discussion

In addition to the symptoms of watery diarrhea, hypokalemia, and achlorhydria classically seen in patients diagnosed with a VIPoma, our patient also presented with a significant hypercalcemia. This was likely secondary to bone resorption as a result of the PTH like activity of VIP. MEN 1 is another rare cause of hypercalcemia in these patients. Diagnosing and appropriately treating hypercalcemia in patients diagnosed with VIPoma is essential to avoid serious complications. Fluids, calcitonin, and bisphosphonates are all useful in controlling the high calcium levels. Although we would expect these patients to present with constipation due to their high calcium levels, the significantly elevated VIP levels counteract the mild to moderate elevation in calcium leading to the classic WDHA syndrome we commonly see.
Hereditary Thrombotic Thrombocytopenic Purpura (TTP) masquerading as Pre-eclampsia

Introduction: Hereditary TTP is a thrombotic microangiopathy caused by a congenital deficiency of Von Willebrand factor (VWF) cleaving protease ADAMTS13. Hereditary TTP is an extremely rare autosomal recessive condition with a prevalence of 1 in a million. We present a young female with a history of recurrent thromboembolic events since early adulthood and an episode of TTP during third trimester which presented as pre-eclampsia. She was subsequently diagnosed with Hereditary TTP after she presented with acute stroke.

Case report: A 35 year old female presented to the emergency room with acute left sided weakness in upper and lower extremities. She had a history of recurrent embolic strokes and deep vein thrombosis (DVT) since adulthood. At the age of 24 years, during her 3rd trimester of pregnancy, she developed uncontrolled hypertension, petechiae and epistaxis due to symptomatic thrombocytopenia (platelet count 3,000/µL), acute kidney injury and proteinuria. Due to concerns for pre-eclampsia, she underwent an emergency Cesarean Section. Post delivery, thrombocytopenia and renal injury did not resolve which prompted ADAMTS13 levels to be checked and were found to be low; consistent with a diagnosis of TTP. She was started on plasma exchange and rituximab and eventually weaned off. Over the next 8 years, she had multiple episodes of transient ischemic attacks (TIA) and DVT despite being on anticoagulants. She tested negative for anticardiolipin antibodies, prothrombin gene mutation, factor V Leiden, deficiencies of protein C and S and antithrombin III. She underwent closure of patent foramen ovale at 33 years.

During the current admission, computed tomography angiography of the head/ neck and magnetic resonance imaging of the brain showed no acute infarct or vessel occlusion. Pertinent laboratory findings were platelets 90,000/µL with normal white cell count and hemoglobin. Electrolytes and creatinine were unremarkable. Doppler studies were negative for DVT. ADAMTS13 levels were found to be low and ADAMTS13 inhibitor levels were undetectable. She was diagnosed with hereditary TTP and started on plasma transfusions every 2 weeks. Patient is currently doing well with no further thromboembolic episodes.

Discussion: Hereditary TTP constitutes < 5% of all TTP cases and presents with a long history of arterial and venous thromboembolic events. Diagnosis is confirmed with severe ADAMTS13 deficiency (<5% activity) in the absence of an inhibitor. The specific ADAMTS13 gene mutation is detected with molecular analysis. Females often present during third trimester or postpartum period of their first pregnancy because VWF levels are increased late in pregnancy. If untreated, pregnancies usually end in spontaneous abortion, stillbirth or premature delivery. One in four patients who develop TTP during pregnancy are diagnosed with Hereditary TTP. Once the diagnosis is established, the patient needs regular plasma infusion every 2-3 weeks to prevent further thromboembolic events and renal failure.
Atypical Presentation of Gastro-gastric Fistula Years after Gastric Bypass.

A 55-year-old Hispanic Male presented to emergency room with multiple bouts of hematemesis after suffering from epigastric pain, and nausea for 3 days. He had no history of NSAID use, peptic ulcer or cirrhosis. His past history was remarkable for gastric bypass for morbid obesity in 2001, and chronic alcoholism. Physical examination was significant for tachycardia, obesity, and hematemesis. Hemoglobin was 12.2 g/dl (baseline 14.7). He received one unit of blood and intravenous pantoprazole infusion. Emergent esophagastroduodenoscopy revealed large blood clots in the stomach and an ulcer at anastomotic site. Repeat upper endoscopy on day 3 showed a gastro-gastric fistula (GGF) connecting gastric pouch to bypassed stomach (Figure 1). 1cm clean based ulcer at mouth of fistula (Figure 2) appeared to cause the initial bleeding. Patient felt well at one-month follow-up, and declined surgical closure of fistula. Biopsy result did not show H. pylori, or neoplasia.

Bariatric surgeries are fairly commonly performed on morbidly obese patients. They cause weight loss by gastric reduction, and malabsorption, thereby preventing and/or reversing type II diabetes. They are mostly performed laparoscopically now-a-days. The Roux-en-Y operation, often called gastric bypass, is the most popular. Gastro-gastric fistula is a rare but important complication of it. With the formation of fistula, gastric pouch opens into the bypassed portion of stomach, thus food once again channels the natural route which ultimately, leads to weight regain, and procedure failure.

The incidence of gastro-gastric fistula is reported at 1.2%-2.6% in the published literature1, 2. Majority of patients (77%-80%) presented with epigastric pain with or without associated marginal ulcer (53%). 44% patient had weight regain2, 3. Gastrointestinal bleeding, and vomiting were the chief complaints in 11% of cases, respectively3. Gastro-gastric fistulae are classified based on their location. Type 1 GGF are located in the proximal part of the gastric pouch and type 2 are near the anastomosis3. Upper endoscopy and contrast imaging is used confirm the fistula. Most early post-op fistulae can be successfully managed endoscopically4. Chronic fistulae are difficult to manage3. Optimal surgical management is controversial. Surgery is considered in case of weight regain, recurrent, or non-healing marginal ulcer with persistent abdominal pain and/or hemorrhage, or recurrent anastomotic stricture5. Laparoscopic remnant gastrectomy with fistulectomy is an option for symptomatic patients if conservative measures fail5.

This case report underscores the importance of maintaining a high index of suspicion for gastro-gastric fistula following gastric bypass, especially in patients with weight re-gain. A focused approach is needed after weight loss surgery since upper gastrointestinal symptoms can be difficult to interpret in these patients. Trainee Endoscopists have to bear in mind that upper endoscopy can be challenging due to the altered anatomy as a result of bariatric surgery.
Hearing loss as initial presentation of Granulomatosis with Polyangiitis: Clinical vignette and review of the literature

Introduction: Granulomatosis with polyangiitis is an antineutrophil cytoplasmic antibody (ANCAs)-associated vasculitis (AAV). AAV is a heterogeneous group of diseases that cause inflammation of the wall of small to medium sized blood vessels. Diagnosing AAV is a challenge as the clinical manifestations vary widely. Lungs and kidneys are the most often involved and when present together often prompt investigation for AAV. However, unusual organ involvement creates a particular diagnostic challenge. Herein, we present a case of GPA presenting with hearing loss, causing a significant delay in the diagnosis and treatment. We also reviewed the English literature from 2000 to 2019 and analyzed similar reported cases.

Case presentation:

A 67 year old female with a past medical history significant for coronary artery disease presented to our ER complaining of a myriad of symptoms over the past 3 months. She initially developed bilateral hearing loss for which she was seen by ENT. Audiometry testing revealed bilateral sensorineural hearing loss and she was treated with prednisone for presumed autoimmune pathology. One month later, she was admitted to an outside hospital for community acquired pneumonia. CT chest showed multiple pulmonary nodules and she was scheduled for an outpatient biopsy. A few weeks later, she developed severe malaise and fatigue prompting her to come to our ER. Labs showed creatinine of 3.81 mg/dl (baseline normal), elevated C-reactive protein 124 mg/L (reference range 0-3 mg/L), elevated erythrocyte sedimentation rate 120 mm/hour (reference range 0-20mm/hour), white blood cells of 17,000/mm, urine analysis showed active sediment (RBC cast and hematuria). Patient received pulse steroids 1 gram/day for 3 days. She underwent kidney biopsy showing pauci immune necrotizing glomerulonephritis. Anti-proteinase-3 antibody was positive with cytoplasmic pattern on immunofluorescence. Cyclophosphamide was added to treatment regimen. Renal function improved to creatinine 1.2 and hearing normalized.

Discussion:

Granulomatosis with polyangiitis (GPA) is a rare ANCA vasculitis. It typically affects the upper and lower respiratory tracts and kidneys (1). Otologic manifestations can predate other signs causing a delay in diagnosis and treatment. The mortality and morbidity of AAV are high if left untreated or if there is a delay in treatment (1). We reviewed the English literature between 2000 to 2019 for any reported cases similar to our case presentation (Table 1). 12 articles were identified which described 23 cases of ear involvement as the initial presentation of ANCA vasculitis. The majority of patients had GPA with positive ANCA antibodies and mixed sensorineural and conductive hearing loss. All cases demonstrated delay in the diagnosis and treatment of AAV.

Conclusion:

Otologic manifestations of ANCA vasculitis present a diagnostic dilemma and can lead to a fatal disease if left untreated. Early recognition and clinical suspicion should be maintained to avoid a delay in treatment.
Introduction:

We report a unique case of bacteremia and meningitis due to Streptococcus salivarius in a patient who presented with fever, headache and seizure. Viridians streptococci accounts for very small percentage of purulent meningitis. Cases of S. salivarius meningitis were reported previously after spinal anesthesia, neurosurgical and GI procedure. Dental procedure was considered the source in our patient.

Case Report:

A 43 year old female with history of seizure initially presented with fever, headache, cough and shortness of breath. She was discharged from hospital twice with oral antibiotics for pneumonia a week prior. On her third visit to the emergency department patient had three episodes of seizures. At admission two sets of blood culture were sent and empiric antibiotics started with Piperacillin/Tazobactam and Vancomycin. CT scan of head showed no acute abnormality and chest x-ray was normal on admission. Patient was adherent to all her home medications. Blood cultures showed gram positive cocci in pair and chains less than 24 hours. At the same time, patient started high spiking temperature with worsening headache and lethargy. Lumber puncture was done. Pip/Tazo was changed to meningal dose ceftriaxone, Vancomycin continued and dexamethasone was added for possible pneumococcal meningitis. CSF was cloudy and cell count showed typical feature of bacterial meningitis, WBC of 5833 with 85% of PMN. Within 24hours the patient became more awake, the headache improved and the fever defervesced. Upon taking further history, patient reported that she had her dental workup done a week prior to these presentations. CT scan of sinuses revealed periapical lucency adjacent to the roots of the posterior most left maxillary tooth. Transesophageal echocardiogram was negative for endocarditis. Two sets of blood culture grew S. Salivarius, sensitive to vancomycin and penicillin.

Discussion:

Viridians streptococci are well known to cause endocarditis but rarely cause meningitis. Cases of meningitis have been described related to the breach in mucus membrane of oral cavity, sinus tract, gastrointestinal tract, urogenital tract or skin. Although S. salivarius in the absence of symptoms and signs of infection could be considered a contaminant, in our patient it was pathogenic. Several doses of antibiotics were administered intravenously prior to the lumbar puncture resulting in a negative CSF culture in the presence of 5833 WBC with 85% Neutrophils. The patientâ€™s recent dental work up and the presence of a periapical lucency in the posterior most left maxillary tooth lends credibility to the fact that this likely was the source of the Strep. Salivarius bacteremia resulting in meningitis.
A case of Hyperhemolysis Syndrome in the Setting of Sickle Cell Disease

Hyperhemolysis syndrome (HS) is a rare and potentially life threatening complication of sickle cell disease. It is a complication of transfusion therapy, which is often required in sickle cell. HS is characterized by the development of severe anemia with post transfusion hemoglobin lower than pretransfusion hemoglobin, intravascular hemolysis, and hemoglobinuria leading to multiorgan failure and ultimately death. We present a case of hyperhemolysis syndrome in a 19 year old male in the setting of sickle cell disease. Patient was a 19 year old male with past history of HbSS positive sickle cell disease. He presented to Emergency Department after a recent discharge for a sickle cell pain crisis returning with severe pain. Patient was admitted to pediatric floors with pain control, but had to be transferred to the pediatric ICU for acute chest syndrome. Patient’s hospital course complicated by frank hematuria. Patient’s hemoglobin dropped to 6.1, and was transfused 1 PRBC. Patient’s hospital course complicated by frank hematuria. Post transfusion, hemoglobin dropped to 4.0. It was suspected patient was experiencing hyperhemolysis syndrome. Patient was started on Methylprednisolone and IVIG as prophylaxis for hemolysis in the event the patient needed further transfusions. Patient started to deteriorate becoming hypertensive, tachycardic and tachypneic, so he was transferred to Medical ICU. Due to hyperhemolysis crisis, transfusions had to be held and blood draws were limited. Patient was started on presser support to maintain BP. Renal function started down trending, so was started on continuous renal replacement therapy. LDH at the time was >34000. Patient Hb dropped to 2.4. Patient went into DIC. Patient was resuscitated, but family elected to cease all medical intervention as prognosis was very poor. Patient was extubated, all medications stopped and patient expired shortly after. Hyperhemolysis syndrome is caused by the destruction of both recipient and donor RBC’s. The mechanism by which these RBCs lyse is still not understood very well. Most cases of hyperhemolytic crisis are probably in the setting of occult splenic sequestration or aplastic crisis. However, our patient, who required transfusions, may have experienced “bystander hemolysis”. A phenomenon consistent with a delayed hemolytic transfusion reactions in which transfused cells and patient’s own cells are hemolyzed. The sickled RBCs are destroyed by antibodies without expressing the specific antigen, leading to worsening anemia.
Resident/Fellow Clinical Vignette

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Respiratory Disease in Migrant Farmworkers

Abstract:
There are limited data on the respiratory status of Hispanic farmworkers in the US. In response to recent changes in Environmental Protection Agency (EPA) regulations, NYCAMH has evaluated Latinx farmworkers (FW) for OSHA Respiratory Protection programs across NY. Health data were extracted from all standardized Spanish (SP) language OSHA questionnaires completed 2017-19. The best of three peak flow (PF) efforts was compared to accepted normal values. 142 questionnaires contained all key information. NY FW do not appear to have abnormal rates of respiratory symptoms or low peak flows. Records from OSHA screenings can provide valuable data for evaluating the health of this population of workers.

Background:
Due to recent changes in EPA regulations, workers exposed to pesticides need to be included in an OSHA-compliant respiratory protection program, which involves:

- detailed health questionnaire
- vital signs
- test of pulmonary capacity (“spirometry or peak flow measurement

The purpose of this study is use these data to assess the respiratory status and function of migrant farmworkers.

Methods:
Extract data from NYCAMH OSHA questionnaires from across NY; January 2017 – March 2019.

To date:
- Data from all complete SP language questionnaires was entered into REDCap
- Best of three recorded peak flows entered into SAS
- Normal PF values derived from regressions using age/gender/height

Results:
142 questionnaires assessed: 97.5 % males, 2.5% females
11.7% smokers
Respiratory Symptoms:
- 2.4 % Any SOB
- 0 % SOB at rest
- 3.0 % Any Cough
- 0 % Cough & phlegm
- 0 % Asthma
- 0 % Wheeze
Peak flow findings: 13% abnormal, 87% normal.
Mean % predicted PF 97.32%.

Conclusions:
To date:
- Rates of smoking are 63% that of the normal US population
- Rates of respiratory symptoms do not appear to be elevated
- PF’s measured in the work place appear to be mildly reduced. This could relate to the choice of reference values.
- OSHA Respiratory Protection Program data may provide useful insight into rates of respiratory problems and other health issues affecting Latinx FW in NY State.
- Data collected relies on the assumption that the FM fully understood the questionnaires
Hyponatremia: A Complication of Trans-Sphenoidal Pituitary Resection

A triphasic water response is seen in patients who undergo trans-sphenoidal surgical resection of a pituitary tumor. This consists of diabetes insipidus (DI) within the first 24-48 hours, followed by the syndrome of inappropriate antidiuretic hormone (SIADH), and a final phase of persistent DI. We describe a patient who had trans-sphenoidal surgery for a pituitary microadenoma, and presented with hyponatremia secondary to SIADH.

Our patient is a 65 year old woman with a history of fibromyalgia, irritable bowel syndrome, hyperlipidemia, and a pituitary adenoma over-secreting insulin-like growth factor 1 (IGF-1) leading to acromegaly. The patient underwent trans-sphenoidal surgery for the pituitary microadenoma. One day after discharge on post-operative day (POD) three, she presented to the Emergency Department (ED) with hypertension, serum electrolytes were normal, and she was discharged. On POD seven, she complained of generalized weakness and was brought into the ED via ambulance. She was severely hyponatremic; Na level of 117 only four days after being discharged from the ED with normal bloodwork. During this one-week interval, she also experienced excessive thirst secondary to dry mouth due to nasal packing. On admission, her urine Na was 82 which is indicative of SIADH. This guided the clinical diagnosis of SIADH and treatment with fluid restriction after the patient received hypertonic saline leading to a repeat urine Na of 21 on day three of admission and complete resolution. Sodium levels must be increased appropriately, not to exceed an increase of 10-12 mEq/24 hours, to avoid central pontine demyelination. The patient was then transitioned to restricted fluid intake of 1200ccs. Within four days she was discharged with a Na of 136 and an uneventful recovery.

Fisher and others described a now classic, triphasic trend of serum sodium abnormalities seen after transection of the infundibular stalk in both animals and humans; DI within the first 24-48 hours, followed by the SIADH, and a final phase of persistent DI. The triphasic response is quite a rare occurrence as published literature details this presentation in approximately 1-2% of cases. In a study by Baskin et al, the majority of cases of hyponatremia following pituitary surgery are solely SIADH-related (71%). As SIADH is prevalent for majority of the triphasic response, when patients present to the ED, clinicians usually observe contrarian features such as volume depletion and can only determine the triphasic response through a meticulous history from the patient.

Our patient was able to recall many of the symptoms she underwent in the week following the surgery. In addition to the history and patient recollection, interpretation of laboratory work and careful consideration of the triphasic response allowed the care team to diagnose SIADH in the absence of normal clinical features in relation to said diagnosis.
Resident/Fellow Clinical Vignette

Andrew Castellano

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A FORGOTTEN CASE OF SJOGRENS RELATED PERICARDITIS

Sjogren’s syndrome is an autoimmune disease often defined by lymphocytic infiltration of the salivary and lacrimal glands; however, subsequent infiltration of visceral organs can also occur. Overt heart involvement in primary Sjogren’s syndrome is rare, but when present, there is a high incidence of pericarditis and diastolic dysfunction seen on echocardiogram.

A 53-year-old male with a past medical history of morbid obesity, diabetes mellitus type II, and chronic fibrous pericarditis with pleural effusion of unknown etiology status post subxiphoid pericardial window five months ago presented to our Emergency Department with a chief complaint of worsening gripping chest pain, aggravated when taking deep breathes or lying supine. Of note, for the past five months the patient had been taking indomethacin for this recurrent pericarditis. On initial presentation the patient was afebrile, tachycardic, and tachypneic and the patient was admitted for recurrent pericarditis of unknown etiology. Echocardiogram showed an ejection fraction of 66% and trace circumferential pericardial effusion. Infectious etiologies including Parvovirus B19, Coxsackie, Mycobacterium Tuberculosis, and bacterial cultures were negative. It was however noted on chart review that the patient had positive antinuclear antibodies (ANA) on his initial presentation months prior and remained elevated with an ANA titer of 1:640. Additional rheumatologic tests were ordered which revealed a positive Rheumatoid Factor of 50 IU/mL and subsequently positive for Sjogren’s antibody anti-Ro (SSA). Resultantly, our patient was started on Corticosteroids and Ibuprofen. He experienced a resolution of chest pain, and was discharged from the hospital with a steroid taper and appropriate follow up.

Discussion

Relapsing pericarditis is most often attributed to an idiopathic or viral etiology. However, autoimmune disease should remain on the clinician’s differential. In addition to routine laboratory tests, clinicians should consider the following immunological variables: ANA, IgM rheumatoid factor, antibodies to native DNA, SS-A, SS-B, Sm, RNP, complement C3 concentrations and serum immunoglobulin. In one study, 23.2% of patients admitted with acute pericarditis with pericardial effusion were found to have a systemic inflammatory disease (SID) such as Lupus or Sjogren’s syndrome. The immune-mediated pathogenesis of this syndrome is suggested by the presence of pro-inflammatory cytokines in pericardial fluid.

Conclusion

Accurately identifying the cause of recurrent pericarditis can increase treatment success rates. Corticosteroids are often considered second-line therapy unless there is a contraindication to NSAIDS or aspirin, or presumed autoimmune etiology. Though autoimmune disease is a less common cause of pericarditis, recognition of this syndrome and addressing the underlying disorder is necessary to prevent recurrence of pericarditis.
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New Evidence of Significant Incremental Benefit of Uninterrupted Direct Oral Anticoagulants vs. Uninterrupted Vitamin K Antagonists during Catheter Ablation of Atrial Fibrillation: A Systematic Review and Meta-Analysis of Randomized Controlled Trials

Background: To assess the incremental benefit of uninterrupted direct oral anticoagulants (DOACs) versus uninterrupted vitamin K antagonist (VKA) for catheter ablation (CA) of atrial fibrillation (AF) on 3 primary outcomes: major bleeding events (MBE), minor bleeding, and thromboembolic events (TE). The secondary outcome was post-procedural silent cerebral infarction (SCI) as detected by brain MRI.

Methods: A systematic review of Medline, Cochrane, and Embase was done to find all randomized controlled trials (RCTs) in which uninterrupted DOACs were compared against uninterrupted VKA for CA of NVAF. A fixed-effect model was used, except when I² ≥ 25, in which case a random effects model was used.

Results: The benefit of uninterrupted DOACs over VKA was analyzed from 5 RCTs that enrolled a total of 2133 patients (male: 71.9%) with AF. A significant decrease in MBE (Relative Risk [RR] 0.54, 95% Confidence Interval [CI] 0.30-0.96, p=0.04) with a Relative Risk Reduction of 46% and an Absolute Risk Reduction of 4.2%. No significant differences were found in minor bleeding events (RR 1.16, 95% CI 0.87-1.55, p=0.31), TE (RR 0.74, 95% CI 0.26-2.11, p=0.57), or post-procedural SCI (RR 1.11, 95% CI 0.79-1.56, p=0.53).

Conclusion: An uninterrupted DOACs strategy for CA of AF appears to be safer than uninterrupted VKA with a decreased rate of MBE. There are no significant differences among the other outcomes. DOACs should be offered as a first-line therapy to patients undergoing CA of AF, due to their lower risk of MBE, ease of use and fewer interactions.
Oral Anticoagulation after Catheter Ablation of Atrial Fibrillation and the Associated Risk of Thromboembolic Events and Intracranial Hemorrhage: A Systematic Review and Meta-Analysis

Background: We sought to examine whether remaining on oral anticoagulation (OAC) after catheter ablation (CA) of atrial fibrillation (AF) is associated with improved outcomes. OAC reduces morbidity and mortality in patients with AF. However, continuation of OAC following the blanking period of CA is controversial due to conflicting published data.

Methods: A systematic review of Medline, Cochrane, and Embase was performed for studies comparing patients with continued OAC (ON-OAC) vs. those in which OAC was discontinued (OFF-OAC). CHA2DS2VASC score had to be available for the classification of patients into high- or low-risk cohorts (CHA2DS2VASC ≥2 and ≤1, respectively). The primary efficacy outcome was thromboembolic events (TE). Intracranial hemorrhage (ICH) was the primary safety outcome.

Results: Five studies comprising 3956 patients were included (mean age 61.1 ± 2.9 years, 72.4% male, CHA2DS2VASC ≤1 50.1%, CHA2DS2VASC ≥2 49.9%). After a mean follow-up of 39.6 ± 11.7 months, OAC-continuation was associated with a significant decrease in risk of TE in the high-risk cohort (CHA2DS2VASC ≥2) (Risk Ratio [RR] 0.41, 95% Confidence Interval [CI] 0.21-0.82, p=0.01) with a relative risk reduction of 59%. ICH was significantly higher in the ON-OAC group (RR 5.78, 95% CI 1.33-25.08, p=0.02, 11/1569). No significant benefit was observed in the low-risk cohort ON-OAC after the blanking period.

Conclusion: Continuation of OAC after CA of AF with CHA2DS2VASC ≥2 is associated with a significantly decreased TE risk and a favorable net clinical benefit in spite of ICH being significantly increased in the ON-OAC group. Continued OAC offers no benefit with CHA2DS2VASC ≤1.
Chew May Cheah

Yelenis Fuertes Yanes, MD, Menatallah Algohary, MD, Sara Ishaq, MD, Natasha Mufuka, MD, Karen Beekman, MD and Anil Kapoor, MD

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Migraine presenting with Episodic Unilateral Mydriasis

Introduction:

Acute onset of anisocoria with severe headache may herald a neurological emergency and warrants extensive neurological workup. We present a rare benign cause of unilateral mydriasis in a patient without neurological pathology.

Case report

A 59-year-old woman with history of migraine headache was referred for anisocoria, headache and right facial numbness. She described the headache as unilateral, throbbing, constant; 9/10 on pain scale and only partially alleviated in a dark room. Her usual headache pain radiated to the right side of the neck and was associated with nausea, vomiting, photophobia and phonophobia. This episode of headache was more intense and new associated symptoms included visual blurring and right facial numbness.

Physical examination revealed asymmetric pupils, (right pupil measuring 7mm and the left pupil 2mm) and loss of consensual pupillary light reflex. The direct pupillary light reflex was diminished in the right eye. Visual acuity (near chart) without correction was 20/30 in both eyes. Extraocular motor function was intact. Peripheral vision was normal and no nystagmus, facial droop or ptosis was noted. There was decreased sensation to light touch on the right side of the face. The remainder of the neurological and physical examination were unremarkable. The complete blood count, comprehensive metabolic profile and thyroid function tests were normal. Cerebrospinal fluid analysis, magnetic resonance imaging of brain and angiography of head and neck were all normal. She was started on topiramate orally, dexamethasone intravenously, metoclopramide, acetaminophen/butalbital/caffeine for Status Migrainosus. The patient’s headache persisted, but with subjective improvement and diminished pain. After 5 days, patient was discharged home to continue topiramate. When seen in neurology clinic one month later, the pupils were equal and reactive to light and headache improved.

Discussion

Anisocoria often raises great concern because this condition may be linked with life-threatening neurologic disorders such as stroke or intracranial bleeding. It is usually seen with anticholinergic medications, third nerve palsy, trauma to the orbit, angle closure glaucoma, infection, aneurysm, inflammation, and certain plants (jimsonweed). Benign episodic unilateral mydriasis (BEUM) is an isolated cause of episodic pupil asymmetry. It occurs predominantly in females between the ages of 5 to 53 years. The underlying pathophysiology remains unclear. However, it is believed to be secondary to parasympathetic insufficiency of the iris sphincter, and sympathetic hyperactivity of the iris dilator. In our case, a 59-year-old female presented with anisocoria, associated with an episode of migraine. Pupil asymmetry resolved spontaneously after headache subsided. There are a few reported cases of benign episodic mydriasis presenting with migraine.

Conclusion

Anisocoria is an alarming sign. Immediate life-threatening events must always be ruled out in the presence of anisocoria. However, other pathologies should be considered including benign episodic unilateral mydriasis.
Dysphagia in immunocompetent patient: toxoplasmosis lymphadenitis

Introduction

Toxoplasmosis is a parasitic disease caused by Toxoplasma gondii. Infection occurs by (1) ingesting infectious oocysts from contaminated food or (2) ingesting tissue cysts in meat from an infected produce or (3) vertical transmission from infected mother to fetus. However, felines are the only animals in which T. gondii can complete its reproductive cycle. Non-feline warm-blooded mammals ingest T. gondii oocysts, allowing the organisms to invade intestinal epithelium, and disseminate throughout the body. Eventually the parasite lay dormant within the tissues for the life span of the host.

Case presentation

A 66-year-old female with history of diabetes, asthma, pulmonary Mycobacterium avium complex (MAC) came to clinic on 09/2018 because of a foreign body sensation when swallowing solid food for 1 month. She denied fever, weight loss, loss of appetite, and odynophagia. Patient was an ex-smoker who used 1 pack per day for 40 years and quit for 2 years. She denied any animal exposure and illicit drug use.

She had pulmonary MAC diagnosed in 05/2017, and the last sputum culture was negative in 09/2018. MAC treatment was continued.

Physical exam revealed a painless, firm, nonmobile lymph node at the left neck. Barium study was negative for structural abnormality and flexible fibrotic laryngoscopy showed normal appearance of larynx. CT neck showed a lymph node, 1.4cm in size, at the level 5 of left neck. After an additional 6 months of MAC treatment, her dysphagia did not improve. Repeat neck CT showed the same size of lymph node. Excision biopsy was performed and showed focal granulomatous inflammation and an immunohistochemistry stain consistent with T. gondii.

