BREAKOUT
ROOM
NUMBER SIX
CV 51 - 60
IS THE PHYSICAL EXAM STILL IMPORTANT IN THE NEW MILLENNIUM?
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Introduction: History and physical exam is the basic yet the most important step in evaluating patients. Sister Mary Joseph nodules are rare malignant metastatic umbilical nodules, usually associated with gastrointestinal (50%) or genitourinary (25%) cancers. The etiology of SMJ nodules remains unclear; however, it has been hypothesized that their presence stems from spread due to extension via the lymphatic system or blood vessels. This condition was named for Sister Mary Joseph (1856-1939), a surgical assistant for Dr. William Mayo, who noted the association between paraumbilical nodules observed during skin preparation for surgery and metastatic intraabdominal cancer confirmed at surgery.

Case presentation: A 67-year-old female presented with abdominal distention, lower extremity edema, and unintentional weight loss of 7 kg of 1-year duration. Vital signs and labs were within normal limits. Physical exam was significant for a supraumbilical subcutaneous mass measuring 3 x 2 x 3 cm, consistent with a SMJ nodule. An enlarged left inguinal lymph node measuring 3 x 2 x 2 cm and bilateral +3 symmetric lower extremity edema were also noted. CT scan showed a large cystic and solid pelvis mass, multiple mesenteric and omental nodules and masses, subcutaneous nodules at the level of the umbilicus, pelvic lymphadenopathy, and multiple splenic masses. The patient underwent total abdominal hysterectomy with radical dissection for ovarian carcinoma, repair incision omentectomy, splenectomy, and embolectomy. She was diagnosed with stage 4 high grade serous carcinoma and was initiated on adjuvant chemotherapy.

Discussion: A detailed history and complete physical exam remain the cornerstone of the patient evaluation. While the level of detail necessary to truly elucidate patient complaints and uncover subtle examination findings can be challenging given the increasing patient loads, paperwork, and non-clinical responsibilities, it is imperative to remain diligent and continue to practice patient-centered medicine. Despite the significant advancements in medical imaging and laboratory diagnostics, rising health-care costs compels the physician to practice guideline directed medicine and order wisely. In this case, a detailed history and physical examination revealed the presence of a SMJ nodule which prompted additional diagnostic workup, and ultimately a timely diagnosis and management.

CV 51

BREAKOUT ROOM 6
ASCENDING AORTIC ANEURYSM IN A CASE OF TERTIARY SYPHILIS

Cardiovascular syphilis was once responsible for 5-10% of all cardiovascular deaths worldwide; however, today it is extremely rare in developed countries. In a study from The Cleveland Clinic looking at more than 1000 aortic specimens, none of the 52 proven cases of aortitis were related to syphilis [1]. Cardiovascular manifestations of syphilis include aortitis secondary to obliterator endarteritis of the aortic vasorum causing destruction, fibrosis, and calcification of the media layer of the aorta. Aortic root aneurysm is a serious complication with high risk of rupture and can occur several years after inoculation in untreated patients [4]. The diagnosis of syphilitic aortitis is confirmed by serologic testing of syphilis and a characteristic location of aneurysm which usually involves the root of aorta. [1-4].

A 59-year old man presented to the emergency department with acute shortness of breath and new onset orthopnea of 2 weeks. Cardiac auscultation revealed a 2/6 systolic ejection murmur and 2/6 diastolic murmur at the right upper sternal border. Laboratory work up revealed normal troponin. Electrocardiogram (EKG) showed anteroseptal ST elevation without acute ischemic changes. Chest radiograph showed cardiomegaly and bilateral pulmonary infiltrates. CT angiography was unremarkable for pulmonary embolism but demonstrated a large aortic root aneurysm measuring 5.9 cm. Echocardiography showed severe left ventricular dilation and aortic insufficiency. Possible causes of aortic root aneurysms including syphilis were investigated. Syphilis screening test was positive which was confirmed by a positive T. pallidum particle agglutination assay (TPPA). The patient was diagnosed with tertiary cardiovascular syphilis without neurosyphilis. He was treated with penicillin and underwent surgical repair of the aortic root one week later. Histopathology confirmed the diagnosis of syphilitic aortitis.

Tertiary syphilis often presents several years after initial infection and usually after a latent phase, making it difficult to diagnose. Patients can have nonspecific neurological or cardiovascular presentations that make the diagnosis even more challenging. Additionally, the serologic testing of syphilis may be negative in 25% of cases due to low titer of circulating antibodies in tertiary syphilis [5]. Therefore, a high index of suspicion is required for early recognition of syphilitic aortic aneurysm in high risk individuals with high risk sexual behaviors [5]. Early treatment with antibiotic therapy and surgical repair of syphilitic aortic aneurysms can help prevent life threatening complications and death.

