TEAM 5 CVs
### Unusual Case of Systemic Hydralazine Induced Lupus

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Drug-induced lupus is an autoimmune disorder caused by chronic use of certain drugs that includes several antihypertensive medications. Hydralazine is the most common culprit and hydralazine-induced lupus syndrome (HILS) occurs in 5–10% of patients taking hydralazine. Here, we present an unusual case of HILS with renal involvement.

A 73-year-old Caucasian female with uncontrolled hypertension, diastolic heart failure, chronic kidney disease, and diabetes presented with progressive dyspnea and lower extremity swelling. On exam she had bilateral coarse breath sounds with rales and pitting lower extremity edema. Labs were significant for pancytopenia, elevated creatinine of 2.8 (baseline of 1.7), non-nephrotic range proteinuria and hematuria. Chest x-ray showed bilateral patchy ground-glass opacities. She was initially managed for pneumonia versus heart failure exacerbation. However, further workup for hematuria and proteinuria showed elevated ESR 48, CRP 32, ANA titer of 1:2560 in homogeneous pattern, low C3 and C4, positive P-ANCA, elevated Myeloperoxidase, positive PR3, but negative C-ANCA. Anti-histone antibody and Anti-dsDNA were also positive. Kidney biopsy showed immune complex related glomerulopathy with single cellular crescent suggestive of drug-induced glomerulonephritis with moderate arteriosclerosis. She had been on hydralazine for almost 3 years. She was managed for HILS with cessation of hydralazine and initiated on high dose steroid with gradual tapering. Her subsequent ANA was reduced to 1:320 in nearly 6 months with negative ANCA studies and improvement in renal function.

HILS is a result of an autoantibody reaction. Unlike classic systemic lupus erythematosus (SLE), HILS does not have diagnostic criteria. Clinical manifestations are non-specific and include arthralgia, myalgia, fever and serositis. Organ involvement is rare but possible. Anti-histone antibody is positive in 95% of the cases and anti ds-DNA can be positive <5% of the cases. Hence, a high index of suspicion is important in the early diagnosis and treatment of HILS, most importantly with cessation of the offending drug.
SEEEING DOUBLE: AN UNUSUAL CASE OF DIPLOPIA
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An 83-year-old male man with past medical history of hypertension, complete heart block s/p dual chamber PPM (2018), nephrolithiasis, BPH, and cataracts began experiencing double vision. Two weeks later, he also developed decreased vision in his left eye. During this time, he was evaluated by several ophthalmologists. Clinical exam was concerning for acute right inferior oblique palsy. He demonstrated restrictive strabismus with limited elevation bilaterally (but worse on the right), chemosis, and restriction was concerning for thyroid-associated eye disease or a carotid cavernous fistula. Cataracts were stable and considered non-contributory. Over the next few days, patient noted color desaturation and more rapid progressive vision loss in the left eye. A CT of the head and brain demonstrated enlargement of the right superior rectus and left lateral rectus muscles, with evidence of displacement of the left optic nerve. Thyroid stimulating immunoglobulins were less than detectable limits. Empiric prednisone 60 mg daily was initiated and CT of the orbits was performed, which showed confirmed compression of the left optic nerve sheath complex. He was directly admitted to the hospital and was treated pulse-dose methylprednisone 1 g daily for 5 days with frequent vision exams. CT chest/abdomen/pelvis was without readily attributable or easily biopsied lesions, full body PET showed FDG avid extracranial muscles with no distant metastasis, so the patient underwent left orbitotomy and biopsy. Preliminary permanent sections and flow cytometry returned with concern for B cell lymphoma. Given the rapidity of vision loss, the patient was transferred to the lymphoma service and immediately started on treatment therapy R-CHOP while awaiting final pathology results. A lumbar puncture ruled out primary CNS involvement. Prophylactic intrathecal methotrexate was administered. He was discharged on a prednisone taper and final pathology was consistent with marginal zone lymphoma. His vision improved rapidly over the next few days. Non-thyroid-related extracranial muscle enlargement (NTR-EOME) is uncommon, representing only 5% of cases in retrospective analysis. Inflammatory, vascular, and neoplastic processes are the most common etiologies, in decreasing frequency. Neoplastic processes represent approximately 20% of all NTR-EOME. In a retrospective study of NTR-EOME, biopsies where malignancy was identified were most commonly lymphoma with evidence of both extranodal marginal zone B cell lymphomas and MALT.

Program Director’s Name:
Dr. Sanjay Desai
(indicating review of abstract)
A CASE OF AUTOIMMUNE ENCEPHALITIS IN A PATIENT WITH METASTATIC NEUROENDOCRINE LUNG CARCINOMA TO THE BRAIN COMPPLICATED BY HSV ENCEPHALITIS. Sayad M, MD. University of Maryland School of Medicine and VA Medical Center, Baltimore, MD.

Autoimmune encephalitis is a severe inflammatory condition of the brain that is immune mediated, involving antibodies against neuronal cell surfaces and presynaptic membranes. This syndrome has been associated with patients with malignancy, immunosuppression, and preceding infection with HSV encephalitis.

A 49 year-old gentleman with history of neuroendocrine lung carcinoma with brain metastases status post resection and chemotherapy initially presented with fever and seizures. He was initiated on high dose dexamethasone, levetiracetam, broad-spectrum antibiotics with vancomycin and meropenem, and acyclovir. CT head was unchanged from prior imaging. A lumbar puncture (LP) was performed and cerebrospinal fluid (CSF) analysis was positive for Herpes Simplex Virus (HSV)-2. The patient was treated with acyclovir IV with improvement in his mental status. He was discharged to home with a to complete two-week course of anti-viral treatment for HSV encephalitis.

Weeks later, the patient presented with persistent altered mental status following prior hospitalization. He demonstrated confusion and short-term memory deficits. Magnetic resonance imaging (MRI) demonstrated new curvilinear enhancement of right temporal lobe concerning for persistent HSV encephalitis vs. post-treatment parenchymal changes. A LP was performed, and CSF analysis demonstrated lymphocytosis without leukocytosis. CSF analysis was negative for malignant cytology, HSV, or other infectious etiologies. There was concern for autoimmune encephalitis vs. paraneoplastic encephalitis. A paraneoplastic panel was collected. The patient was treated with a five-day course of IVIG and high dose steroids with subsequent improvement in mental status. His MOCA score improved to 21, from a pre-treatment score of 18. Given improvement with high dose steroids and negative paraneoplastic panel, the patient’s presentation was ultimately attributed to autoimmune encephalitis. He was discharged to home on a prednisone taper, PPI, and PCP prophylaxis.

The patient was recently seen by his outpatient oncologist. Some short-term memory loss remains; however, the patient can perform all of his activities of daily living.