Serum toxoplasma IgG was positive while IgM was negative. Serum study was negative for HIV. We started her on treatment of trimethoprim-sulfamethoxazole (TMP-SMX) for 2 weeks. After treatment, her dysphagia improved and lymph node decrease in size on subsequent physical exams and imaging.

Discussion

Toxoplasmosis infection in immunocompetent patients are usually asymptomatic, self-limiting and do not require treatment. Clinical manifestations include fever, chills, and myalgia for several days. Some patients may develop generalized lymphadenopathy and the majority are bilateral, symmetrical, nontender cervical adenopathy. Treatment is initiated in patients who are pregnant, having prolonged symptoms, having ocular involvement, pneumonitis, myocarditis, or meningoencephalitis.

In our case, this patient developed new onset cervical lymphadenopathy during MAC treatment. Given patient was under treatment for MAC, may let physician lack of awareness of other differential diagnosis. Pathology of the lymph node showed numerous cysts with bradyzoites and positive T. gondii polyclonal antibody. We administered TMP-SMX (160 mg/800 mg) 3 tablets twice daily for 2 weeks and had significant improvement.
Resident/Fellow Research

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PYLLEPHLEBITIS - REAL PAIN IN THE GUT

Introduction

Hemorrhoidal banding is a commonly performed, effective and safe outpatient procedure for treatment of hemorrhoids. Common postprocedural complications include pain and bleeding. Rarely, it can lead to life threatening conditions, such as pylephlebitis. It is a condition that occurs due to infection of the thrombosed portal vein. Here we describe a case of pylephlebitis, in a 58 year old male following hemorrhoidal banding.

Case

We present a 58-year-old male who presented with a five day history of recurrent, intermittent, worsening upper abdominal pain followed by fever and chills. His surgical history was significant for hemorrhoidal banding, four months prior to hospitalization, with onset of the above symptoms 2 months following the procedure. Physical examination revealed significant epigastric tenderness without peritoneal signs. Initial laboratory work revealed leukocytosis of 11.5 x 10^9/L, total bilirubin of 3.8 mg/dL with direct bilirubin of 2.4 mg/dL, AST 49 U/L, ALT 69 U/L, and ALP 109 U/L. CT scan of the abdomen revealed diverticulitis of the sigmoid colon. Ultrasound of the abdomen was negative for choledocholithiasis or biliary dilation. He was initially started on Vancomycin and Piperacillin/Tazobactam which was then de-escalated to Ampicillin/sulbactam alone after 48 hours of negative blood cultures. His LFTs continued to worsen which prompted further evaluation with a MRCP that identified portal vein thrombosis. He was started on anticoagulation with Apixaban. On day two of hospitalization, blood cultures resulted positive for Streptococcus intermedius and with appropriate antibiotics patient improved for a few days. On day ten of hospitalization, he began developing abdominal pain and fevers. His LFTs at the time were a total bilirubin of 10.8 mg/dL, direct bilirubin of 8.2 mg/dL, AST 150 U/L, ALT 215 U/L, ALK of 328 U/L. A CT abdomen with contrast revealed extensive thrombosis of the superior mesenteric vein and portal vein along with hypoattenuation in multiple segments of the liver consistent with hepatic infarcts. There was high suspicion for pylephlebitis and his antibiotics were switched to ceftriaxone and flagyl. Repeat blood cultures were positive for Enterobacter aerogenes and ciprofloxacin was added to the regimen. Subsequently, patients clinical condition improved, his transaminitis and hyperbilirubinemia resolved and repeat blood cultures were negative. He was discharged on day 23 on apixaban with a plan to complete a 6-week course of ciprofloxacin for pylephlebitis and resultant gram negative bacteremia.

Discussion

Here we presented a case of uncomplicated hemorrhoidal banding resulting in pylephlebitis. Pylephlebitis is commonly associated with diverticulitis and rarely occurs from hemorrhoidal banding. A recent history of hemorrhoidal banding when associated with right upper quadrant abdominal pain, fever, or jaundice should prompt a high index of suspicion for pylephlebitis. Pylephlebitis although rare is fatal if not detected and treated early.
SEX RELATED DIFFERENCES IN IN-HOSPITAL OUTCOMES OF TRANSCATHETER MITRAL VALVE REPAIR: PERSPECTIVES FROM A NATIONAL DATABASE

Background: Little is known about the sex-related differences in the outcomes of transcatheter mitral valve repair (TMVR).

Methods: We queried the NIS database (2012-2016) to identify hospitalizations for TMVR. We conducted a propensity matching analysis to compare hospitalizations for TMVR in men versus women. Our main outcome was in-hospital mortality.

Results: Our analysis yielded 10,014 hospitalizations; 5,299 (52.9%) for men and 4,714 (47.1%) for women. After matching, there was no difference in in-hospital mortality between men and women (3.0% vs 2.4%, p=0.33). Also, there was no difference between men and women in cardiac arrest (2.1% vs 1.3%, p=0.17), cardiogenic shock (3.9% vs 3.5%, p=0.66), mechanical support devices (2.4% vs 2.9%, p=0.45), acute kidney injury (17.8% vs. 14.7%, p=0.08), hemodialysis (1.7% vs 1.6%, p=0.81), respiratory complications (1.7% vs 1.4%, p=0.65), acute stroke (1.4% vs 1.3%, p=0.82), discharges to nursing facilities (12.3% vs 15.2%, p=0.09), tamponade (0.5% vs 0.4%, p=0.69), acute myocardial infarction (2.1% vs 2.4%, p=0.71) and mean length of stay (6.03±8.153 vs 6.08± 8.858 days, p=0.82). TMVR in men was associated with higher incidence of ventricular arrhythmias (7.2% vs 4.1%, p=0.01) and lower incidence of pacemaker implantations (0.4% vs 1.7%, p=0.69).

Conclusion: Real world data showed no sex-related differences in in-hospital mortality for TMVR. Men experienced higher rates of ventricular arrhythmias, while women had higher rates of pacemaker implantations.
Resident/Fellow Clinical Vignette

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Life after tuberculosis: Where did my lung go?

Introduction
Tuberculosis remains a global public health concern. Bronchostenosis is a frequent complication which may not be preventable even with adequate microbiologic therapy. If left untreated, bronchostenosis has the potential to be a significant source of morbidity.

Case
A 24 year old female in the third trimester of an otherwise uneventful pregnancy was referred for shortness of breath and abnormal imaging. She reported completing treatment for pulmonary tuberculosis 2 years previously in India. Patient denied fatigue, cough, fever or chills. Physical examination revealed absent breath sounds in the left lung field. Chest radiography demonstrated left hemithorax opacification with ipsilateral mediastinal shift. Upon completion of tuberculosis treatment the patient was told the chest radiograph was normal. Three sputum smears were all negative for acid fast bacilli and nucleic acid amplification test did not detect M. tuberculosis. Further workup was postponed until the postpartum period. Postpartum pulmonary function tests (PFT) showed a restrictive pattern. Subsequent computerized tomography revealed complete atelectasis of the left lung with compensatory hyperinflation and apical scarring of the right lung. On bronchoscopic evaluation there was complete fibrotic occlusion of the left mainstem bronchus one centimeter below the carina. The patient was asymptomatic and no further intervention was initiated. The patient was counseled on healthy lifestyle and to avoid cigarette smoking.

Discussion
Endobronchial tuberculosis (EBTB) is a complication of pulmonary tuberculosis seen in up to 38.8% of cases. It is frequently undiagnosed due to non-specific clinical presentation. Bronchial stenosis results in up to 90% of EBTB cases. Adequate treatment with RIPE therapy may not prevent the development of bronchostenosis. Normal chest radiography or axial computerized tomography after treatment completion does not exclude the presence of bronchostenosis. High resolution chest computerized tomography (HRCT) provides the best radiologic assessment of the endobronchial tree. Endobronchial involvement was noted in 95-97% cases of pulmonary tuberculosis on HRCT. PFT most often reveal a restrictive pattern, however obstructive and mixed patterns can also be seen. Bronchoscopy is essential in diagnosis and management. If caseation is seen, RIPE therapy is indicated. Corticosteroids remain controversial in preventing stenosis as current literature report mixed results. Fibrotic changes are frequently encountered on bronchoscopy. The patient’s clinical status guides management. Patients with minimal or no symptoms, as our patient, do not require intervention. Symptomatic patients can be treated bronchoscopically with cryosurgery, balloon-dilation, laser therapy, controlled heat application, and stenting, or surgically. Bronchostenosis is a frequent complication of tuberculosis, which may not be preventable even with adequate microbiologic therapy. Some clinicians choose to extend RIPE therapy if EBTB is present. Management of fibrotic changes is guided by bronchoscopic observations and clinical status. Further study is needed to ascertain the benefit of corticosteroids and extended RIPE therapy.
A Case of Acute Tubular Necrosis in Post-Mechanical Thrombectomy

Acute tubular necrosis (ATN) occurs when there is death of epithelial cells in the renal tubules. Common causes of ATN include hypotension and nephrotoxins. One of the nephrotoxins is thought to be hemoglobin, which is a by-product breakdown of mechanical thrombectomy.

We will discuss a case of ATN induced by mechanical thrombectomy. A 44 year old female patient with no significant medical history presented with a large lower extremity deep vein thrombus, which was provoked following a 5th metatarsal fracture. The patient underwent mechanical thrombectomy and developed hematuria the following day. Creatinine level went up from 0.6 to 4, and BUN increased from 7 to 31. Urinalysis showed protein semiquant &gt; 300, small amount leukocyte esterase, nitrites, large amount of blood and bilirubin, and 0-3 RBCs. Nephrology was consulted and suspected hemoglobinuria associated acute tubular necrosis secondary to red blood cell fragmentation from mechanical thrombectomy. Patient did not require dialysis and creatinine level started to trend down after five days. Patient’s renal function recovered fully within 1 month.

Thrombectomy induced ATN cases are rare, and the cases reported have shown favorable prognosis. Mechanical thrombectomy is suggested to be beneficial in patients with large lower extremity deep vein thrombosis. It can reduce the risk of post-thrombotic syndrome, which is a costly problem with multiple burdensome effects on patients. Overall, mechanical thrombectomy procedures can be very beneficial, as long as we keep in mind the risk of ATN and closely monitor patient’s kidney function post-procedure.
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Bronchoscope in the Esophagus?! A Novel Technique for Esophageal Strictures Management

Introduction

Benign esophageal strictures can result from long-standing GERD, ablative or radiation therapy, corrosive substance ingestion etc. Management of strictures could be very challenging. We describe a case of esophageal strictures that was managed with an innovative approach.

Case Description/Methods

A 36-year-old Indian female with a history of esophageal strictures secondary to caustic ingestion 9 years ago presented with complaints of dysphagia. Patient underwent multiple EGD with dilatations in the early years of her stricture formation. Subsequently she was taught to perform self-dilations in India, every 2 weeks to relieve dysphagia. An esophagram revealed a narrow caliber mid and distal esophagus with 3 prominent stricture points. During endoscopy an 8.8mm gastroscope could not traverse the most proximal stricture hence a bronchoscope was used to evaluate the number and size of strictures for optimal use of pneumatic balloon catheter for dilation. The bronchoscope evaluation revealed 3 esophageal strictures at 25cm, 30cm and 35cm from incisors. Serial dilations were performed with a balloon dilator and triamcinolone was then injected at the stricture site post dilation. Following this intervention she was able to defer dilatation for 6 weeks. Using a Bronchoscope a second session of dilatation and triamcinolone injection was repeated 6 weeks later. Therapy was required for one of the three strictures with the others having healed well. Patient has not been performing self-dilations since her first EGD and continues to be asymptomatic.

Discussion

Caustic ingestion and its gastrointestinal sequelae such as stricture formation have been an important public health concern for decades. Damage and complications depend on several aspects: substance concentration and pH, length of time of tissue contact and amount ingested. The primary treatment of a caustic esophageal stricture is endoscopic dilatation, however sometimes management could be challenging. Using a bronchoscope (thin caliber scope), the high grade strictures in our patient were traversed and evaluated optimally prior to execution of the treatment modality. In our extensive literature review, a bronchoscope has never been used before for managing high-grade strictures, which makes our case unique. We believe that this new technique of utilizing a bronchoscope for evaluation of high-grade strictures can provide more targeted therapeutic approach and superior management to the patient.
Kartik Dhaduk

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Purtscher Retinopathy in the Setting of Acute Pancreatitis

Introduction: Purtscher retinopathy is a rare vasoocclusive retinopathy with estimated frequency of 0.24 per million cases in literature. It was originally believed to be associated with severe head trauma; however, it may also occur with other non-traumatic systemic diseases such as acute pancreatitis, fat embolism, SLE and connective tissue disorders. Here, we present a rare case of purtscher retinopathy in the setting of acute pancreatitis.

Case: A 25 year-old female with history of alcohol abuse and previous episodes of acute pancreatitis, presented to outside hospital with severe abdominal pain with vomiting. She was diagnosed with acute alcoholic pancreatitis with laboratory (amylase= 370, lipase= 1600) and radiological evidence (CT abdomen) and underwent appropriate medical management. On hospital day 2, the patient began to complain of dark spots in the visual field and blurry vision in her right eye. A CT head was grossly unremarkable and MRI brain showed no evidence of ophthalmic artery occlusion. Upon transfer to our hospital for higher ophthalmology care, the patient continued to have blurry vision. Ophthalmological exam of the right eye showed reduced visual acuity (vision 20/60) with blurred central vision and reduced color vision. Right retinal exam showed nasal disk elevation with parapapillary cotton wool spots and edema extending centrally to the nasal foveal region confined to zone A. The pattern was consistent with Purtscher-like retinopathy in the setting of acute pancreatitis. Short term steroid course was recommended but the patient left the hospital against medical advice and no follow-up records are available on clinical course.

Discussion: There is a large amount of variation surrounding the preceding illness in purtscher retinopathy. Fundoscopic examination is necessary to establish the diagnosis, it features cotton wool spots, hemorrhages, and retinal edema located predominantly around the optic disc. Fluorescein angiography may provide diagnostic evidence by showing leakage of dye from retinal arterioles, capillaries, and venules. The etiology is not well established but thought to be secondary to complement activation after the systemic injury, potentially causing coagulation and leuko-embolization of retinal small vessels. There are currently no established treatment guidelines in literature. Few case reports reportedly have documented improved outcomes with steroid use however there is controversy, whether or not the use of steroids has a significant impact on the natural course of the disease. The prognosis is variable depending on extent of retinal involvement and can range from loss of vision to delayed recovery by 2-to-4 snellen lines visual acuity.

Conclusion: Purtscher retinopathy is a rare cause of acute pancreatitis-related retinopathy as seen in this case. Physician awareness regarding this rare correlation may provide some benefit to patients by early referral to specialist, meticulous follow-ups and possible course of steroids. Patient should be educated on possible permanent ocular sequelae.
A rare presentation of cardiomyopathy in pregnancy

We report a 33-year-old woman with recurrent rhabdomyolysis in childhood, who presented in her third trimester of her first pregnancy with decompensated heart failure, initially referred to our advanced heart failure clinic with a working diagnosis of peripartum cardiomyopathy. She was later found to have mitochondrial trifunctional protein deficiency, an autosomal recessive fatty acid oxidation disorder caused by HADHA and HADHB gene mutations. This rare disease has a broad phenotypic presentation, however most cases of cardiomyopathy present in the neonatal period. Moreover, the neuropathic phenotype commonly presents by adolescence and has not been associated with cardiomyopathy in adulthood. Following a successful vaginal delivery, the patient underwent cardiac MRI showing diffuse hyperenhancement of all segments suggestive of infiltrative disease that prompted further genetic testing given her childhood history of recurrent rhabdomyolysis triggered by exercise or illness. Skin biopsy eight months postpartum was consistent with mitochondrial trifunctional protein deficiency and the patient was started on a medium light chain fatty acid diet and carnitine replacement therapy. In the first several months postpartum, she continued to have severely reduced cardiac function, however her functional impairment demonstrated by cardiopulmonary testing improved with optimal medical management, strict dietary compliance, and cardiac rehabilitation. One year postpartum she has fully regained cardiac function with normalized left ventricular systolic function. This is the first case report of an adult diagnosis of mitochondrial trifunctional protein deficiency with cardiac manifestations of disease.
Resident/Fellow Clinical Vignette

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EHLERS-DANLOS SYNDROME MAY BE ASSOCIATED WITH NODULAR GLOMERULOSCLEROSIS

Abstract:

Ehlers-Danlos Syndrome (EDS) describes a group of disorders that affect joints, bones, skin and vasculature. Some involve specific genetic components, but others involve many genes in various configurations. We describe a patient with EDS and presumed acute kidney injury, who had unexplained severe nodular glomerulosclerosis.

Case Presentation:

A 58 year old man with unknown medical history was found collapsed, and admitted for rhabdomyolysis with acute kidney injury, hyperkalemia, severe acidosis, hypothermia, and confusion. He had vision loss in his left eye, lobeless ears, a high palate, and bruising on his legs. Initial hemoglobin was 4.4 g/dl, BUN/creatinine were 139mg/dl and 7.5mg/dl, and CPK was 4600 U/L. The patient required emergency hemodialysis for severe metabolic acidosis and fluid overload.

Abdominal and pelvic CT showed splenic, celiac, superior mesenteric, renal and iliac artery aneurysms. Steroids were started for presumed vasculitis, possibly polyarteritis nodosa, and renal biopsy was obtained.

The biopsy showed advanced nodular glomerulosclerosis with global mesangial sclerosis, global glomerular basement membrane thickening, segmental inframembranous hyalinosis, and podocyte effacement and depletion.

When mentation improved, he provided a family history of aneurysmal rupture in his father and brother. Combined with facial features, acrogeria, and evidence of carotid aneurysms, vascular EDS was suspected.

Discussion:

This patient presented with rhabdomyolysis after immobility. Initial concern for his hemodynamic and acid-base status shifted to vasculitis when imaging revealed multiple visceral aneurysms. Only after combining the family history of aneurysms, the patient’s facial features, distribution of aneurysms, and acrogeria, was EDS considered.

Vascular EDS (vEDS) is rare and affects approximately 1:100,000 people (about 4% of EDS cases). This carries a risk of sudden blood vessel and internal organ rupture because of weakened type III procollagen needed for structural stability. This can lead to aneurysms of the iliac, splenic, renal or aortic arteries. There are reports in the literature of spontaneous dissection of the renal artery, renal infarction (mistaken for pyelonephritis) and renovascular hypertension. However, we did not find reports of nodular glomerulosclerosis.

The renal biopsy narrowed the differential considerably in this patient. The most common cause of advanced nodular glomerulosclerosis is diabetes mellitus, with smoking and chronic hypoxic states being less common. He had no history of diabetes; his glycosylated hemoglobin was within normal limits but uninterpretable because of severe anemia. He did not smoke, and there was no reason to assume chronic hypoxia.

It is possible that vEDS is related to the formation of nodular glomerulosclerosis. However, there seems to be no report of this connection in the literature. Further investigation should elucidate any causation of this type of advanced renal damage by vEDS.
DIFFERENT FACETS OF AUTOIMMUNITY: STIFF PERSON SYNDROME WITH GRAVES’ DISEASE

INTRODUCTION

Stiff Person Syndrome (SPS) is a rare neurological condition that is characterized by progressive muscle stiffness, rigidity, and spasm of axial muscles. It has been associated with other autoimmune diseases like Type 1 Diabetes Mellitus and Hashimoto’s thyroiditis but rarely with Graves’ disease. Here, we present a case to report an association between SPS and Graves’ disease and how plasmapheresis used to treat SPS helped in management of hyperthyroidism in the setting of thionamide induced leukopenia.

CASE REPORT

A 34-year-old African American female with a history of Graves’ disease was admitted to the ICU for thyroid storm and severe spasm and myoclonus of left leg. Laboratory evaluation showed thyrotropin (TSH) of < 0.002 mIU/L (reference range: 0.5 - 4.5), fT4 of >7.04 ng/dl (reference range: 0.8 - 1.8) and total T3 elevated to 278 ng/dl (reference range: 80 - 180). Thyroid-stimulating antibodies (TSI) were elevated to 3.25 IU/L (normal range: 0.00 - 0.55). She was started on methimazole (MMZ), hydrocortisone, metoprolol, and Lugol’s iodine. Total T3 normalized in 72 hours and fT4 down trended and normalized in 2 weeks. A comprehensive neurological workup with MRI brain and spine, EEG, lumbar puncture and rheumatological antibodies were negative. She was diagnosed with SPS with elevated GAD antibodies (> 25,000 IU/L). As she was planned for the second round of IV immunoglobulin (IVIg) therapy, repeat labs showed WBC of 1600 cells/mm3 with an absolute neutrophil count of 500 and TSH <0.02 mIU/L, fT4 1.28 ng/dl and total T3 of 186 ng/dl. Methimazole was stopped and alternative definitive therapy for Graves’ disease was needed to prevent another episode of thyroid storm. Due to nonresponse to IVIg, radioiodine ablation therapy was held till after she completed plasmapheresis for SPS. With plasmapheresis, fT4 and T3 levels dropped progressively from 1.28 ng/dl to 0.75 ng/dl and from 192 ng/dl to 107 ng/dl respectively but rebounded back up to 0.94 ng/dl and 159 ng/dl five days after the last session. TSI continued to remain elevated, dropping to 0.39 only after the last session but were elevated back up to 0.94 five days later.

The patient stayed clinically euthyroid. She eventually underwent radioiodine ablation of the thyroid gland.

DISCUSSION

Our case illustrates the unique association between Stiff Person Syndrome and Graves’ disease which has been reported to be rare in literature. Although thionamides are known to cause agranulocytosis, neutropenia has also been associated with IVIg in literature which could have been the case for our patient. Nonetheless, methimazole had to be stopped and another treatment option was necessary for treatment of Graves’. Plasmapheresis used for treatment of SPS helped in regulating thyroid hormone levels and preventing another episode of thyroid storm while she was awaiting radioiodine ablation.
Excellence in ACGME Core Competencies: Strengthening the Mentor-Mentee Relationship

Purpose:
The purpose of this study was to find the best practices, strategies and core priorities of experienced and accomplished physicians so as to provide a reference for residents of graduate medical programs.

Methods:
10 practicing oncology physicians each with at least 10 years of experience were interviewed with open-ended, narrative-based questions related to themes of paths to success, the proper role of a resident, lessons learned, helpful skills, and advice for a new resident/physician.

Results:
Surprisingly, the interviewees’ answers reflected the Accreditation Council for Graduate Medical Education (ACGME) Core Competencies of patient care, medical knowledge, practice-based learning and improvement, interpersonal and communication skills, professionalism, and systems-based practice. The importance of mentorships was also emphasized.

Conclusions:
The interviewees demonstrate an internalized wisdom of ACGME’s Core Competencies, and therefore stand as a valuable resource for residents. Given that mandated mentor programs in many graduate medical programs are ineffectively impersonal and mechanical based on signing off on forms and checklists, residents can follow the interviewees advice and proactively form mentor-mentee relationships with experienced physicians to learn the best paths of success.
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Spontaneous hemothorax secondary to bronchogenic carcinoma- A rare case

Spontaneous hemothorax is a clinical entity encompassing non traumatic accumulation of blood within the pleural space. Although hemothorax is often encountered in clinical practice, spontaneous hemothorax is an infrequent occurrence. Cases of spontaneous hemothorax have known to occur secondary to coagulopathy, malignancy, av malformations, pulmonary infarction and tuberculosis. The neoplasms known to be associated with hemothorax include schwannomas, Hepatocellular carcinoma and soft tissue tumors. lung carcinoma is a rare cause of hemothorax even in the setting of pleural extension. Here we highlight the importance of diagnosis spontaneous hemothorax in case of a primary lung malignancy.

A 75 year old woman with no signifcant past medical history, who presented to the ED with chest pain, was initially diagnosed with a pulmonary embolism and discharged on dabigatran. As a part of the work up for cause of pulmonary emboli in this otherwise healthy woman, a CT chest performed revealed an incidentally found right hilar lung mass and she was scheduled for an outpatient workup. A month later, she developed shortness of breath and presented to the ED before her scheduled appointment. A large pleural effusion was seen on the repeat CT chest. She underwent thoracocentesis, with pleural studies consistent with hemothorax. Eventually, she underwent endobronchial ultrasound with biopsy of lung mass. Based on the histopathology findings, she was diagnosed with adenocarcinoma. We concluded that she likely experienced spontaneous hemothorax secondary to lung adenocarcinoma.

To our knowledge there have been three other case reports of spontaneous hemothorax secondary to bronchogenic carcinoma. Cases of hepatocellular carcinoma with hemothorax have been known to have poor prognosis. Being uncommon, the prognostic implication of spontaneous hemothorax in primary lung malignancies has not been studied. Recognition of hemothorax in these patients can alter the course of management.
Primary hepatic angiosarcoma is a very rare malignancy. Polyvinyl chloride and arsenic have been identified as the causative agent for hepatic angiosarcoma. Here we present a rare case of hepatic angiosarcoma possibly due to prolonged exposure to polyvinyl chloride.

A 75-year-old man with past medical history significant for hypertension, hyperlipidemia and coronary artery disease and occupational history significant for automobile mechanic for five decades presented to his doctor for increasing abdominal distension and shortness of breath for four weeks. He was evaluated for recurrent hemorrhagic ascites of uncertain etiology. He was admitted to the hospital for further evaluation. His physical examination was remarkable for non-tender abdomen with distension and positive fluid thrill. History was negative for alcohol and tobacco smoking. Diagnostic paracentesis was suggestive of non-portal hypertensive etiology for ascites. Ascitic fluid cytology did not reveal any malignant cells. Work up for cirrhosis including the infectious panel, serum ceruloplasmin, and serum ferritin were negative. Markers for malignancy including alpha-fetoprotein and carcinoembryonic antigen were also negative. Doppler ultrasound excluded portal vein thrombosis. Owing to his occupational history of prolonged exposure to polyvinyl chloride, angiosarcoma of liver was considered as probable diagnosis. Computed tomography angiography of abdomen and pelvis showed nodular contour of liver with multiple lesions suspicious for malignancy. Magnetic resonance imaging with liver mass protocol showed exophytic growth along the posterior right liver lobe. Interventional radiology guided biopsy of the omentum was done which on histopathology showed high grade angiosarcoma. The primary was presumed to be angiosarcoma of liver with metastasis to the omentum and thus was staged to have advanced stage angiosarcoma. He was started on taxel-based chemotherapy for palliative intent. He initially improved with the treatment with decreasing abdominal distension and requiring less frequent therapeutic paracentesis. However, he died within 3 months of the initial diagnosis due to multiple organ involvement and infectious complications.

This case illustrates the diagnostic challenge hepatic angiosarcoma poses as it does not have any characteristic tumor markers and presents with non-specific symptoms. Morphological appearances of primary hepatic angiosarcoma varies. It may appear as multiple nodules or dominant mass of diffuse infiltrating lesions. Definitive diagnosis requires high index of suspicion, careful history of exposure to carcinogens and histological examination of the tissue. It has poor long-term survival due to rapid progression, high recurrence and poor response to chemotherapies.
Resident/Fellow Clinical Vignette

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Triple vascular disease: There is no such thing as a “routine catheterization”

Retroperitoneal bleeding and the formation of hematoma after cardiac catheterization is a life-threatening complication. The incidence is 0.15-6% and the mortality rate is 4-12%. The risk factors are a sheath size greater than 7 millimeter, female gender and postmenopausal state, small body surface area, high above inguinal ligament arterial puncture, chronic renal insufficiency, antiplatelet use, heparin use and emergent procedures.

We present the case of a 54-year-old female with a history of asthma and hyperlipidemia who was admitted for Non ST Segment Elevation Myocardial Infarction (NSTEMI). Aspirin, clopidogrel and heparin drip were initiated. She underwent cardiac catheterization which revealed normal coronary arteries. Access was obtained using a 6 French Sheath at the right common femoral artery and closed with a MYNX vascular closure device. Post procedure she had moderate to severe pain at the access site for which she received fentanyl with partial relief of pain. A small hematoma was palpated and pressure applied for 15 minutes and observation for 45 minutes. She was discharged 6 hours after the procedure.

An hour after discharge the patient developed severe right groin and right lower quadrant abdominal pain. At this point she presented to the emergency department at our community hospital. Ultrasonography revealed a deep vein thrombus (DVT) in the right common femoral, superficial and popliteal arteries. Heparin drip was started. Computed tomography (CT) scan showed a large retroperitoneal hematoma. Heparin was discontinued and vascular surgery consulted. Urgent surgery revealed laceration of the posterior wall of the external iliac artery 15 to 20 centimeters from the puncture site which was repaired. She required multiple transfusions including 7 units of packed red cells and 2 units of fresh frozen plasma. An IVC filter was placed for DVT. On day 7 after initial exposure to heparin her platelets started to drop from 237 to 45 x 10^3 uL. The calculated 4 T score was 5 suggestive of intermediate probability of heparin-induced thrombocytopenia (HIT). HIT antibodies were sent which were positive. At this point she developed a low grade fever. On day 12 she started to complain of bilateral calf fullness. Ultrasonography confirmed new lower extremity DVT, Eliquis was started. The length of stay was 20 days.