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ABSTRACT FORM: Must be at least 10-point font. A sharp typeface will help reproduction. Be sure to single-space and STAY WITHIN THE BORDERS!

WHEN THE LINES ARE BLURRED: PITUITARY ADENOMA & APoplexy
Fatima Hafizi, MD; Osejic Oriaifo, MS; Luis Rivera-Ramirez, MD
Pituitary adenomas are relatively common, spontaneously occurring intracranial masses. They may remain asymptomatic or cause symptoms secondary to mass effect or hormone hypersecretion. A rare but potentially life-threatening complication of pituitary adenoma is pituitary apoplexy, occurring in the context of hemorrhage or infarction of the pituitary. While the most common presenting symptoms include severe headache and associated visual disturbances, catastrophic hormonal abnormalities of the hypothalamic-pituitary-end organ axis may occur.

A 31-year-old female with a medical history notable for migraine headache and cabergoline-responsive, prolactin-secreting pituitary macroadenoma presented with a severe frontal headache of progressive severity of one-month duration. Associated symptoms included blurred vision without vision loss or diplopia that had been ongoing for the past few months, without associated nausea, vomiting, or lightheadedness. Conservative management with over-the-counter Tylenol/NSAIDs had failed. Of note, the patient had stopped cabergoline 8 months prior. On presentation, vitals were unremarkable except for an elevated blood pressure. Prolactin was elevated to 475ng/mL and ACTH (14pg/mL) and cortisol (8.8mcg/dL) were normal. MRI of the brain demonstrated a pituitary adenoma that had increased in size compared to prior imaging to 18x15x15mm with subacute hemorrhage. The patient was diagnosed with pituitary apoplexy. Neurosurgery was consulted and recommended initiation of methy/prednisolone with subsequent endoscopic pituitary adenoma resection. Her headaches resolved and she was discharged home on levothyroxine, hydrocortisone and an antihypertensive with outpatient follow up.

Severe headache and blurred vision may be secondary to benign conditions such as migraine, however the presence of additional “red flags” – abnormal neurological exam, progressive severity/frequency – should prompt not only a detailed history and physical examination, but also diagnostic imaging, such as MRI. Pituitary apoplexy is a rare but potentially life-threatening condition that often initially only presents with headache and mild visual disturbances. However, it may be complicated by various degrees of pituitary failure and resultant hormonal deficiencies. Diagnosis and management require dedicated pituitary imaging and evaluation of the hypothalamic-pituitary axis, with subsequent hormone replacement.
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Indicate your participation in research process (4 sentences or less): I initially saw this patient on pulmonary consult service. I then researched pulmonary talcosis further and wrote the case report.

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A CASE OF LEFT CIRCUMFLEX ARTERY ANEURYSM AND SPONTANEOUS THROMBUS IN A 32-YEAR-OLD. Khan Z, MD, Sahbaz J, MD, Shkullaku M, MD, Chahal DS, MD. University of Maryland School of Medicine and VA Medical Center, Baltimore, Maryland.

Coronary artery aneurysms (CAAs) are a rare entity with a mean incidence of 1.65%. CAAs are defined as localized coronary artery segment dilatations of greater than 1.5-fold compared with normal adjacent segments. While frequently associated with underlying atherosclerotic coronary artery disease (>50% of cases), causes like vasculitis (Kawasaki Disease), connective tissue disease, genetic predisposition (polymorphisms), infections, and iatrogenic injury (percutaneous coronary intervention induced) have postulated mechanisms underlying the development of CAAs. While often asymptomatic, angina and associated arterial thrombi are often found in symptomatic manifestations of CAAs.

A 32-year-old man with a history of type 2 diabetes and tobacco use presented to the emergency department with several hours of non-radiating substernal chest pain associated with intermittent dyspnea, nausea, and vomiting. Physical examination was notable for mild distress, sinus pauses with otherwise normal cardiac exam, and clear lungs without the presence of peripheral edema, jugular venous distention or crackles. A chest x-ray was unremarkable. Initial electrocardiogram (EKG) demonstrated normal sinus rhythm without ST changes, but repeat EKG about four hours later demonstrated accelerated idioventricular rhythm (~75 beats per minute). Troponin I initially measured at 0.75 ng/mL but rose to 28 ng/mL in ~6 hours. Patient was taken for urgent coronary angiography and found to have total occlusion of the proximal left circumflex (LCx) artery secondary to extensive clot within an aneurysmal dilatation. TIMI 3 flow was ultimately restored using aspiration thrombectomy and rheolytic thrombolysis, as well as systemic anticoagulation (heparin and eptifibatide). Post-catheterization transthoracic echocardiogram (TTE) demonstrated posterolateral wall hypokinesis with an ejection fraction of 30-35%. The patient now feels back to his baseline on dual antiplatelet (DAPT) and guideline directed medical therapy for heart failure.