It is important to have a high suspicion for autoimmune encephalitis in patients with malignancy and a history of HSV encephalitis. Early treatment with immunotherapy is vital to prevention of irreversible damage that is mediated by immune destruction of neurons.

Program Director’s Name: Susan D. Wolfsthal

(indicating review of abstract)

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A DISSEMINATED CASE OF DIAGNOSTIC DILEMMA
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A disseminated cryptococcal infection as a presenting manifestation of
AIDS is unusual. We present a case of a middle-aged man with
nonspecific symptoms manifesting as a disseminated cryptococcal
infection.

A 59 y/o man presented to the ED with cough and generalized weakness
for 2 weeks. He had subjective fevers and chills. His past medical
history included HTN, Gout and hepatitis C (treated). Social history
included occasional alcohol use and no prior hx of smoking or illicit
drug use. He is currently not sexually active. On physical exam, he was
afebrile, tachycardic at 111, tachypneic at 24 and saturating 91% on RA.
Lung exam revealed mild bilateral crackles at bases. Neurological exam
was normal. Labs revealed a BUN of 43 and Cr of 2.3 (baseline:
17/0.91) with a FeNa of 0.3%. D-Dimer was high at 10.99. V/Q scan
showed no evidence of PE. Chest X ray was normal. Initial diagnosis
was community acquired pneumonia and was treated with ceftriaxone
and azithromycin. His AKI was thought to be pre-renal in nature due to
dehydration and was given IV fluids with no improvement in kidney
function. However, he continued to have persistent fevers without
leukocytosis. A CT chest showed small areas of infiltrate in the right
upper lobe, b/l lower lobes and splenomegaly, CT abdomen showed
lymphadenopathy in upper abdomen and splenomegaly with portal
hypertension. Subsequent labs revealed newly diagnosed HIV (CD4
count 161). CT-guided biopsy of a left retroperitoneal lymph node
showed cryptococcus invasion. Both serum and CSF fluid were positive
for cryptococcal antigen. CT-guided biopsy of left kidney showed focal
glomerulosclerosis suspicious for disseminated fungal infection with
micro abscesses. Blood fungal cultures were positive for yeast. He
received induction therapy with liposomal amphotericin and fluconazole
and subsequently consolidation therapy with fluconazole for 4 weeks for
disseminated cryptococcal meningitis.

AIDS presenting as cryptococcal disseminated meningitis is an
extremely rare presentation. When patients do not improve with
appropriate treatment for their initial diagnosis, further evaluation is
critical to recognize alternative diagnoses.
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EVALUATING CHEST PAIN IN PATIENTS WITH RE-IMPLANTED CORONARIES: CORONARY CARDIOVASCULAR COMPUTED TOMOGRAPHY MAY BE THE TEST OF CHOICE.

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Introduction: The Bentall procedure is cardiac surgery involving composite graft replacement of the aortic valve, aortic root, and ascending aorta, with re-implantation of the coronary arteries into the graft. Compromise of coronary arterial flow as a result of complications of coronary reimplantation is a rare but potentially life-threatening complication of the Bentall procedure.

Case: A 42-year-old female with history of Tetralogy of Fallot, pulmonary atresia and major aortopulmonary collateral arteries who underwent initial repair with a right ventriculotomy to pulmonary artery conduit and ventricular septal defect closure at age 32 with development of severe aortic valve insufficiency due to aortic dilation three years after repair procedure and underwent subsequent aortic valve replacement via the Bentall procedure. Since procedure, the patient has complained of long-standing intermittent exertional chest discomfort and tightness that radiated to the left shoulder which resolved with rest. Echocardiography and ECG were unremarkable. Coronary computed tomography angiography (CCTA) was ordered for further assessment.

CCTA demonstrated severe non-atherosclerotic stenosis of the mid left main coronary artery secondary to severe vessel angulation with poor penetration of contrast through the distal LAD. Given the location of the stenosis with high risk for significant ischemic event or arrhythmogenic sudden cardiac death the decision was made to fix the lesion via percutaneous cutaneous intervention. Coronary angiography confirmed CCTA findings, demonstrating a severely angulated mid left main and resultant 95% luminal stenosis. The stenosis was treated successfully with PCI using a single drug-eluting stent. The procedure was uncomplicated and the patient reported resolution of her symptoms.

Discussion: Patients who are status post cardiothoracic surgery, especially the adult congenital heart disease population, often have post-surgical chest pain syndromes and complex residual anatomy. CCTA is an excellent diagnostic tool for these patients as the superior spatial resolution provides accurate anatomic assessment and can simultaneous evaluate for coronary atherosclerosis as well as post-surgical complications. Post-Bentall coronary stenosis is a known complication of the surgery but is rare and may be unrecognized by treating providers. Evaluation of this complication in symptomatic patients via CCTA is an attractive non-invasive option.
Pulmonary Kaposi Sarcoma despite on HAART Therapy:

Introduction: Kaposi sarcoma is the most common tumor in HIV patients and while cutaneous involvement is the most common manifestation, visceral involvement, while much less common, is not unusual. Here is a case of pulmonary involvement in a patient with Kaposi sarcoma requiring adjunctive therapy to HAART.

Case Report: A 53 year-old male initially presented with a nasopharyngeal mass growing into his orbits a year back biopsy of which revealed diffuse large B-cell lymphoma with positive EBV. Further testing at the time showed patient was HIV positive. Patient was started on R-CHOP therapy and methotrexate with subsequent HAART once in remission. CD4 count at the time was 100. While on therapy, violaceous cutaneous lesions involving the face, chest and inner thigh were noted along with new pulmonary nodules found on CT chest despite an improving CD4 count of 168 and viral load of <20. Biopsy of skin lesions were positive of Skin lesions were biopsied that came back positive for HHV-8 and a subsequent bronchoscopy with bronchoalveolar lavage (BAL) was performed that came back negative for infections along with a negative cytology. Using the positive HHV-8 on skin biopsy, with CT results showing pulmonary nodules worrisome for pulmonary involvement of Kaposi sarcoma, and a negative BAL, a diagnosis of pulmonary Kaposi sarcoma was made. Patient was subsequently started on chemotherapy with paclitaxel along with HAART.