While cardiac catheterization has almost become a “routine procedure in the United States, with over 1 million performed each year, we should still be mindful of the potential serious complications that can occur as demonstrated in this case. Early recognition of patient complaints may allow prompt treatment and prevent downstream complications, lower mortality and healthcare costs.
Sertraline Induced Myopathy in An Elderly Patient

Introduction:
About 7% adults reported at least one episode of Major Depressive Disorder (MDD) annually. SSRI’s are the 1st line treatment for MDD. Although they are much safer than TCA’s and MAOI’s, 55% of patients started on SSRI’s reported side effect, most commonly insomnia or sexual dysfunction. Rhabdomyolysis is a rare adverse effect of SSRI’s, and this paper presents this case.

Case Presentation:
A 74-year-old female with depression presented with limited mobility. Four months ago, she started having frequent falls along with fatigue. Two weeks prior to presentation, she had difficulty standing from a seated position and walking up stairs. On the day of presentation, weakness progressed, and she was unable to stand up from sitting position. History was negative for change in weight or appetite, joint or muscle pain, rash, sicca or Raynaud’s phenomenon. No personal or family history of rheumatologic, neuromuscular or thyroid disease. On further review, she was started on Sertraline 6 months ago for depression. Examination was significant for absent ankle reflexes and 3/5 strength in bilateral shoulder abduction and hip-flexion. Labs (CK) 12,160 IU/L, AST of 326, ALT 51, BUN/Creatinine 29/1.28, ESR 38, Aldolase 95.6, TSH 2.6. ANA titer was 1:160. Rest of rheumatological tests were normal. EMG showed an inflammatory myopathy, a muscle biopsy which showed evidence of a necrotizing myopathic process without significant lymphocytic infiltrates. Possible etiologies including drug/toxin related, autoimmune, or paraneoplastic myopathies. Imaging ruled out occult malignancy. Patient was started on intravenous fluids. On day1, CK trended up to 12,931, at this point Sertraline was stopped. On day2 CK decreased to 9,345 which normalized to 98 on day8. Motor strength improved over the course of hospitalization. She was discharged on day 9 with recommendations not to restart Sertraline. After a week, she was able to walk on her own and CK was down to 30.

Discussion:
It is hypothesized that serotonin is involved in skeletal muscle contraction and relaxation. SSRIs may lead to faster initiation of contraction and longer length of contractions which may results in muscle damage. Although rare, Sertraline can be a cause for rhabdomyolysis and may be suspected in patients where sertraline was recently started, was prescribed in high doses, and/or when no other cause is identified. Sertraline induced rhabdomyolysis has a bimodal distribution. This case may caution the use of SSRIs in elderly patients and highly athletic patients where SSRIs may exacerbate or cause rhabdomyolysis. It is important to consider Sertraline as a possible source of muscle injury with notable consequences such as acute kidney or liver injury.
MYCOBACTERIUM AVIUM COMPLEX INFECTION UNMASKED BY IMMUNE RECONSTITUTION SYNDROME IN AN AIDS PATIENT MIMICKING A LYMPHOMA

Introduction:

Immune Reconstitution Inflammatory Syndrome (IRIS) describes the inflammatory reaction that leads to a paradoxical worsening of pre-existing infectious processes after initiation of HAART (highly active antiretroviral therapy) in HIV patients. The infections may have been previously diagnosed and treated, or they may be subclinical and unmasked by the host’s exaggerated inflammatory response. Effective treatment of the infection is essential in all cases, and deferring antiretroviral therapy may be warranted in very severe cases.

Case:

A 53 years old homeless male with history of polysubstance abuse and longstanding AIDS with non-compliance to medication, presented with one month history of moderate to severe diffuse abdominal pain and low-grade fever with chills and rigors. Physical exam revealed soft, tender abdomen with an ill-defined periumbilical swelling. Patient was started on HAART 12 weeks ago after a long hiatus in treatment. HIV viral load was 1.16 million copies/mL and CD4+ count of 34 cells/cmm at that time. On the current presentation, viral load and CD4+ count was 259 copies/mL and 231 cells/cmm respectively. CT chest showed mediastinal lymphadenopathy and multiple pulmonary nodules. CT abdomen showed conglomerate mesenteric lymphadenopathy, 8 x 5 x 9 cm in size, and enlarged para-aortic lymph nodes. EBV markers revealed EBNA IgG +, EB-Early Antigen IgG +, EBV IgG + and EBV IgM - consistent with a prior EBV infection. These findings were highly suggestive of lymphoma. However, biopsy of mesenteric lymph nodes showed necrotizing granulomatous tissue with acid fast bacillus. Bone marrow biopsy showed no evidence of lymphoma or leukemia. Flow cytometry ruled out any monoclonality in B-cells, T-cells or increased proportion of blasts. Hematopathology was negative for any concerning genetic mutations. Therefore, a diagnosis of Mycobacterium Avium Complex infection was made. Patient was started on Azithromycin 500mg daily with Ethambutol 1200 mg daily while continuing his HAART regimen. Since patient was on Bictegravir-Emtricitabine-Tenofovir (Biktarvy), Rifampin was withheld given the drug interactions. On follow-up visit 2 weeks later, patient was afebrile with considerable improvement in symptoms. He will be treated for a minimum of 12 months.

Discussion:

IRIS risk is associated with a rapid increase in CD4 counts and rapid decrease in viral load after initiation of HAART. Our patient had 7-fold increase in CD4 count and a sharp decline in viral load. There might have been a subclinical MAC infection that manifested after institution of HAART. It is to be noted that such a presentation in HIV can resemble a lymphoma, especially with a history of low CD4+ counts and prior EBV infection. Clinicians should be aware of this mode of presentation of a MAC infection to ensure prompt diagnosis and treatment.
Byler Syndrome or progressive familial intrahepatic cholestasis type 2 (PFIC 2) is a rare disorder (incidence of 1/50,000 to 1/100,000 births) which presents in infancy with intrahepatic cholestasis. It results from mutation in ABCB11 gene located on 2q24 which codes for ATP-dependent BSEP protein expressed in hepatocyte canalicular membrane which exports bile acids. It is referred Byler Disease [PFIC 1] when present in members of the Amish kindred in which it was originally described. There are three subtypes of PFIC; PFIC1 and 2 have few clinical differences despite having genetic distinctiveness. They are caused by absence of gene product required for canalicular export and bile formation and have low GGT levels. PFIC 3 has similar presentation but with high GGT and deficient hepatocellular phospholipid export. Fewer than 200 patients with PFIC1 or 2 are reported in literature.

Clinical features include severe pruritus, jaundice and liver failure within first few years which may be complicated by hepatocellular carcinoma and cholangiocarcinoma (higher incidence with PFIC2). Few patients survive into third decade without treatment. Lab findings include raised transaminases, normal GGT and very high serum bile acid. Histopathology shows canalicular cholestasis with lobular and portal fibrosis and inflammation. Ursodeoxycholic acid and surgical biliary diversion are initial therapeutic modalities and liver transplantation if they fail.

Here we present a 27 y.o. male with Byler syndrome who underwent biliary diversion ostomy at 8 months of age who presented with diffuse abdominal pain and jaundice for 5 days. Pain started around stoma site and progressed to be generalized. He reported poor output from stoma for a week. On arrival, vitals signs were stable. Labs were notable for transaminitis (AST 96, ALT 87) with cholestatic pattern (ALP 344, Tbili 6.4, direct bilirubin 4.4) and no leukocytosis. CT abdomen/pelvis showed constipation with colonic dilation compressing stoma output. US was notable for under-distended gallbladder; CBD was not visualized. Baseline bilirubin from 6 months prior was normal with Tbili 0.6, direct component 0.2. Given the stool burden and reduced output through stoma, patient was started on an aggressive bowel regimen. He had good bowel movements with resolution of abdominal pain. MRCP showed Biliary diversion from the gallbladder fossa to anterior abdominal wall of the right mid abdomen without biliary obstruction. HIDA scan also showed no evidence of biliary obstruction. Vitamin A, D, E were normal however Vitamin K level was less than 0.13. Patient was discharged on a bowel regimen and his LFTs were repeated 2 weeks later which trended down to reference range.

This rare syndrome which typically presents with diarrhea due to malabsorption of fat-soluble vitamins, in our case, presented with constipation leading to colonic dilation which obstructed the canalicular flow, worsening the symptoms of Byler disease.
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**VZV ENCEPHALOMENINGITIS: A PECULIAR CAUSE OF SYNCOPE AND ALTERED MENTAL STATUS**

“Well Doctor, my dad was not like this at home,” is a commonly heard statement among physicians interacting with family members of confused elderly patients. Most of the time, physicians may brush this off as hospital acquired delirium; however, without thorough investigation, physicians may miss an important diagnosis.

Presented is a case of delayed diagnosis of aseptic encephalomeningitis in an elderly male who presented with altered mental status; with which treatment resulted in rapid improvement of symptoms.

**CASE DESCRIPTION:**

An 87 year old male with PMH of BPH, MI, DVT, and history of “skipped beats” presented to the ED for syncope. Patient got up to use the bathroom and subsequently lost consciousness. EKG showed 2nd degree type 1 AV Block, sinus pause, and ventricular escape beat with LAFB. Physical examination was positive for altered mental status. Initial workup was negative - including cardiac markers, CT and MRI brain, Carotid doppler, dementia workup, urinalysis, and chest xray. Patient’s mental status fluctuated between AAO x 0– AAOx 1 - 2. MMSE score was 15, indicating severe cognitive impairment. He continued to have significant multilevel conduction block, requiring pacemaker placement. Echocardiogram and nuclear stress test showed changes from previous MI.

Patient’s neurologic status did not improve and family was insistent that this was not his baseline; patient was AAOx 3 at home. CSF studies from lumbar puncture showed lymphocytic predominance and protein, consistent with aseptic meningoencephalitis. Empiric acyclovir was started. CSF VZV PCR came back positive. Patient continued acyclovir with rapid improvement in mental status. Conduction block improved and pacemaker was not required at time of discharge.

**DISCUSSION:**

VZV is an uncommon cause of CNS infection. Aseptic meningitis is usually due to enteroviridae, while viral encephalitis is commonly caused by HSV. VZV encephalitis was primarily reported in patients with impaired cell mediated immunity or disseminated herpes zoster rash. 35% of cases of VZV reactivation occur in immunocompromised patients; whereas only 0.1-0.2% of cases occur in immunocompetent hosts, as in our patient. Furthermore, VZV reactivation usually presents with a vesicular rash; meningoencephalitis is a rare complication; occurring in 1-3/10,000 cases. Our case describes one of few cases of VZV encephalomyelitis occurring in an immunocompetent host, with an atypical presentation due to lack of rash. This patient demonstrates a common hospital admission for syncope. The most common causes of syncope were ruled out; but the patient persisted to have altered mental status. With repeated insistence by family members, a lumbar puncture was performed. Empiric acyclovir was started with improvement of patient’s mental status in 24 hours. It is our hope that this case will increase awareness of rare presentations of an uncommon disease, enabling clinicians to diagnose and treat a condition to significantly improve someone’s quality of life.
TRAPPED IN THE WRONG BODY: A CASE OF WERNER SYNDROME

Introduction:
Werner Syndrome is a disorder involving pre-mature aging in males and females. First described in 1904 by Otto Werner, Werner syndrome, also known as adult progeria, is a hereditary disease of premature aging apparent around early adulthood. It continues to be a rare disease, affecting 1-20 per million population in the US. Presentation varies among individuals; however, certain clinical characteristics point toward its diagnosis. Presented is a case of a 39 year old female who came to the hospital with a straightforward complaint, but was found to have multiple problems that formulated toward a diagnosis of progeria.

Case Presentation:
Patient is a 39 year old, Caucasian female, who presented to the hospital due to altered mental status secondary to hypoglycemia. Upon further questioning, it was found that the patient was adopted, and biological mother had likely died from cancer at a young age. On physical examination, there was a stark difference in patient’s appearance and age: patient was a small, malnourished female, who looked closer to age 60 than 39. She had prominent wrinkles and bruises on her face and extremities. Her face was small, with a prominent forehead and small jaw, and a pointed, pinched nose with sunken cheeks. She had a high-pitched, raspy voice. Imaging showed carotid atherosclerotic disease, and brain parenchymal involutional and ischemic changes disproportionate for patient age. Hospital course was complicated by sepsis, ESRD, CHF, severe hypothyroidism, anemia, and frequent change in mental status. Patient suffered cardiac arrest, and was eventually made comfort measures by family; patient passed shortly after.

Discussion:
Werner Syndrome is an autosomal recessive disorder diagnosed by clinical criteria, and may be confirmed with genetic testing for the WRN gene, which encodes the WRN protein involved in DNA metabolism. As our patient was adopted, it was not known if her parents had Werner syndrome; however, her mother passing from cancer at a young age may hint toward the possibility of a genetic disorder. Werner Syndrome is characterized by pre-mature aging seen around age 20-30; specifically short stature, skin changes, cataracts, loss of hair, high pitched voice, and bird-like facial features. Cause of death is most commonly due to myocardial infarction or malignancy; average life expectancy is 46 years. We share our case as it illustrates an interesting and unique case in medical literature. It is our hope that by sharing the story of our patient, patients and families in similar situations are counselled on presentation, complications, and prognosis of genetic conditions to improve quality of life of such patients and ease burden of family members.
Resident/Fellow Clinical Vignette

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Daptomycin Related Acute Eosinophilic Pneumonia

Background: Acute eosinophilic pneumonia (AEP) is a fairly uncommon condition, which has been associated with exposure to commonly used medications. We describe herein a patient who developed acute eosinophilic pneumonia after treatment with the lipopeptide antibiotic daptomycin.

Case: A 67-year-old male with a recent diagnosis of methicillin-resistant staphylococcus aureus (MRSA) spinal osteomyelitis and infective endocarditis was treated with daptomycin. Indication for daptomycin use included an allergic reaction with the use of ampicillin and vancomycin. After taking daptomycin for 4 weeks, the patient presented to the emergency department with a dry cough and shortness of breath. Laboratory testing revealed leukocytosis, with eosinophil count of 9%. He was afebrile, with oxygen saturation of 98% on room air. Chest X-ray showed bilateral infiltrates, and CT scan of the chest demonstrated parenchymal consolidation and ground-glass opacities bilaterally. Daptomycin was discontinued, and alternate empiric antibiotic therapy was initiated to cover both the known staph infection and the new bilateral pulmonary infiltrates. Pulmonary consultants added steroids to the treatment regimen. Bronchoscopy was considered, but within 24 hours the clinical condition drastically improved with complete cessation of the cough, and normalization of the peripheral eosinophil count. The patient remained on steroids for a total of 5 days, and follow up chest radiographs documented dramatic progressive improvement of the bilateral lung findings.

Discussion: Acute eosinophilic pneumonia (AEP) is a rare but an important side effect of multiple drugs. AEP is usually diagnosed by clinical findings, laboratory results, and radiologic findings. Most patients present with a cough with or without fever, dyspnea, peripheral eosinophilia, and bilateral lung infiltrates on imaging. Our patient presented with cough, dyspnea and had peripheral eosinophilia with bilateral pulmonary infiltrates on chest radiography. Review of published cases indicates that symptoms of AEP generally appear after 2-3 weeks of starting daptomycin. Bronchoscopy with BAL showing greater than 25% eosinophils helps to confirm the diagnosis. Treatment involves stopping the offending medication and initiating systemic steroid treatment. The condition generally responds dramatically to this approach. The dose and duration of steroids for the treatment of AEP is not clearly understood, although many cases report improvement with a short duration of steroids.
Inferior Vena Cava Atresia; A Rare Cause of Recurrent Deep Vein Thrombosis

Abstract:
Risk factors for venous thromboembolism may be acquired or hereditary. Agenesis or atresia of the inferior vena cava (IVC) is a rare congenital anomaly with an estimated prevalence of 0.0005-1% and classically presents as proximal recurrent deep venous thrombosis (DVT) in a young male. IVC atresia can be implicated in up to 5% of cases of unprovoked venous thromboembolism. We present the case of an obese male who presented with his 2nd deep venous thrombosis and was incidentally diagnosed with Inferior Vena Cava atresia on abdominal imaging.

Case:
37 year old morbidly obese male with history of unprovoked right lower extremity deep vein thrombosis at age 17 on chronic anticoagulation presented with complaints of right lower extremity pain and swelling for one week along with swelling of scrotum and penis with associated difficulty with micturition. Physical examination was remarkable for marked edema of RLE extending proximally to involve scrotum, penis and lower abdomen. RLE was also erythematous in appearance. Doppler Ultrasound revealed extensive DVT with a non-compressible occlusive thrombus in the right common femoral, popliteal, peroneal and posterior tibial veins. CT scan of the abdomen and pelvis confirmed extensive thrombosis involving the right iliofemoral system with surrounding fat stranding. Interestingly, sudden narrowing of inferior vena cava was appreciated beyond the common iliac venous confluence with no definable infrahepatic or intrahepatic IVC which was replaced with a fibrous strand instead. However, suprahepatic IVC was patent and numerous collateral vessels were noted in the abdomen and retroperitoneum.

Patient underwent intraabdominal and bilateral lower extremity venography with pharmacomechanical thrombolysis of the right iliofemoral thrombus. IVC could not be tracked beyond the confluence of common iliac veins- a finding consistent with an atretic IVC. Significant symptom improvement was reported post-thrombolysis. Subsequently, an attempt was made at IVC reconstruction with stenting to allow for long-term venous drainage which was unsuccessful and patient was discharged with regular follow up with radiology.

Discussion:
Atresia of the IVC usually presents as recurrent DVTs with approximately 1/3rd of the patients having an underlying inherited thrombophilia. Our patient did not have any other risk factor except obesity. Although DVT is quite extensive, pulmonary embolism is less common as the dislodged thrombi get entrapped in the collateral system following a more convoluted route to the pulmonary circulation through the azygous and hemi-azygous venous system. Consideration of this diagnosis in the right clinical setting is vital as the recurrence rates of DVT are high. Patients should be advised to wear elastic stockings and avoid smoking, physical exertion or contraceptive use. Our experience clarifies that anatomical abnormalities should be considered in patients who have unexplained, extensive or bilateral DVT despite anticoagulation and represents an under-recognized risk factor for venous thromboembolism.
A CASE OF 61 YO FEMALE WITH AN UNUSUAL MASS IN THE LEFT ATRIUM

Introduction

A mass in the left atrium can be challenging to diagnose. The leading differentials are thrombi, vegetations and tumors which may be primary (benign or malignant) or metastatic (1). We present a case of mass in the left atrium found in a 61-year-old woman with history of bronchial-associated lymphoid tissue (BALT) lymphoma.

Case Description

A 61-year-old female normotensive, nondiabetic, former smoker presented with increasing dyspnea with productive cough and white expectoration for a few days. There was no history of fever, chest pain, palpitations, orthopnea, edema or loss of consciousness. History of significant weight loss and anorexia were present. The patient’s background was that she was initially diagnosed with pulmonary BALT lymphoma for which she was treated with Rituximab. PET scan 2 months prior to the admission showed left lower lobe mass with obstruction of the left mainstem bronchus. Subsequently, she received Obinutuzumab and Bendamustine. On admission, she was afebrile with mild tachycardia, blood pressure 110/60, respiratory rate 25 breaths/minute and SpO2 96% on 2 L nasal cannula. Breath sounds were decreased over the left hemithorax. EKG showed sinus tachycardia. Hemoglobin was 9 g/dl and blood chemistry was normal. Chest X-ray showed opacification of the left hemithorax. CT pulmonary angiogram ruled out pulmonary embolism but revealed thrombi within the left atrium and left inferior and superior pulmonary veins. These were not reported in the earlier PET scan. Apixaban was initiated given the concern for systemic thromboembolism. Transthoracic echocardiogram showed a mass in the left atrium. There was no consent for transesophageal echocardiogram. A cardiac MRI confirmed a bulky left lung mass suggestive of carcinoma invading the left atrium via the left pulmonary veins. There was bulky intraluminal tumor thrombus and extensive mediastinal invasion by tumor. CT-guided biopsy of left lung mass was suggestive of non-small cell lung carcinoma (NSCLC, adenocarcinoma). A diagnosis of dual synchronous cancers, a) BALT lymphoma and b) NSCLC adenocarcinoma with cardiac invasion was made. It was not clear which of the two tumors or if possibly even both, had invaded the left atrium. She continued to receive palliative chemotherapy.

Conclusion

The clinical manifestations of cardiac involvement in lung cancer is relatively uncommon although cardiac metastases have been reported in up to 25% of patients in autopsy studies (2). Hence, clinicians should always be aware of the possibility of cardiac metastasis in lung cancer. Our patient had the unusual presentation of cardiac involvement where the tumor invaded the heart through two of the pulmonary veins. Other routes of spread of lung cancers into the heart include retrograde lymphatic spread, hematogenous dissemination and direct invasion. The tumor invasion to the left atrium through the pulmonary veins is an extremely rare event (3).
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A case of lumbosacral plexopathy secondary to spontaneous hemorrhage from apixaban use

INTRODUCTION

Anticoagulation is used for the prevention of stroke in patients with non-valvular atrial fibrillation. The use of direct-acting oral anticoagulants (DOACs) has made this easier without the need for INR monitoring and fewer side effects. Neurological complications stemming from hemorrhage outside the central nervous system is rare with the use of DOACs. We report a case of lumbosacral plexopathy as a result of acute iliopsoas hemorrhage secondary to anticoagulation use.

CASE REPORT

A 74-year-old female presented with bilateral leg swelling and numbness of the right lower extremity of one-month duration. She had a history of COPD, hypertension, obesity, depression, atrial fibrillation on apixaban. On examination, she had non-pitting bilateral lower extremity swelling greater on the right with erythema of the right thigh. There was reduced sensation of the entire right lower extremity to light touch and pain. Power was 3/5 in the right lower extremity. The left lower extremity was normal on neurological examination. A computed tomographic scan of the lower extremities revealed an acute intramuscular hemorrhage in the right iliopsoas muscle complex and medial musculature of the proximal right thigh with abnormal stranding of the subcutaneous fat of the proximal thigh and right hemipelvis. Anticoagulation was discontinued, and patient began physical therapy. There was moderate improvement in motor function and sensation of the right lower extremity. Hemoglobin was stable after discontinuation of Apixaban. The patient was discharged to a subacute rehabilitation center.

DISCUSSION

Apixaban can be used in the management of venous thromboembolism and prevention of stroke in patients with non-valvular atrial fibrillation. Risk factors for bleeding in patients taking anticoagulation include advanced age, hypertension, duration of use and comorbid conditions including congestive heart failure, diabetes mellitus, renal and hepatic disease. Bleeding may be severe or life-threatening depending on the site and duration of bleeding. Our patient was elderly and hypertensive and had taken apixaban for approximately one year. Lowest hemoglobin during admission was 7.7g/dl and hemoglobin one year before admission was 12.1g/dl. Hemorrhage from apixaban use causing plexopathy has not previously been reported to our knowledge. However, extremity swelling on a patient receiving anticoagulation should raise concern for possible hemorrhage.
Introduction:
Lyme disease (LD) is a zoonosis, transmitted through the tick bite carrying the spirochete Borrelia Burgdorferi. LD is a clinical diagnosis based on physical examination findings of erythema migrans rash in a patient from the endemic area. We present a case of LD, diagnosed through direct visualization of spirochetes on a Wright Giemsa peripheral blood smear with a nonspecific physical examination. There is scarcity in the literature on diagnosing LD by an inexpensive laboratory technique.

Case Presentation:
A 36-year-old male with no past medical history presented with constant fever, and arthralgias for three days. His physical examination was insignificant and did not reveal any skin rash or joint swelling. Laboratory studies including complete blood count, complete metabolic panel, blood cultures, human immunodeficiency virus antibody, Epstein Bar virus antibody were unremarkable. A peripheral blood smear displayed scant visible helical shaped spirochetes on microscopy. The patient then revealed that he visited an area of Uzbekistan where ticks are common but did not recall a tick bite or rash on his body. Intravenous ceftriaxone was started due to the findings on peripheral smear. LD was confirmed after the results of positive enzyme-link immunosorbent assay of 2.74 index with positive Lyme Antibody Western Blot IgM Bands. Polymerase Chain Reaction for Tick-Borne Relapsing Fever was negative. The patient improved and was discharged on cefuroxime.

Discussion:
Spirochetemia is one of the characteristics of Lyme Borreliosis and Tick-Borne Relapsing Fever Borreliosis (RF Borreliosis). RF borreliosis is characterized by an abundance of spiral-shaped spirochetes in the blood with recurring fever whereas LD is characterized by a scant number of helically shaped spirochetes in the blood with constant fever and various tissue involvement, including skin, joints, and the nervous system. Erythema migrans is present in 70% of infected individuals. The diagnosis of LD is made by antibody testing including enzyme immunoassay or Indirect immunofluorescence assay with or without the western blot. However, antibodies are not detected at early stages or may be falsely positive due to the presence of other conditions like Tick-borne Relapsing Fever. The definitive diagnosis is made by the culture of Borrelia from specimens in Barbourâ€“Stoennerâ€“Kelly medium. However, such techniques requires several weeks for positive spirochete growth in a medium. That is where the importance of Peripheral smear can be debated in the diagnosis of LD. Our case illustrated detection of early LD by observation of spirochetes on the peripheral smear in absence of erythema migrans. Accurate diagnosis of LD plays a crucial role in treating the infection and preventing serious complications.
A CASE OF CD-20 NEGATIVE PLASMABLASTIC LYMPHOMA LURKING IN THE SHADOW OF A LEIOMYOMA

CD20-negative diffuse large B-cell lymphomas (DLBCLs) are a rare entity and constitute 1-2% of all DLBCLs. The disease course is characterized by an aggressive extranodal spread with high resistance to routine chemotherapeutic regimens. Plasmablastic lymphoma (PBL) is the most common variant (75% of total cases). We report a unique case of PBL with pelvic organ involvement successfully treated with EPOCH (Etoposide, Prednisolone, Oncovin/Vincristine, Cyclophosphamide, and Hydroxydaunorubicin) regimen.

A 57-year-old African American female with a past medical history of uterine fibroids was transferred to the emergency department with complaints of lower abdominal/pelvic pain and abnormal uterine bleeding. At presentation, she was hemodynamically stable. The abdomen was distended and diffusely tender, without any peritoneal signs. Pelvic examination revealed multiple palpable masses in the uterus with active bleeding from the external os. Laboratory results corroborated with iron deficiency anemia. Tumor markers: CA 125, AFP and HCG were found to be within normal limits. CT abdomen/pelvis with contrast revealed a large right abdominal mass of size 18.52 x 8.61 x 16.79 cm and another parallel heterogenous mass 12.75 x 7.06 x 15.71 cm, with extension into the pelvic region. A subsequent MRI confirmed the left-sided mass to be a leiomyoma, but the right mass was concerning for a malignancy. Exploratory laparotomy with total abdominal hysterectomy/bilateral salpingo-oophorectomy was performed with excision of the draining lymph nodes. The histopathology showed a high-grade tumor morphology with a Ki 67 proliferative index of 92%. There was an immunophenotypic resemblance to plasma cell neoplasms (cells expressed CD79a and MUM1) and a lack of expression of typical B-cell markers (CD19 and CD20). A standard workup for multiple myeloma was found to be negative. Since these lymphomas have a strong association with chronic viral infections, HIV, HHV-8, and EBV were also ruled out. A diagnosis of PBL was established based on immunohistochemistry, Ki 67 index >90% and extranodal clinical presentation. The patient was treated with 6 cycles of EPOCH along with intrathecal methotrexate (CNS prophylaxis). She is being monitored with PET-CT scan every 6 months and is currently in remission for the past 1 year.

PBL poses a diagnostic and therapeutic dilemma because of its complex immunohistochemistry and aggressive clinical course. The overall prognosis is poor (9 to 15 months). Immunocompromised patients are particularly vulnerable with the oral cavity, lymph nodes and skin being the most common sites of involvement. There is still no established standard of care. Less conventional treatment modalities like autologous stem cell transplantation, bortezomib, lenalidomide, and brentuximab have been tried with limited success. Given the rarity associated with this condition, extensive prospective studies are unlikely to be performed. However, their inclusion in larger clinical trials for aggressive B-cell lymphomas may yield promising results.
A Case of Statin-Induced Necrotizing Autoimmune Myopathy

Statins, are one of the most widely used medications for reduction in cardiovascular risk and mortality. Statins inhibit the 3-hydroxy-3-methylglutaryl-coenzyme A reductase enzyme, the key rate-limiting enzyme of cholesterol biosynthetic pathway and inhibits the de-novo cholesterol biosynthesis. Although statins are mostly well tolerated, muscle toxicity related to statins is a frequent phenomenon which includes myalgia, myositis, decreasing exercise tolerance, asymptomatic elevation of creatine phospho kinase (CPK) and statin induced necrotizing autoimmune myopathy (SINAM). The latter one is very rare, affecting about 2-3 patients per 100,000 treated per year.