Coronary artery aneurysms (CAAs) are likely under-identified and often asymptomatic findings, but can be associated with critical occlusion, stenosis, or rupture of coronary arteries. In patients with known CAAs, providers should aim to optimize cardiovascular disease modifiable risk factors (hyperlipidemia, diabetes, blood pressure), and consider referring patients for interventional evaluation depending on CAA size or risk of rupture.
IgM Multiple Myeloma

**Introduction:** Immunoglobulin type M (IgM) multiple myeloma (MM) is a very rare subtype of MM, it accounts for only 0.5% of all cases of MM. Furthermore, IgM MM is hard to differentiate Waldenström macroglobulinemia (WM). It is important to differentiate MM from WM as treatment and prognosis differ.

**Case:** A 67-year-old male presented to the clinic complaining of headaches for one month, preceded by a one-month history of dyspnea on exertion, bilateral lower extremity cramps and heaviness during ambulation. Vital signs were within normal limits. Physical examination was unremarkable. Laboratory investigations showed total protein 10.3 g/dL, albumin 3.6 g/dL, globulin 6.3 g/dL, albumin to globulin ratio 0.6 and creatinine 1.1 mg/L. MM was suspected and further workup revealed an elevated M spike on serum protein electrophoresis (SPEP) from a IgM lambda monoclonal protein of 4.1g/dL, IgM level >5850mg/dL, urine protein 8.8g, urine albumin 44.7%, urine α-1-globulin 5.4%, and plasma viscosity 3.22 mPa/s. Positron emission tomography scan showed focal uptake in the midsternum and mesenteric nodes in the right lower quadrant. Bone marrow biopsy revealed 40-50% lambda positive plasma cells. Fluorescence in situ hybridization analysis revealed a CCND1/IgH t(11;14) gene rearrangement in 6% of cells. He was treated with a total of 8 cycles of cyclophosphamide, bortezomib and dexamethasone (induction therapy), followed by cyclophosphamide for mobilization and autologous bone marrow transplant. Post-transplant the patient was kept on lenalidomide as maintenance therapy. One-year post-transplant, there was no plasma cell infiltration on bone marrow biopsy and no M-spike on SPEP. Patient is currently doing well on lenalidomide maintenance therapy.

**Discussion:** IgM MM is rare, and it can be difficult to differentiate from other IgM paraprotein disease, mainly WM. In one large series of patients with IgM paraprotein, WM was shown to be more common than IgM MM. Several features can be used to differentiate these two conditions including cytogenetics, cell morphology, and presence of bone involvement. In cytogenetic analysis a t(11:14) gene translocation mutation is identified in 83% of cases of non-secretory IgM MM cases but not in WM; whereas, the somatic mutation MYD88 L265P is found in WM but not in MM. In WM cells have lymphoplasmacytic morphology and bone marrow is not involved, while MM is purely plasmacytic and has bone marrow involvement.
RISK FACTORS IN THE DEVELOPMENT OF ACQUIRED METHEMOGLOBINEMIA

Introduction: Acquired methemoglobinemia is a rare adverse effect (0.035%) of some common drugs, including topical anesthetics, dapsone, nitrites and street drugs. A discrepancy exists between routine pulse oximetry, which measures around 85%, and PaO2 levels, which are normal/elevated. The diagnosis may be missed if co-oximetry is not done and if left untreated can be fatal. Here we present a case of benzocaine-induced methemoglobinemia.

Case description: A 60 year old male with no significant past medical history was admitted with bowel obstruction secondary to metastatic colon cancer and managed surgically. Postoperative course was complicated by sepsis from an intra-abdominal abscess. Hemoglobin levels during admission were 7.5-8 g/dL. Benzocaine gel was applied to relieve pain from oral ulcers, and one hour later he complained of chest pain and shortness of breath. He was hypoxic to 79% and tachycardic (124/min). PaO2 was elevated at 308.4 mmHg on 100% FiO2, and the low SpO2 was initially attributed to a malfunctioning pulse oximeter. However, observation that the blood was brown in color prompted co-oximetry testing, which revealed methemoglobin levels of 57%. After one dose of methylene blue, symptoms improved and methemoglobin levels fell to 0.3%.