Discussion: The most common manifestation of Kaposi sarcoma involves cutaneous tissue with visceral involvement being much less common with 3 most common site being lung, GI tract and lymph nodes. In one study done over a 4 year period, of 318 people with Kaposi sarcoma only 19 were found to have pulmonary involvement. Pulmonary Kaposi sarcoma usually has concurrent skin findings in 80% of case. The diagnosis of pulmonary Kaposi sarcoma does not always require a lung biopsy to be made, rather a diagnosis of exclusion can be made once cutaneous involvement has been confirmed and a negative BAL with characteristic CT findings of nodules. With visceral involvement, such as lung, adjunctive chemotherapy with HAART is usually employed.
AUTOIMMUNE ENTEROPATHY IN A YOUNG ADULT MALE TREATED WITH STEROIDS
Rammy Korkor, MD; Marilyn Baird-Howell, MD; Richard Williams, MD; Medstar Health Internal Medicine, Baltimore, Maryland

Autoimmune enteropathy (AIE) is a rare condition characterized by intractable diarrhea refractory to dietary therapy, usually seen in infancy but very rarely seen in adults. At least 50% of diagnosed patients have circulating anti-enterocyte antibodies. Associations with other autoimmune diseases have been described, however most patients do not have an identified abnormality of immune function.

A 24-year-old African-American man presented with concerns of diarrhea, nausea, vomiting, and abdominal pain associated with a 50-pound weight loss over a five-month period. Physical examination was remarkable for cachexia and diffuse abdominal tenderness. The patient appeared malnourished and was not able to tolerate oral diet and hence was started on total parenteral nutrition. Fecal fat was positive. Extensive workup with imaging and laboratory studies for immunodeficiency syndromes, autoimmune disorders, and celiac disease including anti-enterocyte antibodies was negative. Therefore, an EGD was done which revealed severe villous atrophy with marked active chronic enteritis/duodenitis and loss of goblet cells on biopsies of the duodenum and jejunum. AIE was suspected, and within 24 hours of starting corticosteroid treatment, the patient was seen comfortably enjoying a cheeseburger. The prednisone was tapered, and the patient was started on infliximab as an outpatient.

The diagnostic criteria of AIE include 1) severe villous atrophy not responding to dietary restriction 2) circulating intestinal autoantibodies or other autoimmune disease, and 3) absence of a primary immune deficiency. Despite increasing awareness, the pathogenesis, histologic, immunologic, and clinical features of AIE remain uncertain. Histopathology of the small intestine in patients with AIE typically reveals severe villous atrophy and an inflammatory infiltrate involving the lamina propria. In contrast to other etiologies for villous blunting, such as celiac disease, there is typically a milder degree of intraepithelial lymphocytosis. Management of autoimmune enteropathy includes providing nutritional support and adequate hydration, with concurrent immunosuppressive therapy with corticosteroids, infliximab, rituximab, or azathioprine in refractory cases. Recently, biological agents have been introduced, with apparent beneficial effects. Autoimmune enteropathy is a rare heterogeneous disease that should be considered in the differential diagnosis of malabsorption with small bowel villous atrophy after other common causes of malabsorption have been ruled out. Physicians should be aware of this chronic debilitating disease, with more research to establish both etiology and treatment guidelines needed for the appropriate management of AIE.
PANCREATIC INSUFFICIENCY PRESENTING AS FECAL INCONTINENCE

**Introduction:** Fecal incontinence is a devastating disease which can be a manifestation of pancreatic insufficiency and is associated chronic diarrhea due to malabsorption.

**Case Presentation:** The patient was a 44-year-old woman with a history of hypothyroidism, IDDM complicated by gastroparesis and neuropathy who was admitted for severe hypothyroidism accompanied by fecal incontinence. Her symptoms started 2 years ago, resulting in regular use of absorbent pads. As a result, she curtailed her social activities. She denied urinary incontinence or prior abdominal surgery. She also had been experiencing diarrhea for 2 years, but is not sure if the diarrhea preceded the incontinence. She described her stools as watery, sticky, foul-smelling, yellow-colored, and non-bloody. She had bowel movements 30-60 minutes after each meal; therefore, she did not eat properly. She was seen by her primary care physician and was prescribed loperamide, which did not improve her symptoms. Pepto Bismol helped the diarrhea slightly, but not incontinence.

She had cholecystectomy 4 years prior. She denied alcohol use. Review of systems was remarkable for 90 lb weight loss in the past 12 months, skin discoloration in lower extremities, and decreased vision at night. Medications include levothyroxine as well as methadone for back pain. Physical examination was remarkable for dry and thickened skin, coarse facial wrinkles, and angular cheilitis. The anal tonus was normal and there was no rectal prolapse on Valsalva maneuver.

Laboratory tests were remarkable for prolonged PT, elevated INR, hypocalcemia, deficiency of vitamins ADEK, all consistent with fat malabsorption. Fecal analysis was remarkable for decreased elastase, suggesting pancreatic insufficiency. Pancreas appeared normal on abdominal CT. Patient was started on pancrelipase and her fecal incontinence and diarrhea completely resolved within 2 days.

**Discussion:**
Fecal incontinence has been reported to affect 2 to 17% of the general population and up to 50% of the nursing home residents. Evaluation of bowel habits in these patients is critical for further workup, especially to identify the status of chronic diarrhea. In this case, further investigations for the etiology of diarrhea revealed pancreatic insufficiency and malabsorption, which was successfully managed with pancrelipase replacement.
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A CASE OF CYTOMEGALOVIRUS COLITIS IN AN IMMUNOCOMPETENT PATIENT. Gheysens, K. MD; Ajaka, L. MD. The University of Maryland School of Medicine and VA Medical Center, Baltimore, MD.

Cytomegalovirus (CMV) is a member of the Herpesviridae family; infection typically causes flu-like or no symptoms in immunocompetent hosts. However, it is a significant cause of morbidity and mortality in immunocompromised individuals, causing infections such as pneumonitis, retinitis and colitis.

A 54 year-old male with past medical history of idiopathic thrombocytopenic purpura (ITP) presented to medical care following a visit to his home country of Ghana with persistent bloody diarrhea, abdominal pain, anorexia and fevers. Upon arrival to the United States, he sought care at a community hospital, where he was diagnosed with and treated for pan-sensitive E. coli bacteremia with a 7-day course of antibiotics. He continued to have abdominal pain and diarrhea, even after receiving a second course of antibiotics. His pain worsened and he presented to our institution’s emergency room for evaluation.

Physical exam was notable for tachycardia to the 100s and diffuse abdominal tenderness. Lab work from the community hospital was reviewed. Stool studies were negative for H. pylori antigen, Clostridium difficile infection, Shiga toxins 1 and 2, fecal leukocytes, and occult blood. Thick and thin smears were also negative for malaria. An abdominal computed tomography (CT) scan was concerning for colitis; the patient subsequently underwent a colonoscopy. Pathologic examination of biopsy was consistent with CMV colitis.

