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HYPERTRIGLYCERIDEMIA INDUCED ACUTE PANCREATITIS
Shashi Yalamanchili MBBS, Nicole Rapista MD, Arshpreet Kaur MD
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Management of acute pancreatitis caused by gall stones and chronic alcohol abuse focuses on goal-directed aggressive fluid hydration and pain control. However, there is no standard guideline that is currently in place for hypertriglyceridemia-induced pancreatitis. Insulin is one of the recommended treatment options for hypertriglyceridemia-induced pancreatitis with a goal of decreasing triglyceride (TG) levels to <500mg/dL.

A 34-year-old male was evaluated in the Emergency Department with 5-day history of worsening localized abdominal pain that was not associated with food intake. His past medical history was significant for hypertension and hyperlipidemia; however, he was non-compliant with medication or diet. At presentation, serum glucose was 516mg/dL with an elevated anion gap of 16 without urine ketones. He initially received 5 units of regular insulin in the ED and was started on insulin drip per hypoglycemia protocol. Lipase was noted to be five times the upper limit of normal. Contrast-enhanced CT of the abdomen showed localized pancreatitis. A lipid panel was then sent, which initially reported a TG level of >550mg/dL, which led to the suspected diagnosis of acute pancreatitis induced by hypertriglyceridemia. Endocrinology service was consulted. Triglyceride monitoring was done every 12 hours with the subsequent samples needing to be diluted 3 times because it was described to be very lipemic. Repeat TG levels were still reported as >1,100mg/dL and patient was continued on insulin drip even when the anion gap had already closed. Oral Gemfibrozil was planned to be started once the triglyceride levels were below 500mg/dL, however his levels remained elevated. His lowest recorded triglyceride was at 905mg/dL despite being maintained on the insulin drip for 3 days. His abdominal pain has significantly decreased and he was tolerating diet. It was then decided to discontinue the insulin drip and start him on gemfibrozil 600mg BID with close follow up with endocrinology as outpatient. His serum glucose levels were also addressed and controlled during this admission and he was discharged on metformin and insulin.

This case illustrates the challenges in managing hypertriglyceridemia-induced pancreatitis. Current literature suggests discontinuing IV insulin when TG levels are <500mg/dL; however, this was challenging to achieve in this case. Further studies are needed to determine if a strict TG goal is warranted during the initial management.

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SEQUELAE OF AN INFECTED VASCULAR GRAFT

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A 31-year-old woman with superior mesenteric artery syndrome (SMA) and a complex vascular surgical history presented with subacute fevers, chills, and a painful rash on her right foot.

She had previously been diagnosed with superior mesenteric artery (SMA) syndrome and underwent SMA transposition without symptomatic improvement. Two months prior to her presentation, she traveled to Germany for SMA reconstruction and underwent creation of a left femoral artery to femoral vein fistula with a graft for an intraoperative diagnosis of May-Thurner syndrome. After returning to the United States, she experienced one month of fevers, myalgias, and fatigue. Two days prior to her presentation, she developed a painful rash on her right foot.

On physical exam, she had numerous tender, erythematous, raised nodules on the plantar surface of her right foot. She had associated paresthesia but was neurovasculatly intact. On the remainder of her skin exam, she had no nodules on her fingers, painless lesions, or track marks. Blood cultures grew methicillin-sensitive Staphylococcus aureus. Transesophageal echocardiogram revealed a 0.5 x 0.5 cm vegetation on the mitral valve. Her computed tomographic scan of her chest revealed numerous cavity lesions. She was treated with oxacillin but remained persistently bacteremic for more than 5 days prior to culture clearance, prompting a nuclear tagged white blood cell scan which showed significant radiotracer uptake in the femoral arteriovenous (AV) fistula. She was taken to the operating room and was found to have a grossly purulent AV fistula, which was debrided and ligated. Her blood cultures subsequently cleared and she was discharged to complete a six week course of oxacillin.

The nodules on the patient’s right foot are consistent with Osler’s nodes. These thought to be septic microemboli that deposit in the skin, leading to the formation of dermal microabscesses. Osler’s nodes are usually associated with subacute and acute infective endocarditis but may also occur distal to an infected vascular graft. Janeway lesions have similar pathophysiology but are differentiated from Osler’s nodes by the lack of pain.