We are presenting a case of 64-year-old female with history of hypertension, hyperlipidemia and diabetes mellitus was referred by her primary care physician (PCP) after blood work revealed elevated CPK in the range of 5000s. Patient reported gradual progressive weakness of her thigh muscles for last 9 months, now involving proximal upper extremities. The muscle weakness was becoming more profound and limiting her daily life activities. Patient had been taking atorvastatin for last many years, until 9 months ago, as was discontinued for suspicion of statin induced myopathy. Her CPK was then elevated to 500s. However despite the discontinuation of the medication, the levels remained persistently elevated. Review of symptoms was negative for weight loss, dysphagia, joint pain, skin rash and discoloration of digits. Physical examination was remarkable for symmetric 4/5 muscle strength in all proximal extremities. Rest of physical examination was negative. Laboratory results revealed CPK of 7,536 Units/L and aldolase levels of 78 U/L. Thyroid function tests were normal. Electromyography revealed irritable myopathy. Workup to rule out connective tissue disorder i.e, Polymyositis and Dermatomyositis was done. Tests for autoantibodies including antinuclear antibody, rheumatoid factor, anti RNP, anti-Ro/SS-A, anti-La/SS-B and myositis panel were negative. Muscle biopsy revealed clusters of inflammatory cells mainly lymphocytes and some macrophages between muscle fibers, some loss of striation suggestive of necrosis. Age appropriate cancer screening in past had been negative. She was diagnosed with SINAM provided her history of statin exposure and absence of connective tissue disorders.

She was started on prednisone 1mg/kg with suboptimal response initially and was subsequently started on weekly methotrexate therapy in combination with tapering prednisone dose with better clinical outcomes. Unfortunately after a few months she had a relapse and was started on mycophenolate mofetil. She responded well to treatment with near normal muscle strength and is currently in remission.

Till date, no clinical trials of treatment for SINAM have been conducted. The current evidence based consensus for initial therapy includes starting oral prednisone 1 mg/kg per day. Steroid sparing medications includes methotrexate, azathioprine, or mycophenolate mofetil.

It should always prompt physicians if patients continues to have persistent muscle weakness and elevated muscle enzymes, despite discontinuation of statins.
A rare cause of polycythemia.

Introduction:

Polycythemia is defined as an increased concentration of hemoglobin (Hgb) in the peripheral blood; Hgb > 16.0g/dL in women or hematocrit (Hct) > 48% or a Hgb > 16.5g/dL in men (Hct > 49%). Secondary polycythemia can be caused by increased erythropoiesis caused by erythropoietin (Epo) stimulation in response to hypoxia. Pulmonary arteriovenous malformations (PAVMs) are a rare clinical cause of hypoxemia (prevalence of 1 in 2600) (Shovlin, 2014), in which 25% develop secondary polycythemia (Gossage & Kanj, 1998). We present a case of an 83-year old male who initially presented with heat stroke and was found to have a Hgb of 22.3g/dL with persistent hypoxia prompting an investigation which revealed PAVMs.

Case Presentation:

An 83-year old male, with a past medical history of subdural hematoma, presents with fever and transient altered mentation consistent with a heat stroke. Facial plethora and finger clubbing were noted on presentation. While being treated, he was noted to have polycythemia which persisted despite adequate hydration. The hemoglobin/hematocrit was 22.3/67.3. The JAK-2 mutation study was negative and blood erythropoietin level was raised at 23.8 mIU/mL (N = 2.6-18.5mIU/mL). The patient underwent therapeutic phlebotomy which was terminated after 200cc as the patient became cyanotic and oxygen saturation was noted to be 80%.

Lung auscultation was normal and chest radiography revealed no significant lung parenchymal changes. A computed tomography angiography (CTA) was performed and read as a right lower lobe pneumonia. However, pneumonia directed therapy failed to resolve hypoxemia leading to consideration of anatomical right to left shunt. A contrast transthoracic echocardiogram (using agitated saline) was done which revealed no evidence of an intracardiac shunt and an elevated pulmonary artery systolic pressure of 41mmHg (normal <35mmHg).

A closer look at CTA revealed dilated blood vessels in the right lower lobe and left mid zone with a feeding pulmonary artery vessel and a draining pulmonary venous vessel indicative of arteriovenous malformations. The patient and his family declined any intervention.

Discussion:

PAVMs are abnormal direct communications between pulmonary arteries and veins without interpositions of a capillary bed. The pathogenesis is unknown, but the fundamental defect is a right to left shunt from pulmonary arteries to the pulmonary veins, and the magnitude of shunt determines the clinical effects on the patient (Khurshid & Downie, 2002). Shunt related hypoxemia is compensated by a polycythemic response and high cardiac output. While PAVMs are not a common clinical problem, it remains an important diagnosis to consider in cases of refractory hypoxemia and secondary polycythemia as demonstrated by our case report.
Immune Reconstitution Response Syndrome (IRIS) following a change in antiretroviral therapy (ART)

Background:
Immune Reconstitution Response Syndrome (IRIS) is described occurring at the initiation of antiretroviral therapy (ART) or within the first few months. There is a lack of literature on the development of IRIS on a change in ART. As the life expectancy of HIV patients increase, so does the chance of resistance to ART and the need to change it. We describe the case of a patient who developed IRIS after 10 days on switching from one form of ART to another.

Case Presentation:
A 42-year-old woman on ART since 1999 presented to emergency department with a rash on her face, high-grade fever, cough and shortness of breath with bibasilar crepitation and toxic appearance. Her last CD4 count was 15 (two months prior to presentation). She was empirically started on vancomycin, piperacillin-tazobactam, acyclovir and fluconazole. She was also given azithromycin weekly and sulfamethoxazole-trimethoprim daily for prophylaxis. Over the next week, the patient continued to spike fever in spite of negative multiple blood, sputum and urine cultures. On review of records, she was switched to Biktrarvy (Bictegravir-emtricitabine-tenofovir alafenamide) from Odefesy (Emtricitabine-rilpivirine-tenofovir alafenamide) based on her latest HIV PhenoSense GT Plus Integrase drug sensitivity and phenotyping two weeks ago. A new CD4 and HIV viral load were done. It revealed an increase of 3880 % in CD4 counts and a decrease of 95 % in HIV viral load. She was diagnosed to have Pneumocystis Carinii Pneumonia (PCP) and Pityrosporum Folliculitis (a fungal infection) – IRIS skin infection. She was treated with oral steroids and treatment dose of sulfamethoxazole-trimethoprim with no further recorded fever after 24 hours and complete recovery over the next few days.

Conclusion:
Although literature describes IRIS only during the start of ART, a change in ART (based on resistance pattern, compliance issues or any other cause) can also trigger hyper-immune response and lead to IRIS. Large studies on this subset of patients are lacking. Awareness, knowledge, and counseling regarding IRIS should be done and patients closely followed for the next few months to avoid dreadful complications.
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Severe obstructive Esophageal Candidiasis in Poorly Controlled Type 2 Diabetes Mimicking Esophageal Malignancy

Introduction

Esophageal Candidiasis is a very common opportunistic infection in patients with impaired cellular immunity. The main risk factor is systemic immunodeficiency such as HIV with acquired immunodeficiency, chemotherapy, radiation to the neck region, and chronic systemic steroid or immunosuppression therapy. However, patients without systemic immunosuppression may also develop esophageal candidiasis such as patients with diabetes mellitus, achalasia, scleroderma, or those with chronic use of topical inhaled corticosteroids. Here we present a case of severe obstructive candidiasis in a patient with poorly controlled diabetes mimicking malignancy on presentation and imaging.

Case Description

A 60-year-old female with HIV infection compliant with HAART [last CD4: 454 (41%); viral load<20 copies/mL], poorly controlled diabetes mellitus type 2 (last Hemoglobin A1C: 9.6), and a 48 pack-year tobacco history presented to the emergency department with progressive dysphagia to solids for few weeks associated with regurgitation, coughing and substernal chest discomfort along with subjective weight loss. On admission, computed tomography chest/abdomen showed a thick-walled esophagus with large partially necrotic bulky ulcerated distal esophageal mass (Figure 1,2). Esophagogastroduodenoscopy revealed severe candidal pan-esophagitis along with severe esophagitis and edematous mucosa in the distal esophagus with further involvement of the gastric fundus (Figure 3). Biopsy showed marked acute and chronic inflammation associated with periodic acid-Schiff stain positive fungal forms histomorphologically compatible with Candida species with superficial mucosal invasion. Patient was treated with intravenous fluconazole and symptoms rapidly improved.

Discussion

The prevalence of Esophageal Candidiasis in HIV patients has decreased from 42.8% to 16.7% in the HAART era. Symptoms may include epigastric pain, heartburn, odynophagia and dysphagia. Severe cases can lead to esophageal bleeding, strictures, or fistula formation. This case illustrates an unusual presentation of Esophageal Candidiasis causing rapidly progressive obstructive symptoms in a patient with poorly controlled diabetes mellitus mimicking malignancy clinically and on imaging.
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SOLDIER WITH INTERMITTENT BLOODY STOOLS

History and Physical Exam:

28-year-old man presented with bright red blood per rectum (BRBPR) for the past 1 year. He had been seeing blood on toilet paper and in the toilet bowl every few weeks. Episodes would last 1 week at a time. There was no abdominal pain, nausea, vomiting, diarrhea or constipation. There were no fevers, chills, night sweats or weight loss. His only past medical history included Latent Tuberculosis treated with six months of Isoniazid. His current medications included Atovaquone-proguanil prescribed as prophylaxis for Malaria in preparation for trip to Kenya. Patient was born and raised in Kenya. At age 17, he moved to the United States and enlisted in the Marines. He has been deployed to Afghanistan, Japan and Thailand over the past few years. He lives in the state of New York, but often travels to Kenya, most recently about 6 months ago. Vitals were normal. Chest, abdominal and neurologic exams were normal.

Hospital Course:

Complete blood cell count was normal except for 700 eosinophils per cubic millimeter (reference range 100 - 500). Basic metabolic panel was normal. Review of liver function tests showed mild abnormalities over the past 3-4 years: Aspartate Aminotransferase had ranged between 37 to 86 units/L (reference range 6 - 32), Alanine Aminotransferase had ranged between 51 to 93 units/L (reference range 10 - 55). Alkaline Phosphatase and Total Bilirubin had remained normal. Ultrasound of the abdomen was normal. No hepatosplenomegaly was visualized. HIV screen, stool ova and parasite exam, Hepatitis B core antibody and Hepatitis C antibody were negative. Colonoscopy revealed sigmoid erythema, which was biopsied. Pathology revealed intramucosal ova in the sigmoid mucosa consistent with Schistosomiasis. Subsequently, the patient was lost to follow up. Praziquantel was mailed to the patient.

Conclusions:

Differential diagnosis in an individual with peripheral eosinophilia and BRBPR is broad and includes both infectious and non-infectious causes. Inflammatory bowel disease and eosinophilic colitis are examples of non-infectious causes. In this patient with an extensive travel history, infectious causes must be considered. Ancylostoma duodenale, Necator americanus and Strongyloides stercoralis are a few important pathogens that cause gastrointestinal disease and eosinophilia, although usually not associated with BRBPR. Schistosomiasis can cause hematochezia with eosinophilia. Although the discovery of Schistosomes on a colon biopsy was relatively surprising in New York State, it should not be too much of a surprise in this young patient who traveled frequently to an endemic region and now presents with hematochezia and peripheral eosinophilia.
AN UNUSUAL CASE OF POSTMENOPAUSAL BLEEDING

Primitive neuroectodermal tumors are highly malignant neoplasms characterized by small round cells of neuroepithelial origin. Primitive neuroectodermal tumors of the genital tract are rare and lack universally accepted treatment guidelines.

A 66-year-old woman presented to her doctor with a one month history of vaginal bleeding. Her last menstrual period was ten years prior. She denied any history of abnormal pap smears. Associated symptoms included a ten pound weight loss over six months. Physical exam revealed a large pelvic mass. Labs notable for anemia and a normal CA-125. A computed tomography (CT) scan of the chest, abdomen, and pelvis showed a large heterogeneous uterine mass concerning for malignancy. An endometrial curettage was performed, which showed a high grade uterine neoplasm with neuroendocrine features.

A total abdominal hysterectomy and bilateral salpingo-oophorectomy, as well as pelvic and para-aortic lymphadenectomy and infracolic omentectomy was then performed. No evidence of extra-uterine tumor spread was identified. Gross pathology revealed a lobulated tan rubbery uterine mass measuring 15.2 x 13.5 x 13.4 cm with extensive central necrosis and calcification. The final diagnosis was primitive malignant neoplasm, most consistent with primitive neuroectodermal tumor.

The patient initially declined chemotherapy and was treated with external radiation therapy to the pelvis followed by intravaginal brachytherapy. She was asymptomatic for eight months following radiotherapy. However, she then developed severe back pain, requiring hospitalization. Magnetic resonance imaging (MRI) showed multiple lesions at the L3 and L4 vertebral bodies. CT of the chest, abdomen, and pelvis also showed interval development of multiple bilateral pulmonary nodules and a heterogeneous mass superior to the left kidney. She also reported right shoulder pain and was found to have sclerosis, suggestive of metastatic disease. She then underwent five fractions of stereotactic body radiation therapy to the L3 and L4 vertebral bodies and adjacent nerve roots and the right shoulder.

At this point, the patient elected chemotherapy and was started on weekly carboplatin and etoposide. After three cycles of treatment, CT scan demonstrated marked interval improvement of the bilateral pulmonary nodules and left adrenal lesion. Given her excellent response to treatment, her regimen was switched from weekly to two every three weeks. Currently, the patient maintains active treatment and remains symptom free.

This case illustrates the effectiveness of a multimodal treatment approach in primitive neuroectodermal tumors. Although our patient was diagnosed at an advanced stage, she has responded remarkably well to systemic therapy. These results will hopefully serve as a guide for future treatment of this rare disease.
Hepatic glycogenosis presenting as abdominal pain with significant lactic acidosis.

Introduction:

Hepatic glycogenosis is a rare likely under-reported syndrome that occurs in individuals with severely uncontrolled diabetes. This occurs more often in fragile type I diabetics where glucose control is of paramount difficulty. Symptoms usually consist of abdominal pain, anorexia, nausea, vomiting, and tender hepatomegaly. With this comes mild transaminitis AST and ALT <250, without loss of synthetic function from the liver.

Case:

We present a 20 year old thin female with a past medical history of poorly controlled type I diabetes, complicated with recurrent DKA who came to the ER for complaints of severe mid epigastric pain, non radiating to the back with elevated lipase (393), mild transaminitis AST (34), ALT (34), alkaline phosphatase (191), without elevations in bilirubin or INR with normal albumin. Lactic acid noted to be 4.5 on admission which peaked to 12.4 within 2 days of admission. Serum Beta-hydroxybutyrate was undetectable throughout. Imaging studies were negative for mesenteric insufficiency or ischemia however showed significant hepatomegaly (25cm craniocaudal). Immunological, metabolic and rheumatological testing was complete for sources of liver pathology without conclusion and deemed necessary for liver biopsy. Biopsy showed prominent glycogenosis with focal mild steatosis. Patient was continued on strict glucose control with improvement of lactic acid and clinical symptoms, and was discharged home in full health.

Discussion:

Currently it is theorized that hepatic glycogenosis is due to intermittent episodes of hyperglycemia followed by hypoglycemia. During periods of significant hyperglycemia, glucose freely enters the hepatocytes driving glycogen synthesis. This is followed by superphysiologic doses of insulin which further augments glycogenesis. Ultimately leading to significant buildup of glycogen with injury, and steatosis to the hepatocytes. With this can come accompanying lactic acidosis. It is well known that in the liver through the Cori cycle, lactic acid is metabolized. Through this we hypothesize that the liver dysfunction from overt hepatic glycogenosis may have contributed to the inability to process lactic acid in our patient, leading to significantly elevated serum concentrations of lactic acid. Leading us to believe this to be the cause of her persistant lactic acidosis.

Conclusion:

Important to note that young patients with uncontrolled diabetes with painful hepatosplenomegaly and unexplained transaminitis with lactic acidosis it may be important to consider hepatic glycogenosis, requiring tighter control of their hba1c.
A TRIPLE HIT: HEMOPHILIA A IN A FEMALE WITH COMBINED HETEROZYGOUS MUTATIONS AND FACTOR VIII INHIBITOR

Introduction: An X-linked recessive disorder, Hemophilia A usually affects males. We report a rare case of hemophilia A in a female, who is a compound heterozygote and subsequently developed factor VIII inhibitor.

Clinical Vignette: The patient is a phenotypically normal Hispanic female born to unrelated parents after an uncomplicated pregnancy. Family history was noteworthy for menorrhagia in the mother and stroke in her father. She was diagnosed with Hemophilia A at age 6 months after prolonged bleeding from a venipuncture site, requiring hospitalization and cryoprecipitate administration. Early coagulation profile testing revealed a prothrombin time of 12.1 seconds and prolonged activated partial thromboplastin time (PTT) of 87.6 seconds. Mixing study resulted in normalization of PTT. Factor VIII (fVIII) levels were less than 1% and von Willebrand Factor (vWF) activity level was 96%. At age 21 months, she was found to have developed a fVIII inhibitor. Ever since, she has been treated with on-demand factor eight inhibitor bypassing agent (FEIBA), which consists of activated prothrombin complex concentrates (aPCCs). The highest documented inhibitor titer was 35 units at age 3 years. Genetic analysis most interestingly revealed that she is a compound heterozygote with two fVIII gene mutations. She inherited a type I inversion from her mother, who is a carrier of a severe hemophilia A mutation. Her father’s genetic testing was normal. Patient also has a type II fVIII inversion, which she inherited as a new mutation. Her disease has been complicated by hemarthrosis, with ankles and right elbow being the main target joints. When nearing menarche, she was started on Lupron followed by oral contraceptives for suppression of menstrual bleeding. Repeat testing at age 24 years noted a fVIII level of 3% with fVIII inhibitor titer of 8.8 units.

Discussion: Hemophilia A is caused by a mutation of the F8 gene on X chromosome. It occurs almost exclusively in males due to its X-linked recessive inheritance pattern. One of the following mechanisms may explain the occurrence in a female- homozygosity, usually seen with consanguineous parents; uniparental disomy; phenotypic female with an XY genotype; compound heterozygosity or a numeric or structural abnormality in the X chromosome. Development of fVIII inhibitors is poorly understood and treatment requires recombinant plasma factors acting downstream of fVIII, essentially “bypassing” the deficient clotting factor.

Conclusion: Hemophilia A is exceedingly rare in females, with heterozygosity accounting for very few cases. Genetic testing can reveal the causative mutations. A rare side effect of treatment with recombinant fVIII is the development of inhibitory autoantibodies, referred to as fVIII inhibitors. With this phenomenon, the treatment more challenging and typically consists of aPCCs.
INTRODUCTION: Approximately 30,000 cases of confirmed Lyme Disease are reported to the CDC every year; however, some estimates based on clinical laboratory data place the actual incidence of Lyme Disease as high as 288,000 cases annually. Cardiac involvement is a relatively rare complication of Lyme Disease, thought to occur in about 10% of cases in the early disseminated phase of the disease. Atrioventricular conduction abnormalities represent an even smaller percentage, estimated to occur in 1.1% of Lyme Disease cases. When these abnormalities occur, the conduction aberrance that results can progress rapidly to complete heart block, which can be life threatening if ventricular arrhythmias develop and could necessitate a pacemaker if not addressed promptly. This case is presented to illustrate a primary presentation of Lyme carditis to improve recognition of less common symptoms of Lyme Disease and facilitate effective management to avoid serious complications of this disease.

CASE: 43-year-old male sought care at an Urgent Care facility after finding himself with an unusually low resting heart rate in the 30-40 BPM range, lower extremity swelling, and inability to complete his typical exercise routine. During strenuous exercise, his heart rate plateaued in the 70-80 BPM range, when previously it would reach 140-150 BPM. He then subsequently presented to the Emergency Department with the chief complaint of bradycardia and reduced exercise tolerance. Electrocardiogram obtained in the ED showed complete AV block with a junctional escape rhythm and a ventricular rate of 44 BPM. Chest X-ray revealed increased interstitial markings. Patient did not report an antecedent tick bite, but had been treated for cellulitis of the left axilla with cephalaxin a few weeks prior and also had experienced fever with generalized achiness about a week before this presentation. A family member living in the area had previously been treated for tick-borne illness.

MANAGEMENT: Cardiology consult was obtained, and the patient was admitted to critical care. Lyme serology was drawn. Intravenous ceftriaxone was started to treat presumed Lyme carditis. Subsequent ECGs showed mild improvement in heart block to second degree type II heart block with 2:1 AV conduction. The Western blot result was positive for Lyme IgM with three of three bands, and the Lyme IgG showed only four of the required five Borrelia-specific bands. Infectious disease was consulted, and patient was determined to be safe for discharge to continue IV ceftriaxone in the outpatient setting via a PICC line for three weeks.

RESULT: On subsequent follow up with Cardiology three weeks after discharge, AV nodal conductance had completely recovered. ECG revealed sinus rhythm and patient reported normal exercise tolerance.
A case report: Septic Arthritis due to Pasteurella multocida

Introduction: Pasteurella multocida is a gram-negative coccobacillus prevalent in the oral flora of domestic animals such as cats, dogs, and some birds [3,4,6]. It usually causes cellulitis, meningitis, and endocarditis in humans but sporadically results in septic arthritis. Human transmission occurs by bites, scratches, and occasionally through contact with animal saliva [4,5]. Here we describe a case of a middle age male with successful treatment of P. multocida infection of the ankle.

Case presentation: A 63-year-old African-American male presented to the emergency room with nausea, vomiting, and left ankle pain after a fall two days ago. Significant past medical history included lupus arthritis, type 2 diabetes mellitus, autoimmune hemolytic anemia, systemic lupus erythematosus, and surgical repair of left ankle fracture. On examination, he was febrile (102°F), hypotensive (BP 90/60 mmhg), and heart rate of 86 bpm, he had bilateral lower extremity sensory neuropathy, and ulceration of the right medial plantar surface. Labs showed leukocytosis of 15 K/uL, acute kidney injury (creatinine of 2.1 mg/dL), lactic acidosis of 5.27 mmol/L, and procalcitonin of 86.22 ng/mL.

Sepsis protocol was initiated and the patient was given intravenous vancomycin and piperacillin/tazobactam. Blood and left ankle synovial fluid cultures revealed Pasteurella multocida. The synovial fluid analysis showed a high leukocyte count (96,400 mm3) and calcium pyrophosphate crystals, indicative of septic arthritis and pseudogout, respectively. X-ray of his left foot showed a healed ankle fracture laterally with significant arthritis. MRI without contrast of the left ankle revealed widespread osteomyelitis involving the distal tibia, fibula, talus, and portions of the calcaneus. His antibiotics were switched to linezolid and meropenem and the left ankle was debrided. The patient improved with treatment and was discharged with outpatient follow-up.

Discussion: Our patient did not recall any animal scratches or bites but reported to have two cats at home. As the number of domestic pets continues to increase, the risk of Pasteurella infections will also increase simultaneously, especially in patients that are immunosuppressed [1]. Penicillin is the ideal treatment against P. multocida sepsis but in the event of resistance or allergy, intravenous second and third generation cephalosporin, tetracycline, fluoroquinolone, and amoxicillin-clavulanate, can be effective. Oral cephalosporin is not as effective because they result in low blood concentrations [2,3].
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Dyspnea and Subcutaneous Crepitus, a Deceiving Presentation to Rectal Perforation

Pneumomediastinum often occurs from chest trauma or surgical interventions. Colorectal perforations are similarly explained and more obviously diagnosed given obvious physical symptoms such as abdominal pain and tenderness. Acute spontaneous subcutaneous emphysema caused by distal sigmoid perforation with extensive extraperitoneal gas tracking presenting with dyspnea is even more rare and examined in this unique case report.

An 81 year old female presented to our institution with profound dyspnea. Her medical history was notable for severe chronic obstructive pulmonary disease, Non-Small Cell Carcinoma, currently being treated with Nivolumab. She stated that her dyspnea began three days prior and progressed to include fever and wheezing. Her initial vitals and physical exam were notable for TMAX 100.5°F, HR 100-115, appearing in moderate distress, crepitus in the left anterior neck, bibasilar wheezing, diffuse diminished breath sounds, soft, non-tender abdomen, normoactive bowel sounds with a left para umbilical hernia and old stoma site partially reducible but non-obstructing. Rectal wall defect at approx. 6cm no gross blood or palpable masses with crepitus appreciated in perirectal region.

Her laboratory analysis was notable for Lactate WNL, WBC 21.1 k/uL, Hgb 10.8, Sodium 126 mEq/L. Additionally a blood gas revealed Arterial pH: 7.30, CO2: 36, pO2: 206, Bicarbonate: 17.7 and was placed on Bi-PAP. She was then initiated on intravenous methylprednisolone and aeluetro-iptrotopium nebulization. A chest radiograph was notable for subcutaneous emphysema and LUL infiltrate without findings to suggest pneumothorax. A Computed Tomography (CT) of the chest was ordered, which showed extensive extraperitoneal gas that extended along the posterior mediastinum resulting in substantial emphysema in the left neck. This was immediately followed by a CT of the abdomen and pelvis which showed intra and extraperitoneal air with a distal sigmoid perforation just proximal to the rectosigmoid anastomosis. Despite active bronchospasm and extreme high risk for post-operative complications the decision was made to pursue a surgical intervention. The patient underwent an exploratory laparotomy resulting in small bowel resection and a primary anastomosis. In addition to sigmoid resection and sigmoid resection with descending colon colostomy, abdominal washout, lysis of adhesions was performed, and patient was monitored in the ICU. There was continued difficulty weaning off the ventilator, however on POD 7 patient was extubated and placed on BiPAP for continuous support. The remainder of her hospital course was complicated by Sepsis, aspiration PNA, fluid overload, and hyperactive delirium/anxiety. Patient was eventually placed on comfort care and expired on POD 14.

To our knowledge there are few reports of acute spontaneous subcutaneous emphysema caused by nontraumatic perforations of the colon. This report is unique as the patient usually expresses symptoms of abdominal pain, distention, decreased bowel sounds, abdominal tenderness, nausea, and vomiting.
Management of Atypical Chronic Lymphocytic Leukemia Presenting with Extreme Leukocytosis

A 65-year-old male was evaluated for extreme leukocytosis and anemia. He reported exertional dyspnea for several months, denied B symptoms, and had no neurological signs. CBC showed WBC 600K/μL, predominantly lymphocytes, hemoglobin 8.5g/dL, and platelets 92k. Peripheral smear showed numerous lymphocytes. Flow cytometry revealed a clonal population of CD5+, CD23+ B-lymphocytes consistent with CLL, Rai stage-4. CT scans revealed diffuse bulky lymphadenopathy with hepatosplenomegaly. Molecular studies showed mutated immunoglobulin heavy chain and deletion 13q, which ordinarily confer standard risk. Cytogenetics revealed t(11;14) which is the hallmark of mantle cell lymphoma (MCL), but is also present in atypical CLL, and confers a poor prognosis due to an oncogenic IGH/CCND1 translocation. The oral BTK inhibitor ibrutinib was considered as initial treatment, but given the potential for transient lymphocytosis, there was a theoretical risk of hyperleukostasis, which is unusual in CLL unless the WBC exceeds 400K/ul. Benadmustine was therefore started for initial cytoreduction. He had an excellent WBC reduction to 90K/ul after one cycle. He was subsequently started on ibrutinib with continued improvement.

Atypical CLL with t(11;14) presenting with this degree of hyperleukocytosis has never been reported. The t(11;14) is characteristic of MCL, but is identified in 2–5% of CLL cases which have atypical morphology and immunophenotype (CD5+, CD19+, sIg+, FMC7+). This CLL subset has a poorer prognosis and requires prompt treatment. A randomized study of patients with untreated CLL showed significant improvement in 2-year progression free survival with ibrutinib compared with the prior standard of bendamustine plus rituximab (87 vs 74%). However, there is a scarcity of literature to guide the management of patients with markedly elevated WBC counts and the theoretical risk of leukostasis when start ibrutinib.

This case had several unique aspects. To our knowledge, atypical CLL with t(11;14) presenting with this degree of hyperleukocytosis has never been reported. The optimal management of atypical CLL with t(11;14) is not well studied given its low incidence. This case illustrates the dilemma of modern management of CLL in the setting of extreme leukocytosis. In this case, chemotherapy served as an effective cytoreduction prior to starting ibrutinib. Larger studies are needed to determine the optimal initial approach to patients with atypical CLL with markedly elevated WBC.
HYDRALAZINE INDUCED VASCULITIS: A RARE CAUSE OF DIFFUSE ALVEOLAR HEMORRHAGE

Background

Hydralazine, a commonly used antihypertensive medication, is notorious for its role in drug induced lupus. Recently it has been shown to play a large role in drug induced vasculitis as well, resulting in a pulmonary renal syndrome, very rarely a limited pulmonary capillary vasculitis. Hydralazine induced vasculitis, compared to hydralazine induced lupus, has a more serious clinical presentation, often times life threatening. The pathogenesis is unclear however theories include hydralazine forming complexes with myeloperoxidase leading to neutrophil apoptosis which further induces antibody production. The second theory is that slow acetylators develop antibodies easier than fast acetylators. Here we present a case of hydralazine induced vasculitis presenting as pulmonary capillary vasculitis.