Additional work-up for underlying immunodeficiency or malignancy was negative. Serum testing included non-reactive human immunodeficiency virus (HIV), non-reactive human T-lymphotropic virus (HTLV), negative hepatitis serologies, normal IgG levels and low IgM/IgA levels thought to be secondary to his prolonged diarrheal illness. PET CT showed no evidence of malignancy. The patient was started on a three-week course of oral ganciclovir and discharged home. His course was complicated by re-admission 10 days later with toxic megacolon. He was switched to IV ganciclovir, and his symptoms improved; he was discharged again on oral ganciclovir. He completed a six-week total course of antiviral therapy, with complete resolution of symptoms.

CMV colitis is an uncommon imitator of more common colitis and inflammatory bowel diseases. It is unusual for CMV to cause colitis in a healthy, immunocompetent patient, but can occur, as evidenced by this case. Therefore, it should always be considered in the patient presenting with persistent symptoms suggestive of colitis, regardless of underlying immune status.
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SPINAL DURAL VENOUS FISTULAS: AN UNDERRECOGNIZED AND CURABLE MIMIC OF TRANSVERSE MYELITIS
Ting-Jia Lorigiano, MD and Dave Furfaro, MD

Introduction
Vascular arteriovenous malformations and fistulae represent a reversible, yet underrecognized cause of myelopathy. Definitive therapy is often delayed owing to misdiagnosis as transverse myelitis. Here, we present a case highlighting the diagnosis and treatment of myelopathy due to a vascular malformation.

Case Description
A 64-year-old woman with past medical history of intravenous opioid use now on suboxone, hypertension, subarachnoid hemorrhage status post left posterior communicating artery clipping, and seizures presented with six months of bilateral ascending lower extremity numbness and weakness, as well as urinary incontinence. Physical exam was notable for grade 2/5 motor function in bilateral hip and knee flexors, extensors, abductors, as well as in ankle plantar and dorsiflexors, with bilateral numbness and decreased saddle sensation with loss of rectal tone and absent patellar reflexes. Electromyogram and nerve conduction studies demonstrated bilateral radiculopathy, without neuropathy. Lumbar puncture was notable for elevated protein, normal glucose, and white blood cell count of 7, and negative infectious workup. Cerebrospinal fluid immunoglobulin index was normal with negative oligoclonal bands. Cytology and flow cytometry did not suggest malignancy. Systemic infectious, rheumatologic, and malignancy workup was unrevealing. Magnetic resonance imaging of the spine revealed longitudinal, patchy enhancement extending from the 2nd thoracic vertebra to the conus with associated enhancement of the cauda equina, with dilated cervical veins. Spinal angiogram confirmed a spinal dural arteriovenous fistula at T12 subsequently managed with embolization. Post-embolization patient began to improve with inpatient physical therapy.

Discussion
Spinal dural arteriovenous fistula (SDAVF) carry an estimated annual incidence of 5-10 cases per million and cause progressive disability from venous hypertension and congestion, resulting in spinal cord ischemia. Here we describe the workup, diagnosis, and management of SDAVF, and how SDAVF may be differentiated from transverse myelitis by elevated CSF protein without markers of inflammation and characteristic MRI findings of congestion. Spinal vascular malformations are rare and underrecognized, but curable and should be considered when evaluating the myelopathic patient.
NEUROSYPHILLIS: A GREAT MIMICKER
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MRI findings in neurosyphilis (NS) vary in nature and may mimic several common CNS pathologies such as PML, HIV leukoencephalopathy, vasculitis, and myelopathy while presenting as white matter changes, diffuse cerebral atrophy, and parenchymal changes. Hence, NS should be in the differential for a wide range of MRI abnormalities. The prevalence of NS among HIV-infected people is substantial since HIV infection may alter the natural course of syphilis due to the profound defects in cell-mediated immunity it can cause.

A 28-year-old man presented with 2-month history of generalized weakness, unsteady gait, and falls. Anorexia with unintentional weight loss and light-headedness were also reported. Orthostatic vitals were negative with other vital signs remaining unremarkable. The patient did not report any history of HIV/AIDS or syphilis, nor observe any recent rashes. Physical examination revealed wide-based gait, shuffling steps, and decreased strength in right leg. Laboratory data was pertinent for positive HIV antibody and an absolute CD4 count of 27. MRI brain showed marked white matter signal abnormalities in frontal temporal and parietal lobes. CSF serologies for JC virus, cryptococcal antigen, HSV, and ANA were found to be negative. Serum RPR titers were positive and although CSF VDRL was negative, treatment for syphilis with penicillin G was started due to high clinical suspicion and positive serum VDRL. Bactrim was also started for PCP prophylaxis given the low CD4 count. The patient’s weakness improved after treatment, with no further episodes of falls.

NS can present in several ways ranging from cerebral atrophy, white matter lesions, cerebral infarctions, and edema which can be nonspecific to NS as other infections may also present with such findings. These findings, if present in the HIV positive patient, should prompt clinicians towards NS as one of the differentials, as it is easily treated in the antibiotic age. It is important to recognize the diversity of these diagnostic images to promptly treat this infection in time to prevent permanent neurologic dysfunction such as general paresis, locomotor ataxia, and optic atrophy. Follow-up imaging and RPR titers are warranted to ensure improvement in CNS pathology.
ANCA-NEGATIVE MICROSCOPIC POLYANGIITIS; THE PATH TO DIAGNOSIS
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Microscopic polyangiitis (MPA), an autoimmune systemic vasculitis involving small and medium sized vessels often affecting the kidney and lung, is classified as a subset of anti-neutrophil cytoplasmic antibody-associated vasculitis (AAV). Anti-neutrophil cytoplasmic antibody (ANCA) is detected in 90-95% of cases of MPA and its detection has become an essential step in diagnosis; however, rare cases of ANCA-negative MPA have been reported.

A 23-year-old man presented with dyspnea, pleuritic chest pain and hemoptysis for 2 weeks. He was found to be hypertensive (BP 223/136) and tachycardic. Physical exam was unremarkable except for bilateral wheezing on lung exam. CT chest was significant for bilateral centrilobular opacities and hilar/mediastinal lymphadenopathy. He was found to have acute kidney injury and trace proteinuria. He was initially started on antibiotics and steroids and was worked up for autoimmune disease. CRP (86) and ESR (67) were elevated. ANA was weakly positive (1:160); however, other factors and antibodies including anti-ds-DNA, RF, anti-CCP, p-ANCA, c-ANCA, Scl-70 Auto Ab, GBM Ab, SS-A, SS-B, anti-Sm Ab, C3, C4, and anti-histone Ab were negative. Echocardiogram revealed EF of 40-45% as well as moderate concentric left ventricular hypertrophy. Infectious disease workup was unremarkable. Our patient eventually had lung biopsy which revealed extensive intraalveolar hemorrhage with linear PMN collections in alveolar septa, and capillaritis indicative of MPA. He was lost to follow up and subsequently readmitted. He had kidney biopsy for worsening proteinuria, revealing FSGS and changes concerning for MPA as well as tubular atrophy and interstitial fibrosis.