Discussion: Acquired methemoglobinemia is a rare complication of topical benzocaine use, with an incidence of 0.067%. It affects people of all ages with no predilection for gender or ethnicity. Benzocaine is used in procedures such as TEE, EGD and bronchoscopy, and can be used for oral/dental pain.

Methemoglobin levels are normally maintained at or below 1% by the reducing enzyme cytochrome B5 reductase (Cyb5R).

Methemoglobin, formed by oxidation of ferrous (2+) to ferric (3+) ions, is unable to bind oxygen and causes a configuration change of the hemoglobin molecule which increases its affinity for oxygen.

Oxygen delivery is impaired, and a functional anemia develops. Symptoms range from mild to severe, and include cyanosis, dizziness, headache, nausea, dyspnea, coma, confusion and seizures.

Risk factors for acquired methemoglobinemia include active infection, anemia and inpatient status.

In this patient, pre-existing anemia likely exacerbated the symptoms of the functional anemia caused by methemoglobinemia. Similarly, symptoms can be exacerbated by underlying cardiac or pulmonary disease due to diminished reserve.

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Inferior Vena Cava Stenosis as a Cause of Abdominal Ascites
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Inferior vena cava (IVC) stenosis causing abdominal ascites is a well-documented complication of orthotopic liver transplantation, radiation therapy, and liver surgery. However, reports of ascites secondary to IVC stenosis have rarely been described in non-transplant patients without a history of radiation therapy or hepatic surgery. We describe a case of ascites secondary to IVC stenosis in a woman without a history of liver disease.

A 23-year-old woman with a history of systemic lupus erythematosus (SLE) complicated by lupus nephritis and cerebritis and prior DVT/PE on warfarin presented with worsening abdominal distention for two months. Prior to her presentation at our hospital, she had two admissions to an outside hospital, and an extensive workup there was unrevealing. Her symptoms were ultimately attributed to her SLE, and her prednisone dose was increased to 60 mg BID. Despite her increased prednisone dosage, however, her symptoms continued to progress and thus, she presented to our hospital for further evaluation.

On presentation, her vitals were unrevealing. Physical examination was notable for a BMI 31.3 kg/m², a distended abdomen, and palpable splenomegaly. Labs demonstrated a hemoglobin 8.0 g/dL, platelet 105, creatinine 2.0, and normal LFTs. Autoimmune liver serologies including AMA, LKM, and ASMA, were negative. Viral hepatitis studies were negative as well. Ceruloplasmin and A1AT were within normal limits. Liver duplex ultrasound revealed moderate ascites with concern for cirrhotic liver morphology, without evidence of hepatic vein thrombosis. Paracentesis demonstrated a SAAG 1.3, ascitic protein 2.9 g/dL, PMN 65, and negative cytopathology and bacterial/fungal cultures. Hepatology was consulted, and a transjugular liver biopsy was pursued. The hepatic venous pressure gradient was 1 mmHg with a free hepatic vein pressure 42 mmHg and right atrial pressure 15 mmHg, concerning for an obstruction between the hepatic veins and right atrium. Liver biopsy revealed sinusoidal dilation with concomitant hepatic congestion. A follow up MRI liver was obtained, which demonstrated high grade stenosis in the superior portion of the intra-hepatic IVC. Interventional radiology performed an IVC venoplasty with significant improvement in the stenosis post-procedure.

IVC stenosis should be considered as a cause of new onset ascites in a non-liver transplant patient without prior radiation therapy or liver surgery with transudative ascites and high ascitic protein.

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ABSTRACT FORM: Must be at least 10-point font. A sharp typeface will help reproduction. Be sure to single-space and STAY WITHIN THE BORDERS!
THE CASE OF A YOUNG WOMAN WITH FEVER, SORE THROAT, AND POLYARTHRALGIA

History: A 19 year-old woman with a history of ADHD, anxiety, and recurrent fevers presented with fever, sore throat, and polyarthralgia to the Johns Hopkins Emergency Department in February of 2020. In the year prior to admission, the patient reported fevers as high as 101-103°F occurring an average of 3 times per week. She also endorsed intermittent sore throat and waxing and waning polyarthralgia, which did not always align with febrile episodes. She was hospitalized several times for fevers, oral ulcers, and sore throat, and she had undergone infectious and rheumatologic workups. Previously, she was treated with courses of antibiotics and antifungals without symptomatic improvement. On admission, she was nearing the end of a prolonged taper of prednisone. On presentation, she reported worsening fevers over the preceding weeks (Tmax 103°F at home), a sore throat that started 5 days prior to presentation, and worsening polyarthralgia.