Case

This is a case of a 93 year old woman with history of hypertension on hydralazine for three years, who presented with a five day history of hemoptysis. CT chest showed diffuse mixed airspace and ground glass densities throughout the lungs. On bronchoscopy she had diffuse alveolar hemorrhage. Cytology was negative for malignancy. She had an elevated APTT which subsequently led to checking APTT mixing levels, which were abnormal. This warranted a full rheumatologic workup. She had positive Lupus anticoagulant, ANA, ANCA, anti MPO antibodies, anti histone antibody, and positive dsDNA antibodies. Hydralazine was discontinued and she was started on high dose corticosteroids and inhaled tranexamic acid for hemoptysis. She also required several units of transfusion support throughout her hospitalization. Unfortunately her symptoms did not subside, and she was started on azathioprine in addition to a long taper of steroids. Shortly after initiation of immunosuppressant, she symptomatically improved.

Discussion:

This case highlights the importance of being aware of the potential side effects of commonly used medications. Our patient had hydralazine induced vasculitis resulting in diffuse alveolar hemorrhage. It was her elevated APTT that warranted the full rheumatologic workup. Contrasting from hydralazine induced lupus, vasculitis commonly has a positive ANCA titer, with anti dsDNA antibody as well as anti MPO antibody. Generally, hydralazine induced vasculitis will have a mixed renal-pulmonary disease. Our patient had an isolated pulmonary component, without any renal involvement. Unlike other types of drug induced vasculitis and lupus, hydralazine requires aggressive treatment. Cessation of hydralazine alone is not appropriate, patients will require immunosuppressive therapy as the condition can be life threatening.
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SARCOIDOSIS: THE GREAT MIMICKER!

Background:
Sarcoidosis is a multisystem disorder characterized by non-caseating granulomas. Bone and joint involvement itself is uncommon, with bone involvement seen in 5% of cases. Diagnosis of vertebral sarcoidosis is difficult without biopsy. Imaging including MRI can be misleading as sarcoidosis appears as widespread, multiple, lytic lesions, mimicking metastasis.

Case:
This is a case of a 55 year old woman with remote history of pulmonary sarcoidosis. She had sustained an injury to her lower back, following which an MRI of her lumbar spine was obtained. MRI revealed abnormal bone marrow signal intensity with enhancement, involving the lumbar spine, suggestive of multiple myeloma and metastasis. With concerns for malignancy she was referred to hematology/oncology. Serum protein electrophoresis, urine protein electrophoresis, and serum immunofixation were not suggestive of monoclonal proteins. A bone scan did not reveal any osseous metastatic disease. However, a CT abdomen showed multiple enhancing lesions in the lumbar spine suggestive of osseous manifestation of sarcoidosis. A CT guided bone biopsy was obtained, which revealed small non-necrotizing granulomas consistent with sarcoidosis. She was subsequently started on methotrexate for treatment of sarcoidosis, with improvement in her back pain.

Conclusion:
Patients with vertebral sarcoidosis can often be asymptomatic. They commonly present with pain, more so in the upper lumbar area. Diagnosis is often misleading. Vertebral sarcoidosis appears as lytic lesions with peripheral sclerosis, commonly in the vertebral bodies and pedicles, on imaging. Lesions can be multiple in number and widespread, affecting multiple areas of the axial skeleton and often gets mistaken for malignant lesions. Axial skeleton sarcoidosis is becoming more largely recognized, one study of 20 patients with osseous sarcoidosis, revealed that 90% had vertebral sarcoidosis. This case highlights the importance of recognizing the prevalence of osseous as well as vertebral sarcoidosis, and the utility of bone biopsy to clinch the diagnosis.
Resident/Fellow Clinical Vignette

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Pulmonary Embolism after Intravenous Immunoglobulin Infusion in a Patient with Guillain Barre Syndrome: Will Standard Prophylactic Anticoagulation do the Job?

Learning Objectives

- To recognize pulmonary embolism as a fatal complication in Guillain-Barre Syndrome patients treated with intravenous immunoglobulin (IVIG)
- To learn preventive measures for thrombotic events in patients who receive IVIG

Case

55-year-old man with hypertension presented with numbness, tingling and weakness of arms and feet and gait instability for five days. Physical examination revealed right facial droop, reduced motor strength in bilateral upper and lower extremities, absent ankle reflexes and wide-based gait. He had a flu-like illness two weeks prior to presentation.

Cerebrospinal fluid analysis revealed cytoalbuminologic dissociation. Guillain-Barre Syndrome (GBS) - Miller Fisher variant (MFS) was diagnosed. IVIG was initiated at 0.5 G/kg for five days. Lower extremity weakness improved gradually and respiratory status was stable. He was on prophylactic dose of subcutaneous heparin 5000 units every twelve hours. He had no personal or family history of venous thromboembolism.

On Day 9, he suddenly developed dyspnea with hypotension, tachycardia and hypoxia to 70% on room air. Point of care ultrasound revealed a thrombus in transit between right atrium and right ventricle and dilated right atrium with positive McConnell’s sign. He was emergently intubated and tissue plasminogen activator was administered. Repeat Echocardiography did not identify any clot in right heart or inferior vena cava. He was on intravenous heparin drip for anticoagulation and then bridged to warfarin. He was eventually extubated and discharged on Day 20 of hospitalization.

Discussion

GBS is the most common cause of acute flaccid paralysis in the United States with the incidence of 1.2-3 per 100,000 annually. Amongst the GBS variants, MFS classically present with ophthalmoparesis, areflexia and ataxia, or often can present with other features like facial weakness.

IVIG and plasmapheresis are the mainstay of treatment for GBS. Nowadays, IVIG is the preferred treatment because of the greater convenience and availability. However, IVIG is associated with serious adverse effects, including thrombosis, both arterial (myocardial infarct, stroke) and venous (Pulmonary embolism). Possible underlying mechanisms include hyperviscosity, platelet and vasoactive cytokine activation, and vasospasm. Predisposing factors include advanced age, previous thromboembolic diseases, diabetes mellitus, hypertension, dyslipidemia, high dose IVIG or rapid infusion rate, and, as in our patient, immobility.

Measures that potentially reduce IVIG associated thrombosis include avoiding high dose infusion, slow infusion rate, hydration, and use of prophylactic antiplatelet or anticoagulation. However, there are limited studies that evaluate which IVIG dosing protocols are associated with a lower risk of thrombosis. In our case, standard heparin prophylaxis failed to prevent fatal thromboembolism. It raised a concern that standard prophylactic dose may be insufficient to prevent thromboembolism in patients receiving IVIG. Further studies are needed to compare the efficacy of different doses of prophylactic anticoagulation in such high risk patients.
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An unusual case of polyuria and polydipsia

Polyuria and polydipsia more often than not direct the clinicians towards two possible diagnoses, diabetes mellitus and diabetes insipidus. DI typically present with polyuria, nocturia, and polydipsia. The serum sodium concentration in untreated DI is in the high normal range, serum osmolarity is high and urine osmolarity is low, usually <300.

We will present a case of a patient with polyuria and polydipsia was initially diagnosed and treated as diabetes insipidus which turned out to be something different.

Case: 72-year old Caucasian man with polyuria, polydipsia and nocturia gradually worsened in 6-months period. Pt complained of increase in fatigue and sleepiness, also reported leakage of urine and incontinence. Weight has been stable, denied any fever, chills, dysuria, cough, diarrhea, abdominal pain. Pt denied any headache, visual changes, back pain and kidney stones. His most recent HbA1c-5.5. Allergy: Penicillins. Family history “father died of unknown cancer. Social “ex smoker since 1984, denied drinking alcohol, retired lawyer. Home Med: Vitamin C & D, OTC prostate formula.

Physical examination was unremarkable including abdomen and pelvic examination. Labs are remarkable for elevated BUN/Cr 59/2.65(high), Na-146 (high), Cl-112, serum osm-318(high), urine Osm-269 (low). Urine specific gravity- 1.008, UTI-negative. Pt was started on desmopressin for 2 weeks for presumed central DI but had no change in his symptoms. Later AVP-7.1. PSA-6.050 ng/ml. Central DI unlikely due to high AVP level and also lack of response to desmopressin. US kidney shows large post void residual urine volume of 500cc, bilateral hydronephrosis and hydroureter, bladder wall trabeculation and irregularity. At this point, the most likely diagnosis is partial obstruction of urinary tract, causing back-pressure hydronephrosis, renal impairment and eventual loss of renal tubular ability to concentrate the urine with polyuria.

It is true that complete bilateral obstruction or unilateral obstruction of a single functioning kidney such as a renal transplant will result in anuria. However, when the lesion results in partial obstruction, urine output may be normal or increased (polyuria) as in this case. Urinalysis may be completely negative despite advanced obstructive nephropathy. With chronic progressive obstruction (>12 weeks), there is often irreversible and severe renal damage, the resultant polydipsia, polyuria are common features of renal tubular disorder and renal functional recovery may be limited even after relief of the obstruction. Surgical intervention with transurethral resection of the prostate is generally required for debilitating symptoms of obstruction or evidence of renal parenchymal damage. Physicians should keep in mind that while polyuria and polydipsia often indicates diabetes mellitus or diabetes insipidus but they may also be indicative of partial urinary obstruction.
TRAVEL TROUBLE; AN UNUSUAL CASE OF CHOLESTATIC HEPATITIS

Learning objective:

- Outline the importance of travel history in the formulation of differential diagnosis.
- Identify salmonella bacteremia as a cause of acute cholestatic hepatitis without cholangitis.

Case:

A 65-year-old man with no known past medical history presented with one week of fever, right upper quadrant abdominal pain, jaundice, and diarrhea. He had scleral icterus and distended abdomen. The neurologic exam was normal without asterixis. Labs revealed total leukocyte count 8000 u/L, serum sodium 122 mg/dL, Alkaline phosphatase 642 mg/dL, total bilirubin 9.3 mg/dL, alanine aminotransferase 102 mg/dL, aspartate aminotransferase 114 mg/dL and international normalization ratio 1.9. Viral hepatitis serology was negative. Imaging was suggestive of colonic mural thickening and splenic infarction. The liver was 19 cm with no evidence of cholelithiasis or biliary dilatation. Blood cultures grew Salmonella typhi (S. typhi). The patient was diagnosed to have enteric fever complicated by bacteremia and cholestatic hepatitis. A transesophageal echocardiogram was negative for endocarditis. On questioning, the patient revealed recent travel to India. He was treated with ampicillin. His total bilirubin peaked to 19 mg/dL before returning to baseline.

Discussion:

Salmonella infection has an estimated annual incidence of over 20 million cases worldwide with approximately 300 cases per year in the USA. Since humans are the only known reservoir of S. typhi, a travel history to endemic areas or contact with a known typhoid case is useful to establish the diagnosis. S. typhi is acquired through the consumption of contaminated water or food. Classic features include fever, chills, relative bradycardia followed by abdominal pain and appearance of rose spots. S. typhi can affect almost all major organ systems, including hepatobiliary, cardiovascular, respiratory, genitourinary, musculoskeletal, and central nervous systems by bacteremic seeding; however, severe hepatic involvement with acute cholestatic hepatitis is a rare complication. Hepatic involvement in S. typhi was first described in 1899 and the mechanism was postulated to be either direct invasion of the organism or immune-mediated liver injury. Treatment options include fluoroquinolones, third-generation cephalosporins, and azithromycin. Carbapenems are reserved for suspected infection with extensively drug-resistant (XDR) strains.

Conclusion:

- Typhoid fever should be considered when patients from endemic areas present with acute febrile hepatitis.
- Due to the high incidence of complications, it is very important to establish an early diagnosis and rule out the differential diagnosis to improve patient outcomes.
IDIOPATHIC INTRAMURAL HEMATOMA OF THE SIGMOID COLON

INTRODUCTION:

Intramural colonic hematoma (ICH) is usually seen secondary to blunt trauma to the abdomen, use of anticoagulation or with blood dyscrasias. Idiopathic ICH is extremely rare. Due to its rarity and non-specific presentation, the diagnosis is challenging. We present a case of a patient with complications from idiopathic ICH requiring surgical intervention.

CASE:

A 45-year-old man was with a past medical history of quadriplegia from a gunshot wound to the cervical spine, tracheostomy with ventilator-dependent respiratory failure, percutaneous endoscopic gastroscopy (PEG) tube on feeds, recurrent ventilator associated pneumonias, major depression and anxiety disorder was admitted to medicine with septic shock from pneumonia. During the hospitalization he was noted to have abdominal distention and anemia. Computerized tomography (CT) scan of the abdomen was done which showed a hematoma within the wall of the sigmoid colon. He was managed conservatively for the hematoma with clinical improvement and discharged to a nursing home. He presented eight days later with bright red blood per rectum. Repeat CT scan of the abdomen showed large amount of dense peritoneal fluid and free air consistent with perforation. Exploratory laparotomy revealed a distended sigmoid with a large mural sigmoid hematoma and 2500 ml peritoneal fluid secondary to perforation. Sigmoid colectomy and end colostomy with peritoneal washing was done. Pathology revealed an extensive blood clot dissecting the sigmoid colon between mucosa and sub-mucosa leading to perforation.

DISCUSSION:

ICH has been well described in the literature usually secondary to trauma and in patients on anticoagulation or with blood dyscrasias such as hemophilia. Idiopathic ICH is extremely rare comprising <5% of all ICH. Abdominal pain for a few days is the most common presentation. Our patient had loss of sensation below the shoulders and could not perceive the pain from the ICH. In majority of cases, the expanding ICH usually leads to intestinal obstruction. Intestinal perforation, bleeding and hemoperitoneum are some of the other clinical manifestations. Patients are usually managed conservatively. Surgical management is indicated if complications develop. Our patient was also managed with a conservative approach initially and once there was evidence of a complication (perforation), surgical intervention was undertaken.

CONCLUSION:

In conclusion, colonic perforation secondary to an idiopathic ICH is very rare. Given the absence of any known trauma, exposure to anticoagulation or bleeding diathesis, we believe our patient meets the criteria for idiopathic ICH. In our patient, an enlarging mural hematoma dissected through the colonic wall leading to colonic ischemia and then perforation. Patients with an intramural hematoma in the gastrointestinal tract should be monitored closely, and surgical treatment considered if symptoms or signs of complications develop.
Tearinâ€™ Up My Heart: A Case of Bacterial Endocarditis Caused by Non-Typhoidal Salmonella

Intro

Salmonella species are a rare cause of endocarditis. Highlighted is a case of non-typhoidal Salmonella (NTS) endocarditis presenting in a relatively healthy male with history only of opioid abuse on suboxone. Mortality is as high as 42.5% and early recognition and treatment of this condition is vital to patient survival.

Case

A 40-year-old man with a past medical history of opioid dependence on suboxone was admitted with fever, night sweats, diarrhea and chills of three weeks duration. During this period the patient reported twenty-pound unintentional weight loss. Temperature was measured to reach a maximum of 101.6 F on presentation with otherwise normal vital signs. Physical exam was normal with the exception of hepatosplenomegaly. Laboratory data was only remarkable for mild leukopenia. Initial imaging consisted of normal chest x-ray and computed tomography of the abdomen and pelvis which revealed hepatosplenomegaly as well as soft tissue swelling at the site of the patientâ€™s suboxone injections. Extensive serological, bacterial and viral infectious workups were performed without positive findings. Human Immunodeficiency Virus antibody testing was negative. Two sets of blood cultures drawn on presentation one hour apart revealed gram-negative rods which later speciated to salmonella species confirmed by the New York State Department of Health. A transesophageal echocardiogram was performed which revealed vegetations of the aortic and mitral valves as well as trace mitral regurgitation consistent with Salmonella endocarditis. A peripheral intravenous central catheter was placed and the patient was discharged home with a prolonged course of outpatient ceftriaxone and went on to a full clinical recovery.

Discussion

Salmonella species are estimated to cause more than 1.2 million illnesses each year in the United States alone with 23,000 hospitalizations and 450 deaths. NTS is most commonly due to Salmonella enterica. Salmonella is acquired most commonly from ingestion of poultry, eggs, milk products, animal contact and can infect any organ but rarely involves the cardiovascular system. A meta-analysis of 16 case series of 87 cases of bacterial endocarditis SE only accounted for 0.01%-2.9% of cases. In these case series the most commonly involved valve was the mitral valve, unlike in the described patient who also had aortic valve involvement. Mortality of SE and its sequela are estimated to be as high as 42.5% but has been improving as of late. Presentation usually includes fever, murmur and heart failure. Antecedent diarrhea is rare. Patients with HIV, diabetes mellitus and underlying valvular defects have a higher risk of SE. Management is similar to other forms of bacterial endocarditis including extended courses of intravenous antibiotics with or without cardiac surgery. Flouroquinolones or third generation cephalosporins are the antibiotics of choice. Considering its associated poor outcomes, any patient with salmonella bacteremia should undergo echocardiography.
A 38-year-old Caucasian male with type 1 diabetes mellitus presented with anuria, acholic stools, jaundice, scleral icterus and arthralgias. He was febrile to 101.4°F, had a blanching, petechial rash and diffuse tenderness on the lower extremities and scleral icterus. Initial labs showed a hemoglobin 10.2 g/dL, hematocrit 28.9%, platelets 43,000/mm³, sodium 125 mEq/L, potassium 3.9 mEq/L, chloride 90 mEq/L, bicarbonate 27 mEq/L, BUN 17 mg/dL and creatinine 1.0 mg/dL. Total bilirubin 12.1 mg/dL with a conjugated bilirubin 8.6 mg/dL, ALT, AST and alkaline phosphatase were 160 IU/L, 192 IU/L, and 286 IU/L respectively. During hospitalization hemoglobin downtrended as low as 7 g/dL. Hemolysis labs returned positive. Peripheral smear revealed significant inclusion bodies in a small proportion of neutrophils. Abdominal CT imaging showed mild splenomegaly and hepatomegaly without masses. The patient had a markedly elevated ferritin of 32,383 µg/L and soluble interleukin-2 receptor (sIL2R) of 7,274 U/mL. Coombs testing for anti-C3D direct antiglobulins returned positive. The patient underwent bone marrow aspiration with biopsy and results confirmed the presence of hemophagocytosis. Our patient met six out of the eight criteria for the diagnosis of HLH, including fever, splenomegaly, anemia, thrombocytopenia, elevated ferritin, a bone marrow biopsy demonstrating hemophagocytosis, and an elevated sIL2R. The patient underwent treatment with the HLH-2004 protocol which included dexamethasone 20 mg and cyclosporine 250 mg twice daily. Etoposide, which is part of this regimen, was held in light of his hyperbilirubinemia. Given the intraerythrocytic inclusions on his peripheral smear, PCR testing for parasitic DNA was performed to investigate for tick-borne illnesses. He tested positive for Babesia microti DNA on Wright staining, with 2.8% parasitemia. Azithromycin 500 mg twice a day and atovaquone 750 mg once a day were given in addition to his HLH treatment. The parasitemia was eradicated after 5 days of antiparasitic therapy and he completed a 10 day course. The patient’s clinical status improved and the patient was discharged on dexamethasone and cyclosporine; he did not require etoposide for resolution of symptoms. Patient completed cyclosporine and dexamethasone taper after the 2 month follow up appointment and lab values all showed improvement.
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Nivolumab Induced Myasthenia Gravis Crisis and Rhabdomyolysis

Immunotherapy with monoclonal antibodies targeting human programmed death receptor 1 (PD-1) is used to treat an expanding number of malignancies thus increasing the chance of patients developing autoimmune toxicities. We report a case of rare neurological toxicity from use of Nivolumab in metastatic melanoma which presented as a Myasthenia Gravis (MG) crisis. A 77-year-old male with PMH of metastatic melanoma, initially presented in Right eye in 1996 s/p enucleation and radiation, with recurrence in the right lung treated with resection, radiation and 1 dose of Nivolumab who presented to the ED with worsening weakness. Labs showed CPK of 10,366 IU/L concerning for immunorelated rhabdomyolysis. He then developed acute hypercarbic respiratory failure, with ABG: 7.14/99/62/33.7 and was intubated. Physical exam revealed 5/5 strength and 2+reflexes, though was unable to tolerate breathing trials. Labwork was unremarkable except for Acetycholine-Receptor-binding antibodies 0.88 nmol/L, which confirmed suspicion for MG. He was treated initially for MG with high dose steroids and pyridostigmine, but when he failed to improve was treated as immunotherapy-related toxicity with IVIG, plasmapheresis, and Infliximab. Though he required a tracheostomy for prolonged intubation, with the latter treatments, his respiratory status slowly improved to allow him to breathe spontaneously for 5-6 hours at a time. The case highlights the need for a high index of suspicion to suspect immunotherapy-related myasthenic crisis when patients present with respiratory failure as it can be fatal without treatment with one study citing a 30% MG-specific mortality. European Society of Medical Oncology guidelines recommend discontinuing immunotherapy, and starting treatment with steroids, IVIG/plasmapheresis to prevent permanent sequaleae.
HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS (HLH), A RARE COMPLICATION OF IMMUNE CHECKPOINT INHIBITOR (ICI) THERAPY

INTRODUCTION

HLH is a potentially fatal, rare, hematological disorder characterized by immune dysregulation leading to excessive cytokine production by macrophages, natural killer and cytotoxic T cells. Malignancy, infections and autoimmune disorders are the usual precipitating causes. Patients typically present with fever, pancytopenia, and splenomegaly, and have markedly elevated serum ferritin levels and hypertriglyceridemia. The diagnosis of HLH requires 5 of 9 criteria to be met per the protocol used in the HLH-2004 study. Therapy generally involves treatment of the underlying cause, chemotherapy, steroids & allogeneic stem cell transplant. Even after treatment, high mortality is seen.

Immune checkpoints are present on normal cells and help in preventing autoimmunity by downregulating T cells. The expression of these proteins is dysregulated in cancer cells, and immune checkpoint inhibitors (ICIs) help in activating the immune cell against the tumor cells by blocking these checkpoints. ICIs have revolutionized cancer therapy; however, they are associated with multiple autoimmune adverse events. We present an interesting case of HLH due to ICI.

CASE PRESENTATION

A 36-year-old male with metastatic melanoma was on therapy with pembrolizumab, an ICI and propranolol. After 3 cycles, he was noted to have asymptomatic transaminitis (AST-808 IU/L, ALT-1336 IU/L). In the absence of alternate etiology, he was diagnosed with pembrolizumab-induced autoimmune hepatitis and treated with prednisone 2mg/kg, with improvement. Two weeks later, the patient presented to the hospital with necrotizing fasciitis of right groin. He underwent urgent debridement and was empirically treated with broad-spectrum antibiotics, prednisone was tapered due to concerns of delayed wound healing. Wound culture grew MRSA, antibiotics were eventually tailored to ceftaroline. Two weeks into the hospital course, the patient’s condition deteriorated inexplicably; he developed high-grade fever, hypotension, hypoxia, acute kidney injury, pancytopenia and elevated LFTs. Antibiotic coverage was broadened for sepsis, high dose methylprednisolone was started due to the concern of immune-mediated adverse effect. Blood and wound cultures remained negative. His condition stabilized, however he had persistent pancytopenia. Further workup showed ferritin of >40,000 ng/ml, elevated interleukin-2 soluble receptor alpha (s-IL2) (13799 ng/ml), hypofibrinogenemia (57 mg/dl) and hypertriglyceridemia (421 mg/dl). Bone marrow biopsy showed Hemophagocytic lymphohistiocytosis in a normocellular marrow. A diagnosis of HLH was made and methylprednisolone was increased to 115 mg every 8 hours. Over the next 2 weeks, improvement in blood counts, coagulopathy, serum ferritin, and sIL-2 was noted allowing gradual taper off steroids over the next month. Patient continues to be in remission from HLH and melanoma after 6 months of follow-up. ICIs have not been reinitiated to date.

CONCLUSION

HLH from ICI therapy is exceedingly rare with very few cases reported in the literature. Our case is unique as the patient achieved remission from HLH with steroids and did not require any chemotherapy.
LIFE THREATENING HYPERNATREMIA IN A 22 YEAR-OLD MAN WITH CONCURRENT DKA AND HHS

In diabetic ketoacidosis (DKA), hyponatremia is commonly seen due to osmotic diuresis. Therefore, hypernatremia is rare and suggestive of severe free water loss. Here we describe a case with concurrent diabetic ketoacidosis and hyperglycemic hyperosmolar state who presented with life-threatening hypernatremia. A 22-year-old type 1 diabetic male was brought in after 2 weeks of polyuria, polydipsia followed by nausea, vomiting and lethargy in the last 2 days. On admission, plasma glucose was 1230 mg/dL and measured plasma sodium was 158 mEq/L with corrected sodium level of 176 mEq/L. Patient developed acute kidney injury and anion gap acidosis with high serum lactate and ketones. Despite fluid resuscitation per DKA protocol, measured sodium level continued to rise to 180 mEq/L within 19 hours and remained extremely high for 48 hours. Although his blood pressure was always in normal range, patient’s tachycardia persisted at 140 bpm. Shock liver developed with peaked AST/ALT of 2071/729 U/L on day 2. Free water deficit was carefully calculated which showed total deficit of 10L. Decision was made to bolus more isotonic fluid for intravascular volume repletion in addition to maintenance of 0.45% NaCl with KCl according to DKA protocol. Volume-loss-induced tachycardia started to improve. Hypernatremia and hyperglycemia were decreased by a safe rate to avoid cerebral edema. Transaminitis and acute kidney injury also gradually improved. To our knowledge, this is the first case of DKA in adults with hypernatremia to 170s, lethargy, multiorgan failure requiring tedious volume evaluation, resuscitation and recovered without sequelae.
Catch Me If You Can: A Case of Carcinoid Heart Disease

Case Presentation:

We report a 66-year-old female with a history of hypertension and heart failure with preserved ejection fraction secondary to severe tricuspid regurgitation status post tricuspid valve repair a year prior, who presents with vomiting, diarrhea, decreased appetite and weight loss. The patient was recently hospitalized at an outside institution with 2 months of similar symptoms. At that time, workup included MRI imaging significant for multiple ovarian masses, and ascites with hypervascular lesions of the liver. Subsequent liver biopsy was significant for well-differentiated neuroendocrine neoplasm.

On presentation to our institution, the patient demonstrated a normal physical exam only significant for tachycardia without murmurs, and bilateral pitting edema to the knees. EKG was significant for sinus tachycardia, incomplete RBBB, and previously known T-wave inversions. High sensitivity troponin 72 ng/dL, Pro-BNP > 11,000 pg/mL. Chromogranin A was elevated to 1368 ng/ml and 24-hour urine 5-HIAA was elevated to 80 mg/24h. Echocardiography was significant for ejection fraction of 52%, stenotic tricuspid bioprosthetic valve replacement with tricuspid mean gradient of 6 mmHg, right atrial and ventricular enlargement with decreased right ventricular systolic function. Moderate pulmonic insufficiency was also present. The patient was diagnosed with carcinoid heart disease.

Discussion:

Carcinoid tumors are well-differentiated neuroendocrine tumors that arise from neural crest cells. The majority of tumors arise within the tubular gastrointestinal tract, lung and genitourinary tract and can present clinically with both systemic and localizing symptoms including diaphoresis, diarrhea, bronchospasm and skin changes. Patients with cardiac involvement present with symptoms of right heart failure, related to involvement of dysfunctional tricuspid and pulmonary valves. Our patient’s previous presentation of right heart failure and tricuspid regurgitation necessitating valve repair was likely an early manifestation of cardiac involvement of carcinoid that was discovered in subsequent imaging and biopsies.

Isolated acquired tricuspid and pulmonary valve disease is uncommon, hence, carcinoid heart disease should be considered in patients with Carcinoid tumors, especially in the absence of left-sided valve disease. Appropriate surgical intervention with a multimodality cardiovascular approach, including transthoracic and transesophageal echocardiography and cardiac magnetic resonance has demonstrated its utility with regards to guiding management. Furthermore, the choice of surgical valve prosthesis should be individualized based on bleeding risk and possible future interventions. Despite no difference in survival or reoperation rate between mechanical or bioprosthetic valves, bioprosthetic valves are favored given elevated bleeding risk in patients with hepatic dysfunction. Overall, early surgical intervention yields improved survival rates at 1, 5, and 10 years that are 69%, 35%, and 24% respectively.