This case illustrates a rapidly progressive form of MPA involving the lung, kidney and myocardium with negative ANCA results. MPA can present in different ways and doesn’t always have a stereotypic presentation. ANCA plays an important role in diagnosis and prognostication of MPA; however, negative ANCA may delay the diagnosis or lead to misdiagnosis. ANCA may become positive later in the course of the disease; however, in cases with high clinical suspicion for ANCA-associated vasculitis (AAV) and negative ANCA, efforts should be made to obtain a tissue biopsy in order to make a precise diagnosis and initiate appropriate management.

Program Director's Name: Stephanie Detterline MD

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THROMBOSIS IN BLEEDING CROHN'S DISEASE
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There is a strong association between venous thromboembolic events and Crohn’s disease, with a roughly 3-fold increase in venous thrombosis risk compared to the general population, rising to an 8 to 16-fold during flare. Arterial thrombotic events (ATE) have rarely been described. The majority of patients with Crohn’s disease and ATE who undergo hypercoagulability workup do not establish an identifiable coagulopathy.

A 31-year-old man with Crohn’s disease with rectal fistula presented with bloody diarrhea for 1 week. He complained of sharp, intermittent abdominal pain worse with oral intake, as well as intermittent left thigh aching and numbness. He was afebrile, normotensive, and tachycardic to 126 bpm on admission. He had epigastric abdominal tenderness and palpable lower extremity pulses bilaterally. Digital rectal exam was notable for trace redness. At home he had been taking Prenisolone 20 mg twice daily, Mesalamine 400 mg thrice daily, and had received Infliximab 10 days prior to admission. Labs revealed hgb 6.7 g/dl for which he received 1 unit of blood. C. difficile colitis was ruled out. Testing for JAK-2 V617F mutation was negative and CRP was elevated to 38. CTA of A/P demonstrated irregular wall thickening of the sigmoid colon and focal noncalcified plaque in the distal aorta and left common iliac artery with more than 50% occlusion. Due to concerns for ongoing bleeding, anticoagulation was held but prednisone was started. Thereafter, he underwent left iliopsoas thrombectomy and placement of a left common iliac stent. He was started on aspirin and clopidogrel, after which bloody bowel movements resumed; therefore, only clopidogrel was continued with plans to restart aspirin in one month.

ATE is a rare but serious complication of Crohn’s disease. This patient did not have other risk factors for ATE; however, his Crohn’s disease was poorly controlled, which may have been a risk factor for thrombus development. Due to active Crohn’s disease flare, he was not a candidate for medical management and required surgical thrombectomy and stent placement. Though pathogenesis of ATE is not clearly established, it has been thought to be secondary to prothrombotic stimulus from local and systemic inflammation. Long term control of Crohn’s disease for prevention of extraintestinal complications, such as ATE, are important as it may be conservatively managed if there are no complications from the Crohn’s.
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Renal Cell Carcinoma in a Patient with Bilateral Renal Masses

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Introduction: Renal Cell Carcinoma (RCC) accounts for about 85% of renal cancers and 2 percent of cases of renal cancer are associated with inherited syndromes.

Case Report: A 61-year-old male with a family history of Kidney cancer in his father and past medical history of chronic lower back pain (on opioids) presented with hematuria and right flank pain. CT abdomen and pelvis revealed solid masses measuring 8.2 x 6.8 x 8.8 cm and 3.3 x 3.1 x 0.2 cm in the lateral midpole and upper pole of the right kidney respectively. A small exophytic lesion in the anterior aspect of the mid to lower pole of the left kidney was also noted along with extensive retroperitoneal lymphadenopathy suspicious for malignancy. Retroperitoneal lymph node biopsies were performed which showed necrotic cellular material suspicious for a malignant process but not diagnostic. He then underwent Robotic Radical nephrectomy of right kidney with lymph node sampling which revealed Papillary Renal Cell Carcinoma Type 2 with confirmed metastasis to the retroperitoneal lymph nodes on histopathology.

Discussion: Papillary Renal Cell Carcinoma Type 2 tends to have early age of onset (mean age 40 years) and is associated with HLRCC (Hereditary Leiomyomatosis and Renal Cell Cancer), an autosomal dominant syndrome comprising of cutaneous and uterine leiomyomas and kidney cancer. HLRCC is caused by FH (Fumarate Hydratase) gene mutations, with a lifetime renal cancer risk for FH gene mutation carriers to be about 15%. Although there is no consensus for clinical surveillance, abdominal imaging, preferably MRI annually is recommended.

Conclusion: Patients on chronic opiates can mask early symptoms of cancer pain and those with chronic back pain can confound symptoms of retroperitoneal cancers. Those with family history of cancers require cancer screening vigilance.
BILATERAL FACIAL NERVE PARALYSIS: A DIAGNOSTIC CHALLENGE
Sama Alreddawi, MD; Louis Saade, MD; MedStar Health Internal Medicine, Baltimore, Maryland

Facial nerve palsy (FNP) has a wide range of differential diagnoses and becomes a diagnostic challenge when less than 2% of patients present as bilateral FNP. Bilateral FNP is a rare clinical syndrome secondary to many causes such as infiltrative disease, infection, autoimmune disease, trauma, tumor, idiopathic, and very rarely (0.4-2.5%) isolated cranial nerve involvement in diabetes mellitus (DM).

A 65-year-old man with IDDM, hypothyroidism, and Parkinsonism, presented with gradual worsening of facial weakness in the setting of bilateral FNP complicated by dysarthria. Complete workup, which included LP with negative Lyme titers, was unremarkable. Miller Fisher Syndrome and Myasthenia Gravis were ruled out with negative GQ1b and AChR binding and blocking antibodies. Whole body CT was nondiagnostic. Brain MRI revealed symmetric enhancement of bilateral facial nerves with no other cranial nerve abnormality suggesting idiopathic bilateral Bell’s palsy. An EMG/NCS displayed a mixed polyneuropathy usually consistent with diabetic neuropathy. Regardless, a presumed diagnosis of idiopathic bilateral Bell’s palsy was made, and he was treated with 1 week of acyclovir and glucocorticoids with only minimal improvement. The prolonged history of IDDM, comprehensive negative workup, nerve studies exhibiting mixed polyneuropathy, along with lack of improvement to acyclovir and glucocorticoids, leads us to believe that this is a case of DM-induced bilateral facial nerve palsy.