Objective Data:

Physical exam: Prominent cervical lymphadenopathy. Laboratory data: CBC and CMP notable for mild normocytic anemia with Hgb 12 g/dL, ESR 3 mm/hr, CRP 14.4 mg/dL.

Microbiology: Negative blood/urine cultures, rapid strep, respiratory viral panel, ASO titer, Toxoplasma IgM, monospot, EBV VL, CMV VL, HIV Ab/Ag, HAV IgM, HBV core IgM/sAg, HCV Ab, Mycoplasma Ab, treponemal Ab, GC/CT, stool bacterial NAT, cryptococcal Ab, Histoplasma Ab, Beta-D-Glucan, galactomannan, babesia microti Ab. Rheumatology labs: Notable for ANA 1:40 (homogeneous), RF 19.
Normal complement levels, IgG subclasses, CK, anti-Smith, anti-dsDNA, Ro/La, RNP, CCP, tissue transglutaminase, endomysial IgA. Imaging: CT head/neck notable for tonsillitis/pharyngitis without abscess and bilateral large, likely reactive, cervical lymph nodes.

Pathology: Esophageal biopsies with reactive changes, viral and fungal culture/stains negative. Lymph node biopsy with polymorphous lymphocytes, no malignant neoplasm.

Management: The patient met criteria for PFAPA syndrome (Periodic Fever, Aphthous Stomatitis, Pharyngitis, Adenitis) given regularly occurring fevers, cervical lymphadenitis, and pharyngitis in the absence of URI and neutropenia and being asymptomatic between episodes. She is older than expected for onset of PFAPA, although she reports episodes in her teens. She was started on prednisone 60mg with planned taper and colchicine 0.6mg BID for prophylaxis with symptomatic improvement and resolution of her fever. She was discharged home with plan for possible tonsillectomy and confirmatory genetic testing at NIH.
Acute Kidney Injury Secondary to Retroperitoneal Fibrosis

Introduction: Retroperitoneal fibrosis (RF) is a rare condition characterized by inflammatory and fibrous proliferation, usually around the infrarenal abdominal aorta, that is most often idiopathic and immune-mediated, and most commonly causes obstructive uropathy and renal failure in affected individuals.

Case: A 40-year-old female with hypertension presented to the hospital after being found to have a creatinine of 5.96 mg/dL as an outpatient. Over the course of the month before admission, she had progressively developed left leg swelling, bilateral flank pain, nausea and vomiting. She denied irritative and obstructive urinary symptoms, hematuria and constitutional symptoms or taking NSAIDs and herbal supplements. Serum creatinine in 2009 was 0.7mg/dL. Two months before current presentation, she had been treated for acute gastroenteritis and acute kidney injury (AKI) with a peak creatinine level of 9.3mg/dL which had improved to 1.9mg/dL at the time of discharge. During that admission, it was thought that her AKI had been due to severe dehydration, but an ultrasound had shown mild bilateral hydronephrosis without stones. Physical examination was significant for hypertension, mild tachycardia, BMI of 36, and non-pitting edema of the left leg. Laboratory investigations showed BUN 49 mg/dL, creatinine 11.1 mg/dL. Imaging studies showed mild bilateral hydronephrosis, proximal hydrourter, and increased retroperitoneal density seen around the aortic bifurcation. Soon after, she underwent double-J stent placement on the left kidney and a nephroureteral stent placement on the right with resolution of obstruction. Extensive autoimmune workup was unremarkable. A clinical diagnosis of retroperitoneal fibrosis leading to obstructive uropathy was made and she was started on induction therapy with prednisone. Patient did well and at the time of discharge serum creatinine was down to 1.3 mg/dL.

Discussion: The pathogenesis of RF involves a CD4+ cell-mediated immune response, which causes proliferation of B cells and fibroblasts in the periaortic retroperitoneum. Ureteral involvement is common, patients present with flank pain similar to renal colic and acute kidney injury when there is bilateral involvement. Lower limb edema can also occur due to lymphatic compression. Diagnosis is made via imaging studies such as computed tomography or magnetic resonance imaging. Retroperitoneal biopsy is only indicated in cases with atypical localization or with clinical findings suspicious for an alternative diagnosis. The goal of treatment is relief of ureteral obstruction followed by anti-fibrotic therapy to achieve disease regression. Despite its chronic-relapsing course, idiopathic RF has good renal outcomes, and end-stage renal disease progression is very rare with prompt treatment and follow-up.

Conclusion: Although rare, RF should be considered for patients presenting with acute kidney injury secondary to obstructive uropathy with paraaortic mass seen on imaging. Secondary causes should be excluded. The diagnosis of RF can be made clinically, without the need for a biopsy.

CV 60

BREAKOUT ROOM 6