This case emphasizes the importance of early diagnosis of carcinoid heart disease in patients given likely benefit from early valve surgery and other effective therapies.
Cardiac Sarcoidosis Presenting as Silent Complete Heart Block: A Case Report

Case Presentation:

A 39-year-old African American male with chronic kidney disease stage 3 and sarcoidosis presents with an abnormal EKG. The patient was treated previously for urethritis, epididymitis with associated lymphadenopathy prior to developing a bilateral anterior shin rash consistent with biopsy proven sarcoidosis. He is noted to have chronic kidney disease stage 3 and restrictive lung disease that is well maintained on chronic prednisone. Currently he has no symptoms of dizziness, shortness of breath, chest pain or peripheral edema. Initial vitals were significant for bradycardia in the 40’s. Initial laboratory findings were consistent with stage 3 kidney disease, hepatocellular pattern liver enzyme elevation and creatinine kinase of 230 u/L. There was no laboratory evidence of anemia, electrolyte abnormality, thyroid dysfunction or troponin elevation. Electrocardiogram was consistent with complete heart block. Echocardiography was significant for preserved ejection fraction and normal structure. A cardiac MRI was performed demonstrating patchy, late gadolinium enhancement of the right and left ventricle concerning for infiltrative cardiomyopathy.

Discussion:

Cardiac Sarcoidosis (CS) is detected clinically in 5% of cases of systemic sarcoidosis; however, it is thought to be more common given its prevalence of approximately 25% observed retrospectively through autopsy, and even higher in selected populations, such as 58% in Japanese patients. CS most commonly manifests as atrioventricular block or arrhythmias, in addition to heart failure or sudden death. Diagnosis can be difficult owing to sarcoidosis varied manifestations, and as a result clinicians should have a high degree of suspicion in patients with sequelae of sarcoid who present with symptoms and imaging suggestive of cardiac involvement. Our patient, with a known history of sarcoidosis, presented with an EKG significant for 3rd degree AV Block and evidence of infiltrative cardiomyopathy on Cardiac MRI. Our patient has established extra-cardiac sarcoidosis and a cardiac MRI with late gadolinium enhancement, consistent with the diagnosis of CS in accordance with the 2014 HRS Consensus Statement guidelines. Our patient underwent placement of a Biventricular Pacemaker and ICD, and was treated with corticosteroids for chronic immunosuppression. Our patient reminds us of the importance of maintaining awareness of the manifestations of cardiac sarcoidosis, and moreover, that all patients with extracardiac sarcoidosis should be screened for both subclinical and clinical evidence of cardiac sarcoidosis.
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Lyme Pyo-myositis

Lyme disease, caused by Borrelia burgdorferi, has become the most common vector born illness in the United States and is a prominent public health concern in New York State. Since its discovery in the 1970s, the clinical presentation of Lyme disease has diversified. B. burgdorferi has infrequently been reported to be the etiologic agent of myositis, progression to pyo-myositis is not well established. In this case we present a patient with a typical presentation of Lyme arthritis with progression to pyo-myositis.

40-year-old male patient with no significant past medical history presented to an emergency department with a one-week history of pain and swelling in the right knee along with limited range of motion. Two days prior to presentation the patient also developed swelling, erythema and pain in the right calf. The pain was mild and radiated down to his Achilles. No history of venous thrombosis and no systemic symptoms were reported. He was hemodynamically stable and afebrile. Physical examination revealed swollen, erythematous right knee and erythema over right calf.

Bedside ultrasound revealed a right knee effusion and right calf abscess. Formal imaging with CT of the lower extremity showed a 5.9 x 2.7 x 2 cm well-circumscribed collection suggestive of an abscess within the soleus, with adjacent soft tissue thickening. ESR/CRP were elevated at 5.47 and 102.9 respectively. Other labs unremarkable. Patient was empirically started on Vancomycin and Zosyn. The abscess was drained and ultrasound guided arthrocentesis was performed.

Synovial fluid analysis showed a neutrophil predominant septic joint. Gram stain was negative. Culture and gram stain results of the wound culture were also negative. Further history revealed the patient was from a Lyme endemic area with exposure. Lyme serologies were ordered and IgM/IgG were positive. PCR of the synovial fluid returned positive for Borrelia. The patient was treated with 6 weeks IV Vancomycin and Ceftriaxone for lyme arthritis and other bacterial causes of pyo-myositis, as all cultures remained negative. On follow up, patient had clinical resolution of disease and antibiotics were discontinued.

Infectious disease consulted on the patient and a diagnosis of late lyme disease with monoarticular arthritis was established. This is consistent with serological data showing IgM and IgG positivity. Our specialists believed that the lyme arthritis had either precipitated a local inflammatory response predisposing the patient to a pyo-myositis, or B. burgdorferi directly seeded the posterior calf compartment resulting in an abscess. The temporal relation between the development of the arthritis and subsequent development of the abscess days later is consistent with this reasoning. The patient had no other predisposing factors to the development of pyo-myositis and we believe the development of monoarticular arthritis secondary to lyme was either directly or indirectly causative of this patients pyo-myositis.
Resident/Fellow Clinical Vignette

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MASSIVE CK ELEVATION DUE TO SEVERE SEPSIS FROM SHIGELLA ENTEROCOLITIS

Rhabdomyolysis occurs due to a myriad of etiologies including trauma, drugs/toxins, prolonged immobilization, hereditary muscle enzyme defects, metabolic abnormalities and infections. If the diagnosis is delayed, rapid decompensation follows with secondary complications including arrhythmias and acute renal failure. Massive elevation of serum creatinine kinase (CK) is usually seen in acute alcohol intoxication, cocaine use and glucocorticoids. We present a case of massive rhabdomyolysis due to severe sepsis from Shigella Enterocolitis in an otherwise healthy young male. The 24-year-old presented to the ED with acute onset of bloody diarrhea, non-bilious vomiting and abdominal pain after eating at a Chinese buffet. His vitals were notable for tachycardia to 140 and he appeared weak and dehydrated with a diffusely tender abdomen. Initial laboratory assessment showed marked bandemia of 39%, lactate 3.2 mmol/L, creatinine 5.75 mg/dL, BUN 48 mg/dL, AST 2627 U/L, ALT 328 U/L and CK >100,000 U/L, above the lab reference capacity of 100,000 U/L. He had myoglobinuria and a CT abdomen consistent with colitis. Given the degree of CK elevation and renal injury, he was admitted and started on intravenous zosyn and aggressive intravenous fluid rehydration with 500cc/hr maintenance after a 4-liter bolus. Despite these measures he developed anuria; creatinine peaked at 12.92 mg/dL and CK remained unchanged. Hemodialysis (HD) was started on day 3 and his blood cultures returned positive for Shigella Flexneri on day 5 for which antibiotics were switched to intravenous ceftriaxone. Despite continued dialysis and aggressive fluid rehydration, his CK remained above measuring capacity for 4 days. On day 5, levels gradually fell to 86k U/L with eventual normalization to 111U/L by 5 weeks. Dialysis was discontinued and he started to make some urine at the time of discharge.

Rhabdomyolysis complicated by anuric renal failure in the setting of acute gastroenteritis due to Shigella infection is often self-limiting within 3-10 days. However, the degree of severe CK elevation seen here seemed unusual for rhabdomyolysis due to sepsis from Shigella Flexneri alone in an immunocompetent adult. Further work up for concern for an underlying myopathy included thyroid function test, antinuclear antibodies, anti Jo-1 titer, and anti-mitochondrial titers. Results were negative. Electromyogram obtained upon resolution of acute process four months after discharge showed no electrodagnostic evidence of a generalized myopathy. The systemic complication of severe rhabdomyolysis namely mild neurological complications to more severe renal failure require prompt recognition and identification of underlying causes for targeted treatment. We conclude that Shigella Enterocolitis can cause massive elevations in CK resulting in severe rhabdomyolysis and acute renal dysfunction.
BULLOUS LUPUS AS THE INITIAL MANIFESTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS

Background: Bullous systemic lupus erythematosus (BSLE) is an autoimmune blistering disorder that occurs in patients with systemic lupus erythematosus (SLE). The clinical manifestations of BSLE result from the disruption of epidermal-dermal adhesion secondary to antibodies against type VII collagen, the major component of the anchoring fibrils in the cutaneous basement membrane.

Case Report: A 22-year-old-female with Von Willebrand disease (VWD) presented with three weeks of widespread blistering eruption on the trunk, face, and extremities after sun exposure at an outdoor concert. Physical exam was notable for fever (Temp 102.8 F), and purulent filled bullae over the face, trunk, and extremities with crusting lesions on the lips, soft palate and genitals. Initial laboratory test revealed normocytic anemia (9.1 mg/dL), thrombocytopenia (131k/uL), and acute kidney injury (BUN 45mg/dL, creatinine 2.7mg/dL). An Echocardiogram showed moderate pericardial effusion with no evidence of hemodynamic compromise. Broad spectrum antibiotics, pulse dose steroids and intravenous immunoglobulin (IVIG) were given. Subsequent laboratory results revealed ANA (1:1280 speckled pattern), positive anti-Sm, anti-RNP, anti-SSA/SSB, dsDNA, anti-Histone and low C3 and C4 levels. Despite high doses of steroids and IVIG new blistering lesions continued to appear. A skin biopsy showed subepidermal neutrophils infiltrate, direct immunofluorescence showed linear deposition of IgG and IgA at the dermal side with salt-split skin preparation consistent with BSLE. Dapsone was started after desensitization (sulfur allergy) with improvement of the skin lesions. Hospital course was complicated with Streptococcus pyogenes superimpose bacteremia, autoimmune hemolytic anemia and nephrotic syndrome concerning for lupus nephritis, which responded to mycophenolate mofetil and rituximab.

Discussion: BSLE is a rare manifestation of SLE, characterized clinically by a rapid, widespread fluid-filled bullous lesions predominantly on sun-exposed areas, and histologically by subepidermal bullae with neutrophilic infiltrate and immunoglobulin (IgG, IgA, IgM) and C3 deposition at the basement membrane. Although BSLE most commonly presents in patients with preceding SLE, occasionally it can be the initial manifestation of SLE as seen in our case. Immunofluorescence is of particular importance to distinguish BSLE from other blistering disorders like dermatitis herpetiformis and linear IgA dermatitis where IgA is the unique immunoglobulin present on the direct immunofluorescence and from bullous pemphigoid where the antibodies bind to the roof of split, in contrast of BSLE where they bind to the base of the split on the indirect immunofluorescence (salt-split). Dapsone is the treatment of choice, with rapid resolution of blisters. However immunosuppressive therapy may be required when systemic manifestations of SLE coexist. Even though BSLE has not been linked with SLE activity, an association with lupus nephritis has been reported with high mortality rates. This case highlights the importance of the prompt recognition of BSLE as one of the manifestations of SLE and the utility of the immunofluorescence in the BSLE diagnosis.
Resident/Fellow Clinical Vignette

**Muhammad Shabbir**

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**Paroxysmal Atrial Fibrillation presenting as Anterior wall STEMI in an Elderly Female with Zero CAC Score**

Estimated 3% of acute coronary syndromes (ACS) are caused by coronary embolism (CE). In patients with ACS without significant underlying coronary artery disease, embolic etiology should be suspected and sought after. Diagnosis of CE may be difficult in patients with paroxysmal atrial fibrillation, as the rhythm may be normal at presentation. Identification of CE subgroup of acute coronary syndrome patients is important because of increased risk for major adverse cardiac outcomes and importance of systemic anticoagulation in prevention of future embolic events.

A 77-year-old non-smoker female presented with anterior wall ST elevation myocardial infarction (STEMI) due to complete occlusion of the proximal left anterior descending artery (LAD). Left main coronary, circumflex and right coronary arteries were normal (0% stenosis reported). Mechanical thrombectomy was performed and was followed with the placement of two stents in the proximal LAD. Post-STEMI medical therapy was initiated, including aspirin, clopidogrel, beta-adrenergic antagonist, statin, and ACE inhibitor.

Few days prior to her presentation with STEMI, she was seen at the outpatient cardiology office for complaints of palpitations and progressive exertional dyspnea. According to the patient, the heart beat â€“ felt irregularâ€™ for several years. No history of hypertension, diabetes mellitus, dyslipidemia, atrial fibrillation, or cardio-cerebrovascular disease was reported. At the office visit, the 12-lead ECG revealed sinus rhythm with premature ventricular complexes. 24-hour Holter monitor was notable for sinus rhythm with premature atrial contractions (2.1% of beats) and PVCs (1.2% of beats). The patient underwent an ECG stress test where the cardiac rhythm remained sinus but there were non-diagnostic ST and T wave changes. Given the non-diagnostic stress test and the symptoms highly suggestive for angina, on the day prior to her presentation with STEMI, coronary CT angiogram was performed which revealed normal coronary arteries and ascending/transverse aorta with a coronary artery calcium (CAC) score of zero.

When compared to the cerebral and systemic circulation, the coronary arterial vasculature is relatively protected from emboli due to difference in caliber of the aorta and coronary arteries, the acute angle at which the coronaries originate from the aorta, and the fast flow across the coronary ostia. Consequently, CE is a rare cause of acute coronary syndrome overall (2.9%) and STEMI (4.3%).

Clinical presentation of acute myocardial infarction due to CE is indistinguishable from such due to the atherosclerotic plaque rupture and vessel thrombosis. Consequently, the initial ACS management is the same, regardless of the etiology. However, recognition of CE, in addition to specific medical management, is important because of increased risk of cardiac death as compared to the patients with atherosclerotic myocardial infarction. Patients with CE myocardial infarction suffer from significantly increased 5-year risk of adverse cardiac and cerebrovascular events, which may be as high as 27%.
More Than Just Skin Deep: Cutaneous Metastases as Primary Manifestation of a Mullerian Carcinoma

Adenocarcinomas of Mullerian origin are rare malignancies primarily arising in the uterine corpus with the vagina, cervix, fallopian tube and ovary as less common sites of proliferation. Cutaneous manifestations of a visceral malignancy are uncommon and reported in about 0.6-10.4% of patients. Here, we depict a unique case of a gummatous lesion as the first sign of a Mullerian carcinoma.

A 50 year old female with a past medical history of uterine fibroids and umbilical hernia repair presented with right breast pain and persistent umbilical leakage. In the week leading up to this admission, she noticed three painless oval-shaped ulcers under her right breast. Vitals were stable on admission with no leukocytosis on labs. CT of the abdomen and pelvis demonstrated diffuse skin thickening with extensive subcutaneous fat infiltration in the lower ventral and pelvic wall consistent with cellulitis, increased soft tissue densities throughout the deep subcutaneous tissue of the ventral wall concerning for abscesses, and increased small soft tissue nodules in the omentum and extraperitoneal fat. On day 2, she underwent drainage of the abdominal abscess and a punch biopsy of the skin lesion. Tumor markers of the specimen were positive for PAX8, ER, and HER2 which was consistent with a poorly differentiated carcinoma of Mullerian origin. On day 3, she was discharged home on oral antibiotics with planned outpatient follow-up. Surgery recommended further staging workup with no surgical intervention. Unfortunately, the patient failed to present to her outpatient oncology appointments and remains lost to follow up.

Solid tumor malignancies rarely present with skin manifestations with the exception of those of breast and lung origin. Adenocarcinomas of Mullerian origin, which encompass viscera derived from Mullerian ducts have no documented cases with skin involvement, unlike our case in which these lesions were the chief complaint. Cutaneous manifestations of internal carcinomas can resemble either inflammatory or at times mimic infectious processes such as gummatous lesions often seen with syphilis. Unexplained skin lesions though necessitate a high index of suspicion and warrant further evaluation with a biopsy. Management often consists of different modalities including cytoreductive surgery and multi-agent platinum based chemotherapy, however, treatment ultimately remains dependent on the primary tumor. We present this case to raise awareness of this rare, poor prognostic entity to help facilitate early identification and management of a potential underlying malignancy and to arrest further disease progression and avoid poor outcomes.
Mangaiyarkkarasi Sivakuamr
Lincoln Medical Center

A Near Fatal Outcome of Strongyloides Stercoralis Hyperinfection in an Immigrant

Strongyloidiasis is caused by Strongyloides Stercoralis which is a soil-helminth endemic in tropical and subtropical regions. Infection usually occurs through skin contact with soil that is contaminated with larvae. After entering the body and migrating to the small intestine they burrow and lay their eggs. The larvae may mature and re-infect the host by burrowing in the wall of the intestines or the anus through a process known as auto infection. Without treatment and eradication, infected patients are at risk of sustaining a lifelong carrier state with periods of asymptomatic strongyloidiasis.

Immunocompetent individuals usually are asymptomatic carriers or have mild symptoms whereas immunocompromised individuals can have severe symptoms including severe abdominal pain, chronic diarrhea, malabsorption and weight loss, co-infection with gram negative sepsis and even death. It is important for clinicians to have a low index of suspicion for the diagnosis to prevent significant morbidity and mortality.

We present a 45-year-old female from Honduras who immigrated to the United States 4 months ago (stayed in an Immigration camp for over a month) with no past medical history was referred to the emergency department from primary clinic for hypotension and tachycardia. The Patient endorsed a 1-month history of abdominal pain worse in epigastric region associated with bloating sensation, decreased oral intake and had about 20 lbs of weight loss over the same time period. On physical exam patient appeared cachectic, had diffuse abdominal tenderness worse in epigastric area. Digital rectal examination was negative. Labs showed mild leukocytosis wbc 11.9 x 10^3/µL, normocytic anemia H/H 10.3/32.8 g/dl, mild transaminitis AST 45 U/L, ALT 52 U/L, hypoalbuminemia of 1.7 g/dl, and hypoproteinemia 4.7 g/dL, and eosinophilia (12%). A CT of the abdomen and pelvis revealed marked wall thickening of the entire small and large bowel, along with severe hepatic steatosis. Tissue transglutaminase antibody test, HIV, fecal calprotectin was all negative. Random cortisol level was within normal limits (24.4). Labs was also significant for an elevated Transferrin saturation of 57%. Other stool workup was negative, until the Stool ova and parasite study returned positive for strongyloides stercoralis larvae and blastocystis hominis cysts.

The Patient was started on Ivermectin and Albendazole treatment, with resolution of abdominal pain, hypotension and diarrhea. Repeat stool culture on outpatient follow up showed no ova or parasite.

In acute infection, patients may exhibit a local skin rash, dyspnea and cough due to larvae migration into the lungs. During chronic autoinfection, patients may remain asymptomatic or may experience mild gastrointestinal, respiratory or dermatological symptoms with peripheral eosinophilia. In our patient, the severity of infection led to malnutrition and hemodynamic instability that failed to respond to usual resuscitation measures complicated by prolonged malabsorption and protein losing enteropathy leading to severe hypoalbuminemia.
A Phoenix from the Ashes: Survivor from CAPS

Introduction:

Catastrophic antiphospholipid syndrome (CAPS) is a rare and serious disorder, characterized by multi-organ involvement (three or more organ systems) within a short period of time in patients with underlying antiphospholipid syndrome (APS). The term “catastrophic” was first introduced in 1992 due to high mortality rate (50%). CAPS occurs in less than 1% of APS patients. To the best of our knowledge this is the first reported case of antiphospholipid syndrome nephropathy (APLN) as the initial presentation of CAPS.

Clinical Course:

Our case is a 35 year old male with newly diagnosed SLE and idiopathic thrombocytopenic purpura (ITP) who presented with nausea, vomiting and diffuse abdominal pain. He was found to be oliguric with significant acute kidney injury (AKI) requiring continuous veno-venous hemofiltration (CVVH). Renal biopsy showed thrombotic microangiopathic nephropathy without evidence of lupus nephritis. He also developed microangiopathic hemolytic anemia (MAHA) and thrombocytopenia. Thrombotic thrombocytopenic purpura (TTP) and heparin induced thrombocytopenia (HIT) were ruled out due to normal ADAMTS13 level and negative HIT antibody. Autoimmune workup revealed ANA positivity 1:320 homogenous pattern with elevated ds-DNA antibody (718 IU/ml), low C3, as well as positivity for anti-cardiolipin, B2-glycoprotein and lupus anticoagulant. He was on heparin drip for coagulopathy and found to have left intra-jugular thrombus but then developed acute blood loss anemia secondary to GI bleed. He then developed pulmonary edema and ARDS requiring intubation. His course was further complicated by cardiac arrest status post return of spontaneous circulation (ROSC). Echocardiogram revealed dilated cardiomyopathy with moderate to severe mitral regurgitation with valvulitis and inferior wall hypokinesia. He underwent CABG for two vessel disease and mitral valve replacement. CAPS was diagnosed based on positive antibodies with multiple system involvement.

He received rituximab, cyclophosphamide, mycophenolate, pulse-dose solumedrol, without improvement and was transitioned to plasmapheresis followed by IVIG. Following plasmapheresis, he improved and was extubated. But unfortunately he developed septic shock with multi-organ failure including cavitating pseudomonal pneumonia, ESBL bacteremia, and ARDS requiring ECMO. He was weaned of ECMO and was re-intubated due to difficult extubation, later required tracheostomy and PEG tube placement. Due to his severely immunocompromised state and given clinical improvement following plasmapheresis, the decision to avoid eculizumab was made. Despite the severity of CAPS with multiple complications, patient improved and was discharged to lower level hospital for continued care.

Discussion:

The diagnosis of CAPS can be challenging due to the requirement of auto-antibodies in the serum as well as presence of thrombosis in multiple organs. Early recognition of CAPS is crucial as the diagnosis carries a very high mortality. Although CAPS is a serious and life-threatening disease, timely diagnosis and aggressive management can prevent fulminant irreversible complications associated with its poor prognosis.
Rivaroxaban induced Thrombocytopenia

Introduction

Factor Xa inhibitors (Rivaroxaban and Apixaban) are preferred agents in a patient with acute venous thromboembolism for long-term anticoagulation who are not pregnant women, severe renal insufficiency and active cancer. The incidence of thrombocytopenia after initiating treatment with Rivaroxaban was very rare. Herein, we report a case of 39-year-old Hispanic female who developed severe thrombocytopenia after the treatment of Rivaroxaban.

Case presentation

39-year-old Hispanic female with no past medical history presented to the ED after she had a near syncope episode at home. In the ED, vitals were within normal range. Initial labs showed WBC 13.6 x 10^3/ul, hemoglobin 12.1 g/dl, hematocrit 37%, platelet count 111 x 10^3/ul; chemistry, electrolytes and liver function test were within normal range, Troponin 1.190. ABG on room air showed 7.44/31/78/21.1 with A-a gradient 33 mmHg.

Patient underwent CT pulmonary angiogram which revealed extensive bilateral acute pulmonary embolism. Patient was treated with therapeutic dose of Enoxaparin. Enoxaparin was bridged to warfarin. Patient was discharged home on oral warfarin with INR in therapeutic range. CBC checked one week after initiating warfarin showed platelet count 279 x 10^3/ul. Warfarin was switched to Rivaroxaban by the PCP. Patient underwent hypercoagulopathy study which was all negative. CBC done 6 weeks after taking Rivaroxaban revealed that platelet count was 35 x 10^3/ul.

Patient was again admitted to hospital due to thrombocytopenia. Rivaroxaban was stopped and patient was given with heparin infusion. Fibrinogen level was found to be 463 mg/dl which ruled out consumption coagulopathy. Heparin induced antibody was negative. Heparin infusion was bridged to warfarin. Platelet count improved gradually upon discontinuing Rivaroxaban. 4 days after discontinuing Rivaroxaban, platelet count increased to 84 x 10^3/ul from initial 34 x 10^3/ul. Patient was discharged on warfarin with INR in therapeutic range. Upon outpatient follow up, patient’s platelet count continued to improve to 134 x 10^3/ul, 230 x 10^3/ul, 202 x 161 x 10^3/ul and 161 x 10^3/ul on weekly CBC checked

Discussion

This patient had normal platelet count before Rivaroxaban treatment. After 6 weeks of treatment with Rivaroxaban, her platelet count decreased to critically low level, and platelet count recovered upon discontinuing Rivaroxaban. Other causes of thrombocytopenia, including consumption coagulopathy, HIT, were excluded. According to George criteria for assessing drug induced thrombocytopenia, the patient met level of evidence II9 for DITP. The incidence of drug induced immune thrombocytopenia (DITP) is about 10 persons per million annually. Six distinct pathogenic mechanism of DITP have been identified, some involved production of antibodies against platelet. The mechanism of Rivaroxaban induced thrombocytopenia is unknown. Thrombocytopenia during anticoagulation therapy may lead to life-threatening hemorrhage. Close monitor of hemoglobin level and platelet count are crucial for early detection of thrombocytopenia and occult hemorrhage to avoid severe hemorrhagic complications.
Pulmonary Embolism as the initial presentation of Systemic Lupus Erythematosus

Background

Systemic Lupus Erythematosus (SLE) is an autoimmune disease which has a well-known association with hypercoagulability. It is rare, however, for SLE patients to present with pulmonary embolism (PE) as their initial manifestation of the disease. We present a patient with 4 months of recurrent respiratory symptoms, who was diagnosed with SLE only after he presented with venous thromboembolism (VTE).

Case

A 27 year old gentleman with no significant medical history presented to the emergency department with leg swelling, recurrent pneumonia, lethargy, increased breathing difficulties and forty pound weight loss over a four month period. Review of systems was positive for facial rash, but there was no history of lymph node swelling or GI symptoms. He denied any history of chest pain, dizziness, vision change, or hematuria. On physical exam, the patient was saturating well on room air and the remainder of his vital signs were normal. He was found to have a malar rash. The respiratory examination was normal. The right lower limb was swollen and mildly tender. No lymphadenopathy was appreciated.

The investigation showed the patient had anemia, leukocytosis and thrombocytopenia. Urinalysis revealed 3+ blood, 4+ protein, and RBCs and WBCs great than 40. Ultrasound of the right lower limb demonstrated DVT, and CT chest revealed pulmonary emboli involving the lobar and segmental branches bilaterally. Because of the extensive lower extremity DVT, a CT venogram was done, which showed a filling defect in the inferior vena cava and right common iliac vein consistent with thrombus.

Wegener’s granulomatosis, Goodpasture syndrome, Microscopic polyangiitis, Churg-Strauss syndrome and DIC were ruled out. Anti-dsDNA and ANA were positive. His antiphospholipid (APL) antibodies were normal, as were other tests for hypercoagulability, thus he did not meet the criteria for Antiphospholipid Syndrome (APS). The patient was diagnosed as having SLE. Anticoagulation was initiated with heparin drip and transitioned to subcutaneous lovenox after four days. SLE was treated with mycophenolate, hydroxychloroquine, and oral steroids.

Discussion

The clinical presentation of SLE is varied and depends on the type of antibodies present [3]. Pulmonary embolism as the first presentation of SLE is rare. The patients with SLE are predisposed to vascular thrombosis. Presence of antiphospholipid antibodies further increases the risk of thromboembolic phenomena. Initial treatment of PE/DVT in a patient with SLE is not different from the routine treatment of PE/DVT. The duration of the treatment, however, is less clear. Most of the patients in the literature with SLE, even in the absence of APL antibodies, are considered to have similar risk for VTE as APS patients and the consensus for them is lifelong anticoagulation.
MASSIVE POSTERIOR STROKE PRESENTING WITH ISOLATED DIZZINESS

4% of annual emergency rooms visits are for dizziness. While most cases are due to benign conditions like vertigo, approximately 5% of these patients are found to have cerebrovascular events of the posterior circulation. About 20% of all cerebrovascular events involve the posterior distribution.

Diagnosis in these patients can be especially tricky as these patients can present with nonspecific symptoms such as isolated dizziness, diplopia, nystagmus, bilateral leg weakness, or numbness. Furthermore, these patients can present with low or normal NIHSS stroke scores. Additionally, radiological imaging poorly evaluates the posterior fossa. Noncontrast CT head has low sensitivity (26%) for posterior infarctions. These factors culminate in up to 1/3 of diagnoses being missed in the ED.

Case Presentation

A 71-year-old Caucasian female presented to the ED with dizziness and nausea. NIHSS score in the ED was 0; the initial CT head was negative, and serial neurological exams showed no focal neurological deficits. Despite the initial head CT is negative, clinical suspicion for posterior stroke remained high on the differential, and stroke workup was initiated.

MRI head was performed, which revealed a large area of acute infarct involving the right temporal, parietal, occipital lobes, in the vascular distribution of the right posterior cerebral artery territory. Neurological exams were repeated with emphasis on posterior cerebral and cerebellar function, including visual field testing. The patient was found to have left-sided hemianopsia, but otherwise, the exam remained normal.

The patient was treated with guideline-driven CVA therapy, including Statin and evaluated by PM&R without a significant disability. The patient was discharged home with minimal residual deficits with outpatient rehabilitation and neurology followup.

Discussion

Certain stroke presentations have earned the term Stroke chameleons, which is a stroke that presents as another condition when it is a stroke. In the management of stroke, prompt recognition is vital as thrombolytic therapy is time-dependent. Missed stroke or delayed recognition of these stroke chameleons can lead ineligibility for thrombolytic treatment. A composite three portion clinical exam known as HINTS (Head Impulse / Nystagmus / Test of Skew) has been shown to 98% sensitive for detecting posterior stroke syndrome. This clinical exam has higher sensitivity then MRI (83% sensitivity). The HINTS exam evaluates the vestibulocochlear reflexes to differentiate between benign peripheral vertigo and central vertigo (CVA).