DM was reported as the cause of bilateral FNP in 1 out of 43 patients in a case series. Patients with recurrent FNP are 2.5 times more likely to have diabetes. Diabetic neuropathy should be included in the differential diagnosis of unilateral or bilateral facial nerve palsy in patients with complicated DM.
A RARE CASE OF SEVERE LIVER INJURY FROM ADULT ONSET STILL'S DISEASE
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Adult onset Still's disease (AOSD) is a rare multisystem clinical disorder of unknown etiology, characterized by involvement of multiple organ systems including the liver. Hepatic involvement is frequently observed in the course of AOSD with mildly elevated transaminase. Acute liver failure (ALF) is a rare manifestation of AOSD. Treatment usually consist of corticosteroids with addition of disease-modifying anti-rheumatic drugs (DMARDs) and anti-tumor-necrosis-factor (TNF) agents in patients with severe liver injury refractory to corticosteroid treatment. A 22-year-old woman with no significant PMH was diagnosed with AOSD a month prior to admission (fulfilling all four major and all five minor items of Yamaguchi's criteria), was treated successfully initially with pulse dose corticosteroids but then developed elevated LFTs. While on prednisone taper, she presented with syncope, fever, arthralgia, abdominal pain, and scleral icterus with lab findings suggestive of markedly elevated liver enzymes, LDH and serum ferritin. Work up, including autoimmune and infectious tests, excluded all potential other causes of liver failure. Bone marrow biopsy ruled out hemophagocytosis and, given elevated levels of IgG, she underwent a liver biopsy for suspicion of autoimmune hepatitis (AIH). Biopsy showed centrilobular hepatocellular loss involving 20% of the hepatic parenchyma with no significant inflammation or fibrosis consistent with toxic injury as well as treated AIH. Since the patient was already treated with corticosteroids, differentiation between AIH and AOSD hepatic injury was not possible. Serologic work up was negative for AIH, and the diagnosis was presumed to be AOSD-associated liver injury. Anakinra was added to her treatment regimen which led to dramatic improvement in liver function.

Our case demonstrates that AOSD-associated liver injury which is refractory to corticosteroid treatment can be treated successfully with anakinra. Further study is needed for clarification of histologic features of AIH versus AOSD which will aid in choosing appropriate and targeted therapy in patients who are refractory to corticosteroids.
A Case of Severe Alpha-1 Antitrypsin Deficiency

Introduction: Alpha-1 antitrypsin (AAT) deficiency is a relatively common, yet underdiagnosed, genetic condition which can lead to early-onset emphysema and liver disease. AAT is the prototypic member of the serine protease inhibitor (serpin) superfamily of proteins, and a deficiency leads to unregulated activity of multiple proteases. In the lung, alveolar damage is caused by a multifactorial process.

Case Presentation: A 30 year old man presented with acute COPD exacerbation, with rapidly worsening of respiratory status with increasing home O2 requirement. Medical history was significant for smoking and asthma with tracheomalacia. He had limited work exposure as installing home sidings and denied bird exposure. Past work-up was performed to identify the cause of early-onset COPD. At that time, CT chest revealed diffuse moderate to severe emphysema with slight lower lobe predominance. Pulmonary Function Testing showed obstructive pattern. Genetic testing demonstrated PiZZ, indicative of homozygosity for the Z allele. Bronchodilators and systemic steroids were started, and he was placed on Alpha1-proteinase inhibitor with enzyme replacement therapy infusions. However, his lung disease continued to progress, and he eventually required Bilateral Orthotopic Lung transplant (BOLT), with accompanying immunosuppression and prophylaxis.

During the hospitalization, he had extensive complications, including bleeding, prolonged respiratory failure requiring tracheostomy, ECMO, cannula-related bacteremia, and tracheostomy-related issues, all made worse by presence of childhood-tracheomalacia. His poor prognosis was discussed with the family and the decision was made to initiate comfort care measures.

Discussion: Current estimates place the prevalence of AATD at 1 in 2000 – 5000 in the US. Since it can result in early-onset emphysema, the American Thoracic Society recommends genetic testing for AATD in all symptomatic adults with emphysema, COPD, or asthma with airflow obstruction that is incompletely reversible after aggressive bronchodilator treatment. Early detection can prevent progression to severe emphysema and lung parenchymal destruction by treatment with intravenous infusion of purified pooled human plasma AAT, also known as IV augmentation therapy.
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Paraneoplastic Dermatomyositis as the Presenting Symptom of Intrahepatic Cholangiocarcinoma

Introduction: Paraneoplastic syndromes (PNS) are a heterogeneous group of disorders with varying incidence based on tumor type. For solid tumors overall, the incidence of neurological paraneoplastic syndromes is far less than 1%. Cholangiocarcinoma associated para-neoplasia are rare with only a handful of cases noted in the literature.

Case: A 34-year-old male presented with a 1month history of diarrhea, weakness, and 75lb weight loss. Associated symptoms included upper and lower extremity weakness, right abdominal pain and nausea. On examination the abdomen was mildly tender in the right upper quadrant and neurological examination was normal. Laboratory tests/studies demonstrated INR 1.2, CK 1,436 U/L, and CA19-9 2,327 U/ml. Computed tomography of the abdomen showed numerous hypodense liver lesions. Abdominal magnetic resonance imaging (MRI) revealed peritoneal carcinomatosis. Liver biopsy was consistent with moderately differentiated adenocarcinoma, immuno-stains favored stage 4 intra-hepatic cholangiocarcinoma. On physical exam, on hospital day 6, he was noted to have proximal muscle weakness +3/5 in the upper and lower extremities, 5/5 strength in the distal muscles, all reflexes were 2+, he was unable to stand or ambulate. Electromyogram and nerve conduction studies (EMG/NCV) demonstrated proximal myopathy. At this time a fleeting purplish rash was noted on his arms, legs and trunk. He was started on high dose steroid therapy for a paraneoplastic dermatomyositis. Myositis antibody panel was negative. On hospital day 8, he became diaphoretic and lethargic. He was found to be bradycardic with HR in the 30s-40s and normotensive. EKG demonstrated high degree AV block. A cardiac catheterization ruled out coronary artery disease, and he required placement of a permanent pacemaker for paraneoplastic heart block. Patient was started on palliative chemotherapy and discharged home. Unfortunately, he had 2 repeat admissions for worsening muscle weakness unresponsive to steroids and became bed-ridden. Patient passed away less than 2 months after initial cancer diagnosis.

Discussion: Paraneoplastic syndromes associated with solid tumors are rare and carry a poor prognosis. Dermatomyositis is a clinical diagnosis; many patients are negative for antibodies. As up to 30% of dermatomyositis cases are associated with malignancy, a thorough evaluation is essential with history and physical exam, age-appropriate screening, and CT scan of chest/abdomen/pelvis in high risk patients. Treatment is directed at the underlying tumor followed by glucocorticoids or IVIG to treat the paraneoplasia myositis and cutaneous changes.

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A RARE CAUSE OF PERSISTENT DYSPNEA
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Clinically amyopathic dermatomyositis (CADM) is a rare disease with little or no evidence of myositis but with the typical skin findings of dermatomyositis1. CADM has been associated with rapidly progressive interstitial lung disease (ILD), PE, or even malignancy that can be fatal. A 32-year-old Hispanic man with a history of morbid obesity presented with dyspnea on exertion for 2 weeks, with fatigue, cough, and wheezing. He had low grade fever (100.4°F), tachycardia (100 bpm), tachypnea (40/min), and was hypotensive (106/76) with no overt weakness. On physical exam, he had periumbilical erythema and Gottron’s papules on his elbows. On day 1, he required high-flow nasal cannula oxygen (HFNC) set at 50%/50L. A chest CT showed bilateral patchy infiltrates consistent with bilateral pneumonia and H. influenza A was positive. He was treated with oseltamivir along with azithromycin and ceftriaxone and started on CPAP for presumptive OSA given his obesity. Despite treatment, he had persistent dyspnea, hypoxemia, and tachycardia with oxygen requirements nadired at HFNC at 40%/10-15 L. On day 4, his symptoms prompted repeat imaging with a chest CTA which showed bilateral PEs and he was subsequently started on rivaroxaban. His labs were notable for WBC 14 and BNP 3757 which prompted an echocardiogram (TTE) that exhibited a normal RV, PA pressure 55-60 mmHg, and EF 55-60%. The patient was started on daily diuretics with some improvement. TTE was repeated after diagnosis of PE and was notable for new right heart strain without evidence for LV dysfunction. On day 7, his dyspnea and O2 requirement persisted, and a repeat chest CT showed almost no radiographic improvement in his infiltrates. Further work up was notable for elevated CK (~600), aldolase, and ferritin. Due to clinical suspicion for CADM, antibody testing was done which showed a positive anti-Ro52 antibody. He was then started on prednisone 60 mg daily and azathioprine and his oxygenation requirements improving shortly thereafter.

It is important to consider CADM in patients presenting with persistent dyspnea and imaging findings suggestive of ILD, despite treatment of the more common causes of dyspnea, including pneumonia and PE.
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Esophageal Perforation: A serious complication of black esophagus

Introduction
Black esophagus, also known as Gurgit's syndrome, necrotizing esophagitis, or acute esophageal necrosis, is an uncommon clinical entity. Most patients present with hematemesis, coffee-ground emesis, or melena, accompanied by dysphagia, epigastric pain, chest pain, lightheadedness, or syncope. It has a high mortality largely due to severity of the underlying disease, and carries a poor prognosis.

Case Description
A 69-year-old woman with morbid obesity, hyperlipidemia, and recurrent urinary tract infections presented with shortness of breath after episodes of non-bloody vomiting and diarrhea. She also had right-sided pleuritic lower chest pain radiating to the back. Afebrile, normotensive at 119/96, tachycardic at 112, tachypneic and hypoxic, she initially required oxygen support through a nonrebreather mask, and ultimately intubation and ventilator support. Lungs were clear to auscultation. Chest radiograph showed pneumomediastinum with subcutaneous emphysema, and computed tomography of the chest additionally revealed pneumothorax, pleural effusion, and complete collapse of the right lower lung lobe, without discrete esophageal injury.

Esophagoscopy revealed nonbleeding severe esophagitis proximally, with diffuse circumferential black discoloration of friable esophageal mucosa with spontaneous bleeding in the middle to the distal third, consistent with acute esophageal necrosis. A perforation measuring 20 mm was found in the distal third of the esophagus. Mediastinal exploration and washout of the right chest were performed, followed by chest tube and esophageal stent placement. After 30 days, esophageal stent was removed. The patient also underwent decompressive gastrostomy and jejunostomy. However, due to chronic hypoxic respiratory failure, tracheostomy was required. After 3 months of hospitalization complicated by intrathoracic hemorrhage, pneumonia, and gastric venous fistula, she was eventually discharged.

Discussion
Perforation is a serious complication of black esophagus, which may lead to mediastinitis, mediastinal abscess, empyema, or sepsis. Because of the potential for rapid clinical deterioration, especially among patients with underlying comorbidities, awareness and prompt recognition of this condition are essential.

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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!
PERITHEPATITIS IN A MALE WITH GONOCOCCAL INFECTION. Foppiano Palacios C, MD; Kathari Y, MD. The University of Maryland Medical Center and Baltimore VA Medical Center, Baltimore, MD.

Disseminated gonococcal infections typically cause dermatitis, tenosynovitis, polyarthritis or oligoarthritis. In women, it can cause pelvic inflammatory disease (PID) and is associated with FitzHugh-Curtis syndrome. FitzHugh-Curtis syndrome is a form of perihapatitis that can cause liver capsule infection. Classically, it is described as causing violent-string adhesions around the liver, presenting with right upper quadrant pain in female patients with PID or salpingitis. However, perihapatitis from disseminated gonococcal infection has not been well described in male patients.

A 36-year-old HIV-negative man with a past medical history of tricuspid valve endocarditis, untreated hepatitis C, and poly-substance abuse presented with chills, generalized malaise, night sweats and subjective fevers. He had dysuria, but no other urinary symptoms or discharge. Urinalysis was notable for 3+ leukocyte esterase and greater than 50 white blood cells; urine culture grew 20,000 colony forming units of Neisseria gonorrhoeae. He was treated empirically for gonococcal and chlamydial infections with ceftriaxone 250 mg intramuscularly once and azithromycin 1g orally once.

He developed right upper quadrant (RUQ) abdominal pain, and worsening transaminitis during his hospitalization. His baseline from one month prior to admission showed aspartate aminotransferase (AST) 76 units/L, alanine aminotransferase (ALT) 66 units/L, and alkaline phosphatase (ALP) 134 units/L. Treatment, 4 days later revealed peak AST 259 units/L, ALT 284 units/L, and ALP 211 units/L. CT abdomen and pelvis showed diffuse hepatic steatosis and a 1.8 cm ill-defined hypodense lesion in segment seven of the liver. MRI of the abdomen showed no evidence of malignancy or cirrhosis. Diagnostic studies did not yield any other etiology for hepatic inflammation, including hepatitis A and B, RPR was nonreactive, and blood cultures remained negative. He was started on ceftriaxone 1g intravenously every 24 hours for the presumed diagnosis of disseminated gonococcal infection causing perihapatitis. This was evidenced by RUQ abdominal pain and rising transaminitis, both of which began to improve with initiation of ceftriaxone.