Widespread education and implementation of the HINTS exam can provide a highly sensitive rapid, bedside clinical tool for early screening and detection of posterior cerebrovascular events. Our case demonstrates the necessity of implementing early detection screening tools such as the HINTS exam considering the tendency of posterior strokes to present as Stroke Chameleons or with nonspecific symptoms and the propensity of current screening methods to miss cerebrovascular events involving the posterior circulation.
Background: Anti NMDA receptor encephalitis (anti NMDA-R encephalitis) is a relatively novel discovery made in 2007. There is a significant dearth of understanding about this condition and its variable presentation which often leads to a delay in diagnosis. We present an interesting case of anti NMDA-R encephalitis which, despite ominous clinical deterioration, resulted in rapid recovery within 12 hours of treatment once diagnosed accurately.

Case: A 25 year old female with no medical history presented with headaches, neck stiffness, fever and photophobia. General and neurological examination was initially non-localizing. CT head was normal. Lumbar puncture revealed lymphocytic pleocytosis, but gram stain, bacterial cultures, encephalitis panel and multiple sclerosis work up were negative. MRI revealed hyperintense T2-flair signal possibly suggesting encephalitis. Clinically, she deteriorated over the next couple of days with worsening in mental status, motor function, memory and speech. Antibiotics and antivirals were started but she remained febrile. Serum NMDA antibodies were negative. LP was repeated to check for fungal etiologies and NMDA antibodies and was unrevealing. Vasculitis work up was negative, as were chest and abdominal CT scans. EEG showed moderate encephalopathy without epileptiform abnormalities. LP was repeated again to check for paraneoplastic and autoimmune etiologies including NMDA antibodies which resulted to be positive. She was treated with intravenous immunoglobulin and methylprednisone for 5 days. Within 12 hours, she experienced substantial improvement in cognitive function with almost complete recovery of motor function and speech. A transvaginal ultrasound as well as CT abdomen and pelvis (done to rule out an ovarian teratoma) was negative.

Conclusion: The incidence of NMDA encephalitis has not been prospectively measured, however a multicenter population based study suggests that it accounts for less than 3-4% of all causes of encephalitis. It has many presentations varying depending on age and gender. Majority of cases are considered to be paraneoplastic, most often associated with discovery of ovarian and lung tumors. Idiopathic cases represent about one third of the reported cases so far. Though the pathophysiology is not entirely understood, antibodies to the NR1/NR2 subunits of NMDA receptors may lead to decreased NMDA-R mediated synaptic currents and hence neuronal death. Furthermore, the diagnosis should not be ruled out based on unremarkable serum findings and hence multiple CSF examinations may be needed in order to confirm positive NMDA-R antibodies. First line therapies include removal of tumors (if present), steroids, IVIG and plasma exchange. Lastly, though it has been well documented that treatment response may be slow and drawn out, recognition of clinical features and accurate diagnosis can lead to a dramatic response to medical treatment.
Resident/Fellow Clinical Vignette

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Disseminated CMV causing Hemophagocytic lymphohistiocytosis

Introduction: Hemophagocytic lymphohistiocytosis (HLH) is an aggressive and life-threatening syndrome which occurs due to excess activation of the immune system with an incidence of 1.2 cases per million patients per year. Primary HLH is due to a genetic defect and while secondary HLH occurs in immunocompromised states such as infections, malignancy, organ transplantation or rheumatological disorders. It has a multiorgan involvement and presents with fever, rash, lymphadenopathy, hepatosplenomegaly, cytopenia, elevated ferritin and deranged liver function tests. Untreated patients have survival of few months and the mortality rate is almost 47%. Since the presentation is often mistaken for sepsis, diagnosis is delayed. We present a case of CMV-associated HLH in an immunocompromised female, which has been rarely described in the literature.

Case: A 46 year old female with a history of Crohn’s disease and psoriatic arthritis presented with complaints of fever and chills for the past nine days. Her medications included 6MP, sulfasalazine, and plaquenil. She was febrile and tachycardic on presentation. Pertinent laboratory findings were white count 2,500/uL, hemoglobin 12.3 gm/dl, platelets 76,000/uL, total bilirubin 2.1 mg/dL, ALT 395 U/L, AST 251 U/L, ALP 408 U/L, ferritin 1552 ng/ml. Serum electrolytes and creatinine were unremarkable. Computed tomography (CT) scan of the thorax showed bibasilar atelectasis while CT abdomen showed splenomegaly. She was tested positive for CMV IgM with elevated CMV PCR (693,000 IU/mL). She was started on broad-spectrum antibiotics and Ganciclovir. Remainder of the workup for bacterial, viral and fungal infections was negative.

The patient had persistent fever over the next 4 days and her pancytopenia was worsening. Ferritin was noted to be rising at 3296 ng/ml with TG of 252 mg/dl. Since the suspicion for HLH was high, bone marrow biopsy was done which showed florid hemophagocytosis. Subsequent workup for HLH showed markedly elevated CD25 activity, reduced fibrinogen of 150 mg/dl and reduced Natural Killer cell activity. Treatment for HLH as per the HLH-94 protocol with eight weeks of induction therapy with Etoposide and Dexamethasone was initiated. She has marked improvement in her pancytopenia, liver functions and ferritin one week after initiation of treatment. The patient is currently doing well.

Discussion: An episode of HLH is usually triggered by an infection or alteration in the immune system which causes excessive cytokine release by macrophages leading to tissue destruction. There is impaired cytotoxic function of Natural killer (NK) cells and cytotoxic T lymphocytes which are unable to eliminate the activated macrophages. As per the HLH-2004 trial, the patient met eight out of eight criteria required for diagnosis of HLH. Disseminated CMV associated with HLH is rare in adults and physicians must have high suspicion for HLH in immunocompromised patients with multiorgan involvement for early diagnosis and to reduce mortality.
A rare case of selective IgM deficiency associated with Hashimoto’s thyroiditis

Introduction: Selective IgM deficiency deficiency (sIgMD) is an uncommon immunodeficiency disorder, with fewer than 300 cases reported in literature. Patients commonly present as repeated infections with extracellular and intracellular bacteria, viruses, and fungi. In rare cases, it can be associated with autoimmune, malignant or allergic conditions. We present a case of sIgMD with Hashimoto’s thyroiditis in an otherwise asymptomatic young male and its management.

Case: A 39 year-old-male with history of Hashimoto's thyroiditis and hypothyroidism was referred by his primary care physician after his IgM levels were found to be consistently low for over a year. The patient reported of few episodes of bronchitis every winter but was completely asymptomatic on presentation. He did not recall any recurrent sinusitis, otitis, pneumonia, meningitis during childhood. He was an active smoker and smoked a pack-per-day for the past 20 years. His immunoglobulin assay showed low IgM levels (22 mg/dl) while IgA was normal (151 mg/dl) and normal IgG (1095 mg/dl). His immunoglobulin levels one year ago also showed similar pattern. His hemoglobin, white blood count, platelets, creatinine and serum electrolytes were unremarkable. Serum kappa/lambda free light chains along with serum and urine electrophoresis showed normal findings. Serum celiac panel and autoimmune workup was negative. The patient was educated to seek urgent medical attention in case of fever or other infectious symptoms. The patient is currently doing well and is regularly being followed up in clinic.

Discussion: Recurrent infections as the presenting manifestation occurs in more than 80% of patients with sIgMD, however certain patients can remain asymptomatic. Some of these bacterial infections can be life-threatening. There is an increased frequency of allergic and autoimmune manifestations in sIgMD. The role of IgM deficiency in the development of autoimmune diseases remains unclear but it could possibly be due to the deficiency of natural antibodies/autoantibodies, which could predispose to pathologic autoantibody formation. Hence patients with sIgMD are at risk for systemic lupus erythematosus, Hashimoto's thyroiditis, autoimmune glomerulonephritis, and rheumatoid arthritis. Several malignancies and hematologic diseases that have been reported in association with IgM deficiency include multiple myeloma and paraproteinemias, clear cell sarcoma, Bloom syndrome, lymphomatoid papulosis, immune thrombocytopenia (ITP), and lymphocytic leukemias.

IgM deficiency causes impairment of IgG antibody responses against pneumococcal and meningococcal polysaccharides, hence vaccines must be administered to prevent these infections. Patients with recurrent infections need to be treated with prophylactic antibiotics. Additionally, intravenous immune globulin has an excellent clinical response in case of recurrent infections. Through this case we highlight that asymptomatic patients with sIgMD should be monitored routinely for change in clinical status. They should also be tested for hematological and autoimmune disorders.
Hemophagocytic Lymphohistiocytosis: A Delayed Diagnosis due to the Absence of Bone Marrow Hemophagocytosis on Initial Bone Marrow Assessment

Background: Hemophagocytic Lymphohistiocytosis (HLH) is a rare and life-threatening disease due to excessive immune activation and is characterized by clinical and laboratory evidence of extreme inflammation. We present a case of HLH that was initially undiagnosed due to the absence of hemophagocytosis on initial bone marrow assessment.

Case: A 77-year-old male presented with nonproductive cough and a fever of 38.6°C. Laboratory results showed pancytopenia with hemoglobin of 11.6g/dL, leukocyte count of 1,300/μL (absolute neutrophil count 900), platelet count of 95,000/μL and splenomegaly on abdominal ultrasound. The patient was started on empiric broad-spectrum antibiotics but remained febrile. Infectious workup was negative. Laboratory results revealed ferritin of 2200ng/mL, fibrinogen of 142 mg/dL and triglycerides of 149mg/dL. HLH was suspected. However, bone marrow biopsy was nonrevealing. On day seven of hospitalization, the patient spiked a fever of 39.4°C and became hypotensive requiring vaspressors. He developed progressively worsening anemia (hemoglobin 6.7g/dL) and thrombocytopenia (platelet count 44,000/μL). Repeat bone marrow biopsy revealed a hypocellular marrow and extensive hemophagocytosis. Subsequent results showed ferritin of 8200ng/mL, fibrinogen of 63mg/dL and hypertriglyceridemia of 280mg/dL. A diagnosis of HLH was made, and treatment was initiated with etoposide and dexamethasone. The patient died two days later due to uncontrolled gastrointestinal hemorrhage.

Discussion: HLH is a challenging diagnosis due to the variable clinical presentation. A potential misconception is that hemophagocytosis is pathognomonic for HLH. The sensitivity and specificity for hemophagocytosis based on a small study is 76.7% and 77.8% respectively. Hemophagocytosis may be absent in the initial bone marrow biopsy at disease onset (or possibly missed by sampling) and may appear later on in the disease course, as was seen in our patient. Based on the diagnostic guidelines from HLH-2004 trial, five out of nine criteria is needed to diagnose HLH: Fever ≥38.5°C, splenomegaly, cytopenia (2 of following: anemia, thrombocytopenia, ANC <1000/microL), ferritin >500 ng/mL, fasting triglycerides >265 mg/dL or hypofibrinogenemia <150mg/dL, hemophagocytosis in bone marrow, low NK cell activity and elevated sCD25. In our patient, the initial bone marrow biopsy was negative for hemophagocytosis. However, he met the first five criteria required for a diagnosis of HLH. Ideally, treatment should have been initiated even before the repeat bone marrow biopsy. We should keep in mind that a very high ferritin level especially levels >10,000 g/dL is highly sensitive and specific. In addition, sCD25 is a very useful inflammatory marker and correlates very well with current disease activity. Clinicians should consider HLH in very ill patients with concurrent cytopenias and elevated inflammatory markers especially ferritin. Our case serves as a reminder that clinicians should maintain a high degree of suspicion even if the bone marrow biopsy does not reveal hemophagocytosis, and treatment should not be delayed looking for this single feature.
A Sweet Diagnosis: Sweet Syndrome Presenting As Neutropenic Fever in a Patient with AML

Introduction: Sweet syndrome (acute febrile neutrophilic dermatosis) is a rare, inflammatory disorder characterized by the abrupt appearance of erythematous papules, plaques or nodules on the skin. As the name of the disease suggests, fever and neutrophilia frequently accompany the skin lesions. However, Sweet syndrome can occur in the absence of neutrophilia. We report a case of Sweet syndrome in a patient with neutropenic fever.

Case: A 68-year-old female with a history of treatment-refractory acute myelogenous leukemia (AML) presented with fever, and a small black, tender nodule under her left breast with surrounding induration. She was chronically neutropenic (ANC 0-200), and receiving prophylactic acyclovir and fluconazole. She improved after seven days of broad-spectrum parenteral antibiotics and was discharged home with three days of oral doxycycline. Ten days later, she presented with a fever of 38.3°C and ulceration of the left breast skin nodule. Ecthyma gangrenosum was suspected, and vancomycin and cefepime were started. Routine, fungal and mycobacterial blood cultures were negative. Despite eleven days of antibiotics, the left breast lesion did not improve, and she developed a second lesion in the right inguinal fold. Skin biopsy of the left breast lesion revealed diffuse neutrophilic infiltration of dermis and absence of vasculitis. Stains for bacteria, mycobacteria, and fungal organisms were negative. A diagnosis of Sweet syndrome was made, and antibiotics were discontinued. Systemic corticosteroids were not initiated due to concern for increased infection risk with ongoing neutropenia, and further treatment of her AML was not advised. She remained afebrile, and ultimately chose to return home.

Discussion: Sweet syndrome is classified into three subtypes: classical/idiopathic, malignancy-associated, and drug-induced Sweet syndrome. According to diagnostic guidelines proposed by Von den Driesch, two major, and two or more minor criteria should be met to diagnose Sweet syndrome. Major criteria include sudden onset of painful erythematous nodules, and histopathological evidence of dense neutrophilic infiltrate without evidence of leukocytoclastic vasculitis. Minor criteria include pyrexia greater than 38°C, association with underlying hematologic malignancy or predisposing conditions, excellent response to systemic glucocorticoids, and abnormal laboratory values (ESR >20 mm/hour; CRP positive; WBC >8,000; >70% neutrophils). Our patient fulfilled both major criteria and the first two minor criteria. A review of 118 patients with Sweet syndrome demonstrated that 21% had a hematologic or solid tumor malignancy, and the most common malignancy associated with Sweet syndrome is AML. Sweet syndrome can occur in the absence of neutrophilia, as was seen in our patient with neutropenia. In patients with hematologic malignancies presenting with fever and erythematous skin nodules, Sweet syndrome should be considered as an alternative cause of fever other than infection. Unlike systemic infection, effective treatment of Sweet Syndrome involves treatment of the underlying malignancy and/or corticosteroids, and not broad-spectrum antibiotics.
Secondary Hemophagocytic Lymphohistiocytosis following Influenza Vaccination

Introduction:

Hemophagocytic Lymphohistiocytosis (HLH) is a life-threatening syndrome of immune hyperactivation, characterized by dysregulation of lymphocytes and histiocytes. It occurs as a familial or sporadic disorder and can be triggered by infections, hematologic malignancies, and autoimmune conditions. Given its variable presentation and association with other pathologies, HLH is difficult to diagnose. Interestingly, there have been case reports of HLH associated with vaccinations (dTaP, measles, and MMRV). Here we present a case of HLH following an influenza vaccination.

Case Presentation:

A 76 M with hypertension, hypothyroidism, and hyperlipidemia presented to the hospital with one week of intermittent fevers, generalized malaise, and was incidentally found to be anemic. Of note, the fevers started one day after receiving influenza vaccine. He was admitted for presumed sepsis and empirically treated with vancomycin and piperacillin-tazobactam. His clinical course deteriorated rapidly within the first two days of hospitalization. The patient continued to spike fevers despite empiric broad spectrum antibiotics and required transfusion on day three of hospitalization. On day five, he developed new thrombocytopenia. Antibiotics were expanded to vancomycin, micafungin, doxycycline, and meropenem given the thrombocytopenia and the fevers. Ferritin continued to rise from initial value of 400 ng/ml to 11742 ng/ml over the course of the first week. Triglyceride were elevated at 465 mg/dl. Bone marrow biopsy showed hemophagocytosis, raising the suspicion for HLH. He was immediately treated with dexamethasone, followed by etoposide. He defervesced after the first dose of etoposide. Subsequent tests showed soluble IL-2 receptor were was also elevated at 62900 u/ml.

Extensive infectious workup including Epstein Barr virus, Cytomegalovirus, West Nile virus, HIV, hepatitis panel, Herpes virus, tick-borne diseases, tuberculosis were negative. Tests for autoimmune disorders such as systemic lupus erythematosus, rheumatoid arthritis, Sjogren’s syndrome, collagen vascular diseases returned negative. Primary HLH was less likely given his age. Based on the clinical data and absence of other identifiable triggers, HLH appears to have been triggered by influenza vaccination. Hospital course was complicated by neutropenic sepsis secondary to fungemia and polymicrobial bacteremia, respiratory failure, and acute renal failure. The patient responded to the etoposide regimen and was discharged to a rehabilitation facility. Patient is clinically well during outpatient follow-up.

Discussion:

Patients that present with HLH tend to be critically ill and deteriorate rapidly without treatment. Therefore, HLH should be included in the differential diagnosis of patients presenting with fevers, pancytopenias, and elevated ferritin. To the best of our knowledge, this is the first case report of HLH associated with influenza vaccine in an immunocompetent patient. The vaccine may have disrupted the immune homeostasis in a susceptible host triggering overwhelming macrophage activation. Although the prognosis of HLH is generally poor, early detection and treatment of HLH can show drastic improvements in morbidity and mortality.
Resident/Fellow Clinical Vignette

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Nivolumab Induced Combined ACTH and GH Deficiency: A rare, life-threatening but easily correctable adverse event

Background:
Nivolumab is a monoclonal antibody, which binds to a human programmed cell death protein-1 (PD-1) on the T-cells, and helps to restore T-cell mediated anti-tumor activity. There has been evolving observational data reporting autoimmune adverse events related to these immunotherapy drugs. In this case report, we present a patient who developed combined adrenocorticotropic hormone (ACTH) and growth hormone (GH) deficiency likely secondary to autoimmune (AI) Hypophysitis as an adverse event of Nivolumab therapy, which she had received for her malignant melanoma.

Case:
A 46-year-old female presented to the outpatient clinic in November 2018 with decreased energy, fatigue, myalgia, shortness of breath on exertion, recent weight loss of 60 lbs. progressively worsening over 3 months. She had a history of stage IIIIC malignant melanoma (BRAF Wild type) on her right face, for which she underwent excision of the lesion and started on immune checkpoint inhibitor (ICI)- Nivolumab [IV 240 mg q2 weeks] in March 2018. After 10 cycles of therapy, she developed optic neuritis which was presumed to be immune-mediated and immunotherapy treatment was stopped. After 2 months she was admitted to the hospital with a diagnosis of DKA, which after extensive workup was presumed to be immune-mediated secondary to Nivolumab. She was started on an insulin regimen for this diagnosis. She denied any skin changes, dizziness, hair loss, menstrual changes and leg swelling. Physical examination was benign except for orthostatic hypotension. Her laboratory analyses showed Cortisol <1.0 [3.0-16.0 μg/ml], ACTH <5.0 [7.2-63 pg/ml], IGF-1 37 [49-240 ng/ml], FSH 6 [3-33 mIU/ml], LH 4 [<17 mIU/ml], Prolactin 8 [<30 ng/ml], TSH 0.76 [0.34-3.00 mIU/ml], free T4 1.1 [0.6-1.6 ng/dl]. High dose ACTH (0.25 mg IV Cosyntropin) stimulation test revealed subnormal response with baseline cortisol level <1.0, 30-minutes cortisol level of 1.2 and 60-minutes cortisol level of 1.6, indicating primary/secondary adrenal insufficiency. Based on undetectable ACTH, secondary adrenal insufficiency and GH deficiency was diagnosed. She was treated with oral prednisone with remarkable improvement in her symptoms within a few days.

Conclusion:
AI Hypophysitis secondary to immunotherapy is an established entity. The objective of our report is to highlight a rare but life-threatening entity called combined ACTH and GH deficiency secondary to Nivolumab induced AI Hypophysitis, where only ACTH pathway is affected amongst all the available endocrine pathways in the anterior pituitary. Oncologists, endocrinologists, primary care physicians need to be familiar with this potentially life-threatening and easily correctable adverse event.
Successful treatment of cardiac electrical storm with cardiogenic shock using ECMO and Impella

Electrical storm is a life-threatening syndrome, defined by three or more sustained episodes of Ventricular Tachycardia (VT) and/or Ventricular Fibrillation (VF) leading to implantable cardioverter-defibrillator (ICD) shocks occurring over a 24-hour period. We present a rare case of a young woman without any history of structural heart disease, myocardial infarction or inherited arrhythmic syndrome with a dramatic clinical presentation of electrical storm refractory to standard treatment and was managed successfully.

A 30-year-old woman with a past medical history of alcohol abuse collapsed at her home. Initial rhythm on EMS arrival was noted to be ventricular fibrillation. CPR was initiated, the patient was shocked, intubated and transported to the hospital. Soon upon arrival to the hospital, she was noted to develop VF again, was cardioverted successfully and amiodarone drip was started. In the interim, she developed hypotension and needed vasopressors. Of note, the patient’s family reported that in the past month patient had multiple emergency department visits for dyspnea and decreased exercise tolerance along with upper respiratory tract symptoms. She was also exposed to a sick child a few days prior to the presentation. The initial lab work revealed cardiac troponin I level of 2.7 ng/ml and no electrolyte abnormalities. Baseline EKG showed sinus tachycardia, premature ventricular contractions and QTc interval of 420 milliseconds. The patient continued to have multiple runs of VT and VF requiring electrical shocks and further addition of lidocaine drip. An emergent bedside echocardiogram showed ejection fraction (EF) of 6-10% with global cardiomyopathy. Emergent coronary angiogram was performed which showed normal coronaries and severely depressed EF. While in cardiac catheterization laboratory patient again had multiple runs of VT/VF requiring 15 shocks. She received mechanical hemodynamic support with Impella 2.5 (percutaneously inserted ventricular assist device) and Veno-arterial (VA) ECMO. In addition, the patient was started on high dose steroids for suspected myocarditis. The patient’s electrical storm terminated shortly after receiving mechanical cardiac support. Although cardiac biopsy could not be obtained to confirm the diagnosis of myocarditis, a cardiac MRI performed after clinical stabilization showed sub-epicardial late gadolinium enhancement involving the anterolateral wall of the LV midlevel segment and inferolateral wall of the LV midlevel segment suggestive of myocarditis. Thus, the patient likely had fulminant myocarditis which leads to the electrical storm and hemodynamic instability resulting in cardiogenic shock.

This case highlights the importance of timely identification and management of electrical storm. Myocarditis rarely presents with an electrical storm. Beta-blocker plays a key role in managing the activated sympathetic system, but it could not be utilized because of cardiogenic shock complicating the course. The timely utilization of a combination of ECMO with Impella saved the life of this young patient.
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**Recurrent Pneumothorax in a Young Woman with Catamenial Pneumothorax**

Catamenial pneumothorax (CP) is a rare clinical entity presenting with recurrent pneumothorax in women of reproductive age. Presence of ectopic endometrial tissue in thorax leads to its activation with each menstrual cycle. We report a unique case of a young female who presented with recurrent spontaneous pneumothorax despite video assisted thoracoscopic surgery (VATS).

A 36-year-old female was sent to emergency department for management of a large right sided pneumothorax on a CT chest done as outpatient for evaluation of chest discomfort. She had a past medical history of endometriosis, DVT, catamenial pneumothorax after having VATS procedure about 4 months prior to this presentation. She was hemodynamically stable with SpO2 97% on room air. CT chest revealed a very large loculated right pneumothorax with left mediastinal shift and extensive pleural thickening. Initially she underwent a pigtail catheter placement in the largest pocket. Despite the pigtail drainage, patient continued to have air leak and mediastinal shift. Thus, a decision was made to perform VATS again. Pleural biopsy and VATS pleurodesis were performed with placement of 12F, 19F, 28F chest tubes. Mediastinal shift resolved but she continued to have small pneumothorax. On day 16, she was discharged with a chest tube for outpatient follow-up. Chest tube was removed after one week follow up in the clinic. She continued to have small loculated right sided pneumothorax. Pleural biopsy obtained during VATS, showed positive paired box gene 8 (PAX8) in epithelial lining compatible with presence of endometrial tissue in pleura. She is currently on progesterone only mini pill and is closely followed up in the clinic.

Catamenial pneumothorax (CP) is an uncommon cause of recurrent pneumothorax. Histologic finding of endometrial tissue on pleural biopsy is often required to confirm the diagnosis. CP is usually managed with hormonal therapy and VATS but needs further investigation. Even after a confirmed diagnosis, CP poses a treatment challenge as it can continue to cause recurrent large pneumothoraces despite VATS and hormonal therapy. Patients can develop pleural thickening and loculations which makes management of future pneumothoraces even more difficult. No clear guidelines exist on management of CP. With the disease affecting young women with significant impact on quality of life, it is prudent that more research should be done for CP management.
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Purulent Pericarditis Caused by Bacteroides Fragilis: A Rare Complication of Cholangitis

Introduction:
Pericarditis involves inflammation of the pericardium and viral infections are typically the most common cause. However, about 1% of cases are due to a bacterial infection and carry a poor prognosis if untreated. The bacteria identified most commonly are Staphylococcus aureus, Streptococcus pneumoniae, Haemophilus influenza, and Neisseria meningitidis. However, infections caused by anaerobic bacteria have also been reported. The etiology of the infection may be hematogenous or extension from a contiguous location. Historically, pneumonia was cited to be the most common source, but this has significantly decreased in the post-antibiotic era.

Case Presentation:
An 89-year-old female with a history of cholangitis four months prior to arrival presented with anorexia and confusion for three days. On physical exam the patient was toxic-appearing with jugular vein distention and distant heart sounds on auscultation. The patient’s clinical condition rapidly declined and she developed septic shock. She was intubated and placed on mechanical ventilation. She was started on vasopressor medications and broad-spectrum intravenous antibiotics. An emergent bedside transthoracic echocardiogram showed a large pericardial effusion with evidence of right ventricular collapse. Emergent pericardiocentesis was performed with drainage of 700 mL of yellowish fluid. Subsequent cultures of the pericardial fluid isolated Bacteroides fragilis. Antibiotics were adjusted to ceftriaxone and metronidazole based on the culture sensitivities. After 48 hours of admission, a computed tomography scan of the abdomen and pelvis with contrast revealed a liver abscess with a fistulous tract reaching the pericardium. The patient declined drainage of the liver abscess. She received four weeks of antibiotic therapy with resolution of her pericarditis.

Discussion:
Purulent pericarditis is a rare condition with multiple etiologies described in the literature. Anaerobic bacteria present a challenge due to the difficulty in isolating organisms as well as its increased resistance to antibiotics. Several reports have documented contiguous or hematogenous spread of anaerobic organisms leading to pericarditis. Bacteroides species are part of the normal flora of the human intestine and cohabits this location with other bacteria without frequent pathogenicity. However, significant infections can occur when the bacterium is displaced outside of the intestines and into the surrounding tissue or bloodstream. To our knowledge, this is one of the first reported cases of suspected B. fragilis liver abscess causing pericarditis. Prompt diagnosis and treatment are necessary as cardiac tamponade and constrictive pericarditis are common complications of purulent pericarditis. The mortality rate is estimated to be around 20% to 30%. Pericardiocentesis or pericardiotomy is indicated if there is evidence of tamponade or a high clinical suspicion of purulent pericarditis. Intrapercardial thrombolysis is an option in cases where loculated fluid is present or when prevention of constrictive pericarditis is needed.
Thyroid eye disease in Hypothyroidism

Introduction:

Thyroid eye disease (TED) is commonly seen in Grave’s disease and affects around 25% to 50% of patients. TED has also been rarely reported in hypothyroidism like the case described below.

Case:

This is a 76 year old female with past medical history significant for former smoking, hypertension, diabetes mellitus and hypothyroidism who presented with complaints of eye popping out of her sockets. She previously had been having annual eye examinations without any complaints. She was found to have floppy eyelids and pigmentary and degenerative changes in her left retina. The floppy eye lids were attributed to obstructive sleep apnea and a sleep study was recommended. She later presented to primary care physician with complaints of eyes popping out from her left eye socket and excessive dryness due to inability to close her left eye while sleeping. On examination of eyes, she was noted to have increased bilateral inferior scleral show and upper lid retraction in left eye. She was sent to the ophthalmologist who also reported the same findings and diagnosed her with mild thyroid eye disease in the setting of hypothyroidism with left eye proptosis. There were no signs of extra ocular movement limitation, lagophthalmos, exposure keratopathy or optic nerve compression. Her TSH was 13.4 at that time and her TPO antibodies and TSH receptor antibodies were negative. Her TSH was normalized subsequently with increased levothyroxine dose. She was prescribed eye lubricants and is being managed conservatively.

Discussion:

Thyroid eye disease (TED) also called thyroid associated ophthalmopathy or Grave’s ophthalmopathy is the most common orbital disease, affecting 25%-50% of patients with Grave’s disease. The pathogenesis behind TED is thought to be fibroblast activation by antibodies that act against the TSH receptor or IGF-1 present in the retro-orbital tissues that causes fibroblast activation, excess production of hyaluronic acid and edema resulting in proptosis. However, this can also be seen in hypothyroid or euthyroid state. In patients with Hashimoto’s thyroiditis, eye changes occur in the absence of TSH-receptor antibody production. These findings suggest that a specific link between thyroid autoimmunity and ophthalmopathy has not been established in all situations. There have been small studies that have raised the possibility that autoimmunity against calsequestrin and collagen XIII may play a role in the pathogenesis of TED. The role of genetic factors also remains unclear and larger studies are needed to explore these areas.
Residents Fellow Clinical Vignette

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Now wreaking havoc on the central nervous system; Aseptic meningitis, a rare complication of immune check point inhibitors.

Immune checkpoint inhibitors have revolutionized the management of solid organ tumors. They are well known for their endocrinologic toxicities but neurologic adverse effects are less frequently documented. Aseptic meningitis is a rare complication of immunotherapy with an estimated incidence of 0.1-0.4%. We present a case of a 64 yo F who presented with fever, headache and recurrent altered mental status and was found to have aseptic meningitis secondary to use of Nivolumab and Ipilimumab in the absence of infectious symptoms and negative lumbar puncture.

64 yo F with history of melanoma metastatic to the brain on dual immunotherapy with Nivolumab and Ipilimumab presented to the Emergency Department with fever, chills, upper respiratory symptoms and headache that started on a road trip from North Carolina to Rochester with sick friends. She was febrile to 101.6 but otherwise hemodynamically stable. She had a negative respiratory gram stain and culture as well as atypical viral panel but received a dose of Vancomycin and Zosyn in the Emergency Department. She clinically improved over the next 2 days, however, she subsequently developed sudden onset altered mental status so she was started on Ampicillin and Acyclovir and received a dose of Methylprednisone for presumed meningitis. She had no meningeal irritation signs and denied recent use of NSAIDs. Routine blood tests, including a blood count and biochemistry were within normal limits. CT scan head showed pansinusitis and stable vasogenic edema surrounding known metastatic foci within the temporal lobes bilaterally but no signs of meningeal inflammation. She subsequently underwent a lumbar puncture which showed 139 cells with 35% neutrophils, glucose of 56 and protein 95 and negative encephalitis PCR panel. CSF was negative for presence of melanoma cells. She was thought to have partially treated bacterial meningitis and discharged home on antibiotics but she returned 2 days later with recurrent altered mental status along with headache and stiff neck. Repeat CT showed stable frontal lobe metastasis and resolved sinusitis. Antibiotics were held this time and her altered mental status cleared spontaneously over the next couple of days.

Presentation of immune checkpoint inhibitor induced neurotoxicity occurs within the first six weeks of treatment and our patient had just received his first dose of Iplimumab a week prior to presentation. His clinical picture was initially suspicious of a bacterial illness given his exposure to sick contacts and presence of respiratory symptoms but negative CSF analysis and rapid clinical recovery made a bacterial cause less likely. Moreover, CSF findings of mild leukocytosis along with elevated protein and negative cultures for bacteria or viruses was consistent with an aseptic picture. With the widespread use of immunotherapy nowadays, hospitalist physicians should be aware of the potential toxicities including neurologic side effects.
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When hoofbeats lead to Zebras instead of Horses

Autoimmune glial fibrillary acidic protein (GFAP) antibody has long been known as a bio-marker for necrotizing meningoencephalitis in dogs but meningoencephalitis associated with IgG binding to GFAP among humans was not known prior to 2016. The disease is rare with an unknown incidence but a slight female predominance and tends to present in patients over 40 years of age. We present the case of a previously healthy young male who was diagnosed with GFAP associated astropathy after a long struggle of recurrent hospitalizations and exhaustive clinical work up for recurrent altered mental status.

57 yo M presented to the Emergency Department with altered mental status and declining situational awareness which was preceded by a month long history of global headaches and low grade fever. Family denied any recent travel, pet exposure, insect bites or consumption of raw meat/fish. Labs were significant for normal complete blood count and biochemistry. ESR, CRP, RPR, ANA, HIV and urine drug screen were negative. CT head and MRI brain were unremarkable. EEG showed diffuse encephalopathy but no elliptogenic foci. He was started on Vancomycin, Ampicillin and Acyclovir. A lumbar pucture was performed but patient became obtunded shortly after the procedure requiring intubation. CSF analysis revealed 344 cells, 99% lymphocytes and normal protein. Viral encephalitis panel was unremarkable and patient was diagnosed with lymphocytic meningoencephalitis. Patient improved clinically and was discharged but returned 1 week later with acute encephalopathy. Neurologic exam revealed abnormal gait but was otherwise non focal. MRI brain showed abnormal leptomeningeal enhancements within the infra- and supratentorial regions. Repeat Lumbar Puncture showed nucleated cells 323 with 96% lymphocytes and he was started on Acyclovir and Doxycycline. Lyme antibody, legionella, histoplasmosis, alpha-1 antitrypsin, ceruloplasmin, heavy metal, AFB and viral encephalitis panel testing was negative. EEG revealed epileptiform discharges from the left posterior quadrant so he was started on anti-epileptics. Autoimmune encephalitis panel tested positive for Glial fibrillar acidic protein (GFAP) and he was started on IV steroids and IV immunoglobulin which led to clinic improvement and discharge to rehabilitation medicine.

GFAP astrocytopathy usually involves the cerebra, meninges, spinal cord and optic nerve, and manifests as fever, headache, encephalopathy, myelitis, and/or abnormal vision. Like other autoimmune encephalopathies, GFAP tends to occur more commonly among patients with underlying malignancy with ovarian teratomas being the most common. No cancer was found in our patient. Knowledge regarding neuropathology is limited as no post-mortem reports have been published thus far. Although common things remain common, our case suggests how hospitalists may occasionally come across a diagnosis that can only be solved with a high degree of suspicion and ability to hunt for zebras in a stable of horses.
PULMONARY VEIN THROMBOSIS ASSOCIATED WITH A LARGE HIATAL HERNIA

Introduction

Pulmonary vein thrombosis (PVT) is a rare condition. The limited existent literature consists mainly of case reports, so the incidence rate is unknown. However, it is potentially fatal with the risk of acute systemic arterial embolism, thus merits clinician awareness. We report a case of PVT secondary to compression from a large hiatal hernia.

Case presentation

An 82-year-old male was referred to the emergency department after incidental finding of left lower lobe pulmonary vein thrombosis (PVT) on his chest computerized tomography (CT) for his large hiatal hernia follow-up. He was discovered to have an asymptomatic large hiatal hernia 7 years ago and was treated conservatively. However, 17 months prior to admission the patient developed intermittent epigastric pain and dyspnea on exertion (DOE). Chest CT at that time showed a large hiatal hernia compressing his left lung with associated atelectasis, but no vascular abnormalities. Due to his age, he not considered a surgical candidate for hiatal hernia repair. Although epigastric pain and DOE persisted, extensive work-up, including pulmonary function tests, nuclear cardiac stress test, coronary angiogram, and a barium swallow were all negative, leading to the repeat CT scan of his chest done the day of admission. In the ED, lower extremity Doppler study was negative. Physical exam was notable for abdominal distention but no tenderness. He was treated with Lovenox and has bridged on to therapeutic doses of Coumadin. After several weeks of anticoagulation, his epigastric pain and dyspnea resolved, and he remained free of systemic embolic events.

Discussion

PVT is a rare condition associated with significant morbidity. Although many patients are asymptomatic, PVT can manifest with nonspecific chest pain, cough, hemoptysis, and dyspnea. Complications include pulmonary edema or infarction, or rarely, systemic embolism, including limb ischemia, stroke, and splenic infarction. Physical exam is often not helpful. Etiologies based on case reports have identified anatomic changes as the main cause, including lobectomy, lung transplant, and compression phenomena from large hiatal hernias or masses. Malignancy and hypercoagulability are potential risk factors. Rare cases in sickle cell and atrial fibrillation have also been reported. Diagnosis relies on chest CT with IV contrast, trans-esophageal echocardiography, pulmonary angiogram or MRI. At this time, there are no guidelines available for PVT treatment. In most cases, anticoagulation was initiated. For close monitoring, many clinicians are more comfortable with heparin or low molecular weight heparin bridging to Coumadin. Chest CT follow up studies are often performed. Anticoagulation has been continued until clot resolution for those with an identifiable etiology. For those whose PVT etiology cannot be fixed, indefinite anticoagulation seems reasonable. Clinicians should keep PVT in mind due to its nonspecific symptoms but potentially severe complications.
New York Chapter
American College of Physicians

Annual Scientific Meeting

Resident / Fellow/Medical Student
Quality- Patient Safety
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RICHMOND UNIVERSITY MEDICAL CENTER

Study in Reducing Blood Culture Contamination Costs

Background: True positive blood cultures are crucial to instituting necessary life-saving measures. However these same measures can be harmful and costly when cultures are falsely positive resulting in unnecessary antibiotics, delayed procedures, prolonged hospital stays, and increased costs. National target contamination rate is set at 2-3%. This study started with a goal to reduce our hospitals 2014 contamination rate, 5.07%, to the minimal.

Methods: A multi-disciplinary approach was pursued. Mandatory video tutorials on proper sterile techniques, online certifying assessments, and customized blood collection kits were instituted. Using data from electronic medical records, contaminants were identified via chart reviews completed in 2014, 2015, 2016, and 2018. Personnel with high contamination rates were identified and retrained.

Results: Over a 5 year time period, the implemented methods resulted in a gradual reduction of contamination rates from 5.07% in 2014, 4.28% in 2015, and 2.56% in 2016, and, finally, this last year, 1.96% in 2018, below National Targets. Though cost data from previous studies are lacking, it can be inferred that the low contamination rate of 2018 significantly minimized the associated costs of wasted antibiotics and unnecessary hospital days. In a 6 month period of 2018, contaminations resulted in an expenditure of $5,043.52 for antibiotics and $72,732 for hospital days. The cost would likely have been higher if house staff was not already accustomed to accounting for the possibility of contamination in treatment planning.

Conclusion: Blood culture contamination is a patient safety and healthcare cost issue. It increases the risk of potential harm and overall costs. In order to reduce the blood culture contamination rates, we provided ongoing education to staff, utilized customized blood culture collection kits, maintained surveillance, and provided performance feedback to personnel. As a result, over a 5-year span, contamination rates were reduced from 5.07% to our current below the national rate of 1.96%. It should be noted, however, that each yearly rate has been determined by a different study team so there is a concern that a difference in surveillance methods may affect the true rates. Nonetheless, the trend is undeniable showing that the measures implemented to reduce contamination are effective.
**IS IT TIME TO SAY GOODBYE TO YOUR PPI?: DEPRESCRIBING PROTON PUMP INHIBITORS IN AN ACADEMIC PRIMARY CARE PRACTICE**

**Purpose:**
To identify and evaluate patients on proton pump inhibitors (PPI) and to deprescribe if appropriate. Deprescribing is the planned process of reducing or stopping medications that may no longer be of benefit or may be causing harm.

**Methods:**
In a Patient-Centered Medical Home (PCMH) primary care practice at Albany Medical Center, ten Internal Medicine residents identified and evaluated their patients during office visits to determine whether it would be appropriate to deprescribe PPIs. During a 12-month period, we evaluated and identified patients who were being prescribed longer courses and/or higher doses of PPIs than necessary. Patients with a history of poorly controlled acid reflux, Barrett’s esophagus, chronic non-steroidal anti-inflammatory use, severe esophagitis, or bleeding ulcer were excluded. After identifying those patients appropriate for deprescribing, we used a resident-created shared decision-making tool to explain the risks and benefits of PPIs and to formulate a deprescribing plan. The deprescribing plans included educating patients in non-pharmacologic management, tapering or immediately discontinuing PPI, or transitioning to a histamine type-2 receptor antagonist (H2RA). We followed up with patients undergoing one of these plans in the office or by phone after 5 weeks to assess the efficacy of deprescribing.

**Results:**
93 patients were identified as being prescribed PPIs. 40 patients (43%) were identified as being appropriately prescribed PPIs and excluded from the study. 53 patients (57%) were identified as appropriate for deprescribing. In this group, the average duration of PPI use was 4.5 years. Of the 53 patients, 36 patients (68%) agreed to a trial of deprescribing. At the conclusion of the study, 24 of the 36 patients (67%) had been successfully deprescribed with well-controlled symptoms, reduced or complete cessation of PPIs, or transition to H2RAs.

**Conclusions:**
This pilot study demonstrates that in our primary care practice, a significant number (57%) of patients taking PPIs were being prescribed longer courses and/or higher doses of PPIs than necessary. Our results also show that PPIs can be safely and effectively decreased to a lower therapeutic dosage, deprescribed completely, or transitioned to a H2RA. This study highlights the importance of the primary care physician’s role in ensuring that patients are prescribed PPIs appropriately in order to minimize long-term side effects. The next phase of this study is to expand this intervention across our practice by increasing awareness of all of our providers in identifying and evaluating patients for appropriateness of deprescribing PPIs.
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Documenting Obesity in Patient Chart and its Positive Impact on Health: A Retrospective Study from Adult Primary Care Center

Background:

Obesity is a major preventable public health concern with increasing global prevalence. It is a risk factor for health problems such as diabetes, hypertension, vascular disease, and sleep apnea. Primary care providers (PCPs) are well positioned to identify and treat patients with obesity. Physician awareness of the diagnosis with appropriate documentation of the condition sets the stage for ideal care. In this study, we assessed the impact of obesity diagnosis documentation on various patient care factors in the Adult Primary Care Center (APCC).

Method:

Patients with a BMI >30 were included in the study and charts (n=171) were reviewed for variables such as patient weight, BMI, blood pressure, HbA1c, thyroid function, activity level and nutrition class referral. Initial visit and follow up visit data from 2017-2018 (mean duration between visits, 477 days) were compared between 2 groups: those with obesity documented (n= 67) and those without obesity documented (n= 104) in the patient chart. Data were collected by retrospective chart review from electronic medical record system and analyzed using Microsoft Excel and SPSS for t-test and chi square test. Significance was accepted for P<0.05.

Result:

Of 171 patients included in the study, 67 patients (39%) had obesity documented as a diagnosis in the health record. Of those, 44 patients (65%) were referred to a nutritionist and 20 patients (30%) attended the nutritionist class. Those with obesity documented compared to those without obesity documented had mean weight loss of 2.6kg v weight gain of 0.7kg (P=<0.05), rate of screening for diabetes (61% vs. 44%; p=0.019), thyroid dysfunction (48% vs. 29%; p=0.012) and referral to nutritionist (66% vs. 25%; p=<0.0001). Further improvement of “good” functional status was evident in obesity documented patients (initial vs. follow up visit; p-value) (25% vs. 30%; p<0.05) but mean change in systolic BP mmHg (2.08 vs. 0.25; p=0.29), diastolic BP mmHg (0.36 vs. -0.79; p=0.55) and HbA1c value (-0.67% vs. 0.13%; p=0.16).

Conclusion:

In this study, we found that documentation of obesity in the health record was associated with weight loss and other favorable care measures such as rates of diabetes screening and nutritional assessment.
Hypertensive Retinopathy Screening Rates in an Urban Primary Care Clinic

Introduction: Hypertensive retinopathy is a serious, but preventable, complication of chronic hypertension that is associated with an elevated risk of stroke even after hypertension is controlled. Consequently, the Joint National Committee on Prevention, Detection, Evaluation, and Treatment of High Blood Pressure (JNC 8) recommends yearly ophthalmology screenings for retinopathy in patients with hypertension. Given this recent change, we hypothesized that we had not adequately been screening patients for hypertensive retinopathy in our urban outpatient primary care teaching clinic.

Methods: An anonymous electronic survey was sent to all categorical internal medicine residents at an urban teaching hospital in New York City in November of 2017 asking them to estimate how often they screen their hypertension continuity patients for retinopathy. Additionally, the charts of all adult patients at our resident clinic with an active diagnosis of essential hypertension on their problem list, who were seen for an office visit of any kind from January 1, 2016 through December 1, 2017 were reviewed to determine if these patients were referred to ophthalmology as well as the indication for referral. Charts were also reviewed for an uploaded ophthalmology visit note to account for patients who did not require referrals. There were no exclusion criteria.

Results: A total of 37/81 (46%) residents responded to the survey. Of those who responded, 19% (7) said they "always" screen hypertensive patients for retinopathy, 16% (6) selected "most of the time," 49% (18) selected "sometimes," and 16% (6) responded "never." During the designated time period, 546 patients had an active diagnosis of essential hypertension, 168 (33%) of which were seen by an ophthalmologist within the past year. However, of the 168 seen by an ophthalmologist, 100 (60%) were seen for management of chronic diseases like diabetes and rheumatoid arthritis, 41 (24%) were seen for an acute ocular complaint, and 18 (11%) were being followed for a chronic ocular problem. Only 9 (5%) were referred specifically for hypertensive retinopathy screening.

Conclusion: Only one third of hypertensive patients at our resident clinic are followed by an ophthalmologist and only 5% were specifically referred for hypertensive retinopathy screening.

This low screening rate is concerning because hypertensive retinopathy is preventable with early identification and intervention. We have begun to improve management of hypertensive retinopathy at our clinic through resident education on the importance of ophthalmologic screening and bimonthly email reminders to reinforce that knowledge.
Screening for nephropathy in patients with hypertension: A survey of annual screening frequency in an urban clinic

Introduction: Hypertensive nephropathy is a potentially fatal complication of chronic hypertension. Consequently, the Joint National Committee on Prevention, Detection, Evaluation, and Treatment of High Blood Pressure (JNC 8) recommends yearly microalbuminuria screening for nephropathy in patients with hypertension. We hypothesized that residents at our urban outpatient primary care teaching clinic were not adequately screening patients for hypertensive nephropathy.

Methods: All categorical internal medicine residents at an urban teaching hospital received an electronic survey asking them to estimate how often they screen their continuity patients with hypertension for nephropathy. A chart review was conducted of all adult patients at the resident clinic with an active essential hypertension diagnosis on their problem list from January 1, 2016 through December 1, 2017. Patients with diabetes, chronic kidney disease and other diseases that cause nephropathy were excluded from the sample. Charts were evaluated for a microalbumin/creatinine order within the past year, the presence of microalbuminuria, and a prescription for appropriate therapy with Angiotensin Converting Enzyme-inhibitor (ACE-i) or Angiotensin II Receptor Blocker (ARB).

Results: A total of 37 out of 81 residents responded to the survey. Of those who responded, 19% (7) said they always screen hypertensive patients for microalbuminuria, 16% (6) selected “most of the time,” 38% (14) selected “sometimes,” and 27% (10) responded “never.” During the designated time period, 546 patients had an active diagnosis of essential hypertension, 301 of which were excluded for having additional risk factors for nephropathy. Of the remaining 235 patients in the sample, 33% (78) had been screened with a microalbumin/creatinine ratio in the past year and 22% (17) of them had microalbuminuria. Among the patients who had microalbuminuria, 82% (14) were prescribed an ACE-i or ARB.

Conclusion: Only one third of hypertensive patients were screened for microalbuminuria at our resident clinic during the study period. Although patients received appropriate treatment once microalbuminuria was identified, this low screening rate is concerning because hypertensive nephropathy is preventable with early identification and intervention. We plan to improve screening rates for hypertensive nephropathy at our clinic through resident education and expect that increased screening will improve patient outcomes.
Using Constant Observation Status as an Interactive Treatment Modality

Background:
Patients with mental health issues, both in the psychiatric inpatient unit as well as on the medical floors, are often placed on increased Levels of Observation, or Constant Observation (CO) Status, during which the patient is paired with a Nursing Assistant who remains at their side 24/7. The current use of this status does not fully utilize the potential of staff to increase the quality of patient care. Staff typically has minimal guidance in how to engage with their patients, documentation practices are underdeveloped, and transfer of information to other team members is not maximized. Patients are not always clear on the reason for CO status or the role of the staff who sits with them.

Objectives:
Our objectives are to (1) Clinically define CO status as an interactive treatment modality; (2) Clarify the role of Nursing Assistants as members of the patients’ care team; (3) Enable and Empower Staff to better aid their patients; (4) Focus on improved patient outcomes for patients on CO status; (5) Enhance the level and descriptiveness of communication between various team members for this group of patients.

Methods:
To correct for these shortcomings, a comprehensive Training Program was developed. The Program consists of informational booklets, instructional techniques, video scenarios and discussions, question and answer sessions, and refresher courses throughout the year. The Program focuses on defining and explaining Mental Illness on a basic human level, fostering greater empathy and understanding of our patients, without over-complicated terminology. The Program also teaches exercises on how to communicate with patients, de-escalate crises, and minimize Behavioral Emergencies. Included in the Program is how to work with patients with Special Needs, such as Intellectual Disability, Visual or Otological Difficulties, Mutism, and more.

Anticipated Results:
We are expecting a significant increase in positive outcomes, including: (1) reduced use of manual restraints; (2) reduced requests for PRN medications; (3) reduced incidents of violence, both between patients as well as between patients and staff; (4) decreased allegations made against staff; (5) improved relationships between patients and caregivers; (5) decreased number of patients on CO Status at any given time; (6) decreased in average length of CO status; (7) decreased length of stay in general; (8) increased cost effectiveness; (9) improved patient outcomes. These variables have been measured before training was implemented, and will be continuously measured throughout the course of training and beyond.
A QUALITY INITIATIVE TO INCREASE SCREENING FOR OBSTRUCTIVE SLEEP APNEA (OSA) IN AN ADULT GENERAL MEDICINE CLINIC

BACKGROUND: OSA is an increasingly prevalent but under diagnosed condition with serious clinical consequences. OSA is often associated with hypertension, heart failure, depression, motor vehicle accidents and other chronic conditions. The aim of our project was to increase screening, diagnosis, and treatment of OSA in a general medicine resident clinic.

METHODS: We conducted a Quality Improvement (QI) project at an academic primary health care center after Institutional Review Board (IRB) approval. This was a retrospective pre and post intervention study done between August 2018 and February 2019. To begin with, the residents were instructed to screen their patient panel to identify patients with Body Mass Index (BMI) greater than 30; patients with diagnosis of OSA or documented STOP-BANG score (loud Snoring, Tiredness, Observed apnea, high Blood Pressure, Body mass index, Age, Neck circumference, and Gender) were excluded from the study population. Over the course of the next 6 months residents were educated regarding the utilization of STOPBANG score on all their obese patients at risk for OSA. Patients with high scores were referred for polysomnography. The primary outcome measured was the number of patients screened for OSA and the secondary outcome measured was the number of patients with a new diagnosis of OSA.

RESULTS: Our clinic has 104 residents with an overall patient population of 4,867. 39.01% (1899/4867) were diagnosed with obesity with a BMI of >30. 707 patients already carried a diagnosis of OSA or had a documented STOPBANG score. The pre-intervention screening rate was calculated as 37.23% (707/1899). Following our intervention of resident education, an additional 186 patients were screened resulting in a post-intervention screening rate of 47.02% (893/1899). Of the 186 patients, 79 underwent polysomnography, 86% of them were tested positive i.e., at the end of our study 68 patients were newly diagnosed with OSA.

CONCLUSION: OSA has been recognized as an important cause of medical morbidity and mortality. Effective diagnosis and treatment of the disorder has been associated with significant improvement in quality of life. Our study utilized regular feedback and education on OSA to increase screening rates and diagnosis which parallels with the provision of quality patient care. We focused on improving awareness among the residents for utilization of screening tools which further contributed to the increasing diagnosis of patients with OSA.
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HPV vaccination rates in lupus (SLE) and primary care patients in the clinic following a new FDA update in Gardasil-9 use in adults up to 45.

Background/purpose: The incidence of cervical dysplasia and cancer in women with SLE is higher than the general population. Routine HPV vaccination has 90% cervical cancer coverage and has been able to produce a protective response with seroconversion rates exceeding 79% in SLE women. Recently the FDA approved Gardasil 9 use for adults up to age 45, expanding the number of patients that could benefit from prophylactic HPV vaccination. We wanted to evaluate after the release of these new guidelines if age appropriate HPV vaccination is being done in SLE patients in comparison with general population.

Methods:

A retrospective chart review was done for an 8-month period to compare vaccination rates between primary care patients and SLE patients following the new FDA approval. Subjects were women ages 18 to 45. In the primary care clinic, 5 patients were randomly selected each month and were compared with matched SLE patients that followed at least twice per year in the lupus clinic. The health care maintenance section was reviewed to assess vaccination status.

Results:

A total of 40 patients from primary care clinic and 35 SLE patients from lupus clinic where compared between October 5th, 2018 to May 30th, 2019. Using the Fisher exact test 17.50% from the primary care clinic (95% CI (7.3-32.8)) and 14.30% from the SLE clinic (95% CI (4.8-30.3%)) had completed HPV vaccination HPV (p=0.76). Both groups had poor rates of vaccination by the guidelines and the difference in the vaccination rate was not statistically significant.

Conclusions:

Prophylactic HPV vaccination should be encouraged in all patients up to age 45 as part of primary prevention measure to decrease disease burden related to HPV infection. Special attention should be taken in high-risk populations such as SLE women. With new updates in Gardasil-9 use, more women will benefit from HPV vaccination, and this will require that primary care doctors/gynecologist and rheumatologists share equal responsibility to ultimately increase vaccination rates. Based on this data we are piloting a prospective quality improvement project to improve the HPV vaccination rates in our primary care and rheumatology clinics.
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Hard of Hearing Patients Experience Increased Length of Inpatient Hospital Stay

Purpose: To assess differences in length of stay (LOS) data for hard of hearing (HOH) patients as compared to that of hearing patients.

Background: Disabilities increase LOS in internal medicine inpatients (3), often subjecting patients to greater financial and clinical burden. Reduced LOS decreases the risk of hospital acquired complications, enhances quality of care, and increases hospital operational efficiency (1). Longer hospitalizations in people with impaired activities of daily living, such as vision and hearing disabilities, may also be negatively associated with functional recovery (4). Few studies have evaluated the specific impact of sensory disabilities on LOS. Visual impairments have been seen to increase LOS by 2.4 days (2), but the impact of hearing impairments on LOS of both elderly and non-elderly patients is unknown.

Methods: Data from hearing and self-identifying HOH patients over the age of 18 in all Albany Medical Center inpatient units were collected from January 2017 through May 2019. Analysis was based on all patient visits with discharge dates within this period. LOS and expected length of stay (eLOS) were compared between HOH and hearing patient populations. The effect of hearing aid location on these variables was also evaluated.

Results: Across all age groups above age 24, HOH LOS and eLOS were significantly higher than those of hearing patients (p<0.01 for all). HOH patient LOS was greater than their respective eLOS (p<0.001). The LOS and eLOS of HOH patients without hearing aids were longer than those of HOH patients wearing hearing aids (p<0.001 for both). Those wearing hearing aids had LOS and eLOS values comparable to those of hearing patients (LOS p=0.24, eLOS p=0.15).

Conclusions: On average, HOH patient eLOS was one day longer than that of their hearing counterparts and their actual LOS was greater than their eLOS. Of the HOH patients evaluated, over 90% did not possess hearing aids. For those that did, wearing hearing aids matched HOH LOS and eLOS to those of hearing patients, across all age groups. As the first study to examine these variables in HOH patients ages 18 and older, this analysis will increase awareness in hospital systems regarding a disparity in care among HOH patients and encourage quality improvement for this population.
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The Effect of Inpatient Palliative Care Consultations on the Costs of Care

Purpose: The purpose of this study is to characterize the financial effect of inpatient palliative care consultation and to assess if the effect is correlated with length of stay (LOS).

Methods: This study was a prospective pre/post analysis of patients who received inpatient palliative care consults at Albany Medical Center (AMC). Data was collected by the AMC Data Analytics Department on patients’ LOS, CMS-Expected LOS, and Total Hospital Costs in the three days before and after a palliative care consult. Comparisons were made between total costs before and after palliative care consults to determine the financial impact. A regression analysis was used to determine how the timing of palliative care consults impact patients’ overall length of stay. Moreover, comparisons of patients’ LOS to CMS-expected LOS were conducted for early and late palliative care.

Results: Patients who received a palliative care consult (n = 104) had an average LOS of 14.1 days, but an average CMS-expected LOS of 6.0 days. Regression analysis showed that for every day sooner a patient received a palliative care consult, their LOS decreased by 1.34 days (p < 0.001). Furthermore, patients who received a palliative care consult within 5 days of admission had a significantly lower LOS (p < 0.001) and a LOS significantly closer to that predicted by the CMS (p < 0.001) relative to the cohort who received a later palliative care consult. The average daily hospital cost one day before a palliative care consult were $905 higher compared to one day after the consult (p < 0.001). Similarly, average total aggregated costs three days before a palliative care consult were $2,315 higher compared to three days after the consult (p < 0.001).

Conclusions: Inpatient palliative care consultation offers hospitals significant cost-savings by decreasing patient LOS and decreasing costs per day. It also offers the opportunity for hospitals to approach the targets for LOS suggested by CMS. Although the reimbursement for palliative care consultation may not be as lucrative as procedure-based care, these data suggest that the return on investment for palliative care will be seen as an increase in net income due to decreased cost of care. Given that palliative care has also been shown to offer better quality of care in previous research, hospitals should still invest in the resources necessary to offer palliative care early to the most complex and costly patients.