Although perihapatitis in a male with gonococcal infection is rare and not well described, it is important to include in the differential diagnosis of RUQ abdominal pain seen with transaminase elevations in a patient with recent sexually transmitted infection.
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**Immune checkpoint inhibitor induced central adrenal insufficiency**
Kaleb Lambeth MD, Beatriz Wills MD, Susan Lin MD. Department of Medicine, The Johns Hopkins Hospital, Baltimore, MD

A 57-year-old gentleman with a past medical history significant for melanoma treated with ipilimumab and nivolumab. He was undergoing immunotherapy for roughly 3 months prior to developing a diffuse headache associated with nausea and vomiting. His oncologist prescribed a course of anti-emetics, which were ineffective. He also received a course of dexamethasone out of concern for intracranial metastases causing increased intracranial pressure. Although he experienced symptomatic relief with steroids, they were discontinued when subsequent brain MRI was negative for metastatic disease. Shortly after discontinuation of steroids, his symptoms returned. After two weeks of ongoing nausea and vomiting, he presented to our emergency department. His initial vitals were significant for tachycardia and orthostatic hypotension. His physical exam was notable for dry mucus membranes and an inguinal mass. Initial labs were significant for hyponatremia and hypochloremia. Early morning cortisol was low, ACTH was low, and his serum cortisol responded to ACTH stimulation. A pituitary protocol Brain MRI was obtained and significant for a subtle hypoenhancing lesion within the pituitary; otherwise unremarkable. He was diagnosed with central adrenal insufficiency thought to be secondary to hypophysitis caused by his immune therapy. He was treated with intravenous dexamethasone, and his symptoms resolved. He was discharged with oral hydrocortisone, and on follow-up his symptoms were controlled.

Immune therapy for the treatment of cancer is a relatively new option for various malignancies. In 2011, the FDA approved Ipilimumab for the treatment of metastatic melanoma. Ipilimumab is a cytotoxic T-lymphocyte associated protein-4 (CTLA-4) inhibitor. CTLA-4 is a surface receptor on T cells that is upregulated by activated T-cells and is a key receptor in transmitting inhibitory signals to dampens immune activation. Nivolumab is a PD-1 inhibitor which can also be used to treat melanoma. PD-1 is a receptor on the surface of T cells that is upregulated after prolonged activation, once it binds to its receptor it also dampens the immune response. Immune related adverse effects can affect any organ system, most commonly it effects the skin. However, Hypophysitis is characteristic of ipilimumab, and occurs with a well-defined time course. Endocrinopathies typically appear after the 6-7th week and are often irreversible.

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MYOCARDIAL INFARCTION WITHOUT CORONARY ARTERY DISEASE
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Myocardial infarction with no obstructive coronary atherosclerosis (MINOCA) is defined as having evidence of an MI with ≤ 50% stenosis of coronary arteries and has a prevalence of 6% among patients found to have acute MI. While there are numerous causes of MI, two of the rare causes are coronary artery embolism and thrombophilia.

A 70-year-old African-American woman with history of bilateral breast cancer status post radiation therapy and pulmonary embolism (PE) not on anticoagulation, presented to the ED with chest pain. EKG showed ST elevation in the inferolateral leads. Troponin level on arrival was 2 and peaked at 128 ng/mL 24 hours after arrival. The patient went for emergent cardiac catheterization and was found to have a wrap-around LAD, with 100% occlusion of the apical LAD, apical hypokinesis, and a left ventricular ejection fraction of 30% All other arteries had minimal non-obstructive disease. Despite the finding of the apical LAD obstruction, due to non-obstruction of most of the arteries with <50% stenosis and the patient’s history of PE, the diagnosis of MINOCA was made, secondary to coronary artery embolism. TTE with bubble study was negative for any intracardiac shunt. A holter monitor was placed to evaluate for paroxysmal AF and did not show any evidence of arrhythmia. Due to the patient’s history of recent PE and now embolic STEMI, she underwent a hypercoagulability workup which demonstrated normal Factor V Leiden, but an elevated Factor VIII activity of 354 (normal 55-175). She was discharged on long-term anticoagulation. At three-month follow up, the patient had a further drop in EF to 20% on echocardiogram, and considering multiple episodes of ventricular tachycardia, required ICD placement.

While factor V Leiden is the most common cause of thrombophilia, multiple studies have been done demonstrating that an increase in factor VIII levels is also a cause of thrombosis, rather than a consequence. Further, an association has been found in the African-American population, specifically in obese women. This case highlights the potential that MI may have causes other than coronary atherosclerosis, and a hypercoagulable workup should always be kept in mind.
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Trichosporon beigelli: A Rare Case of Fungal Septic Arthritis

Introduction: Septic arthritis is an important consideration in the differential diagnosis of swollen joints. The diagnosis cannot usually be made solely based on the history and physical examination. Laboratory findings such as leukocytosis, elevated inflammatory markers and synovial cell count often aid clinicians in making the diagnosis, but the lack of sensitivity and specificity often make these studies unreliable. Here, we present a rare case of fungal septic arthritis.

Case: A 68-year-old female presented with a one-week history of left knee pain and swelling. The pain was 8/10 in severity, constant, non-radiating, throbbing and worsened with movement. She denied any trauma, recent intra-articular injections or previous surgical interventions. Functionally, the patient was independent with all activities of daily living and used a wheelchair for mobility outside of her house. On physical examination she was febrile and tachycardic, the left knee was not erythematous, but it was warm to touch, tender to palpation and an effusion was present. Erythrocyte sedimentation rate and c-reactive protein were elevated. Arthrocentesis and synovial fluid analysis revealed 35cc of straw-colored fluid, WBC 37.8K with 96% polymorphonuclear cells and no crystals. Left knee MRI showed a complex loculated effusion with septations. Synovial culture grew Trichosporon beigelli. Patient underwent arthroscopic irrigation and debridement. She was discharged to sub-acute rehab with improvement in her symptoms, she completed 4 weeks of antifungal therapy.

Discussion: Septic arthritis is largely caused by bacteria, Staphylococcus aureus is the most commonly isolated organism. Fungal infections account for <1% of cases with the most commonly reported organism being Candida albicans. Fungal arthritis is primarily seen in immunocompromised patients and is difficult to diagnose and treat promptly given its indolent course. In a 20-year retrospective study by Girmenia et al. Trichosporon beigelli was reported in 10 out of 52 cases of septic fungal arthritis. These infections occurred almost exclusively in immunocompromised patients such as those with hematological malignancies, on chronic steroids or undergoing chemotherapy. To our knowledge, this is the first case of fungal septic arthritis caused by Trichosporon beigelli in a non-immunocompromised patient. Azoles are the mainstay treatment for Trichosporon as demonstrated by in vitro studies when compared to amphotericin B. The cure rate is estimated to approach 50-60% with the use of voriconazole.