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An Unusual Presentation of Burkitt Lymphoma

Introduction: Burkitt lymphoma (BL) is a highly aggressive mature B-cell neoplasm with three distinct clinical forms: endemic, sporadic and immunodeficiency-associated. Clinical presentation varies depending on the specific form but can be as subtle as a case of numb chin syndrome (NCS) as the only presenting symptom.

Case: A 29-year-old, previously healthy male, presented complaining of generalized bony pain and myalgia for two months. He had relocated to the United States from Togo two months before symptom onset. One month before presentation he developed a gradually progressive, non-productive cough and decreased hearing on left ear with a feeling of fullness. Dental pain and hypoesthesia over his chin had developed more recently. Physical examination findings included: bulging tympanic membranes with tender tragus bilaterally, hypoesthesia over the chin and no palpable lymphadenopathy. Laboratory tests showed WBC 11.1 K/μL, platelets 22 K/μL, erythrocyte sedimentation rate 92 mm/h, Epstein Barr virus IgM antibody 65.2 U/mL. Lactate dehydrogenase >2,500 U/L and uric acid 12.8 mg/dL. Computed tomography of the head, chest, abdomen, and pelvis revealed multifocal pneumonia and bilateral mastoiditis. Appropriate antibiotic therapy was started but the patient failed to improve. Peripheral flow cytometry revealed immature lymphoid proliferation composed of monoclonal B-cells expressing CD19, CD20, and kappa light chain. CSF examination was negative for malignant cells. Bone marrow biopsy revealed a hypercellular bone marrow and CD10 positive B-cell lymphoma. IGH-MYC t(8;14)(q24;q32) fusion gene by FISH was positive in keeping with BL. The patient was started on hyper-CVAD regimen. One week after the initiation of the regimen, bone marrow biopsy was repeated and showed 90% resolution of the malignant clone. The patient is currently under treatment with the same regimen.

Discussion: NCS, also called mental nerve neuropathy, can occur from a lesion anywhere along the course of the trigeminal nerve. Among the causes of the NCS is the presence of malignancy. In a systemic review, 21% of the patients with NCS secondary to malignancy were found to have BL. There have been previous clinical and histopathologic reports about the association between leukemia and hearing loss, facial palsy, and ear infections. However, clinical presentation of BL with otologic symptoms is unusual.

Conclusion: NCS as the initial presentation of BL is not uncommon. This case report aims to bring awareness about unusual presentations of BL, as these are misleading and may delay the correct diagnosis and proper treatment, thus promoting disease progression.

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Title: Retrospective NSCLC in the setting of Neuropathy and Calcium Channel Antibodies

Introduction: The presence of P/Q subtype of Voltage Gated Calcium Channel Antibody (VGCC-Ab) is a known association of Lambert-Eaton Myasthenic Syndrome (LEMS) in Small Cell Lung Cancer (SCLC). Although positive in various paraneoplastic and neurological disorders, there is little evidence of these channels and antibodies in Non-Small Cell Lung Cancer (NSCLC).

Case description: 71yo male was referred for elevated VGCC-Ab P/Q with a 3-year-long history of symptoms of neuropathy inconsistent with LEMS. He also described a 10-year-long history of non-productive cough, without hemoptysis. Never smoker, he admitted to diffuse arthritic pain but denied weight loss, night sweats, or masses. There had been occupational exposure to asbestos and bronze dust. His past medical history was significant for Hypertension, Hyperlipidemia and Atrial Fibrillation. He had a family history of lung cancer in both parents and pancreatic cancer in his brother, all of whom were smokers.

Vitals and Physical Exam (other pertinent labs) including hba1c were all within reference range. Over the course of multiple outpatient visits, a repeat VGCC-Ab was negative, however a CT scan revealed a left upper lobe lung mass correlating with a subsequent PET scan finding of intense hypermetabolism in the speculated lung mass and left hilar lymphadenopathy. He underwent mediastinoscopy and left upper lobectomy with biopsy resulting a moderately differentiated adenocarcinoma. The adenocarcinoma was staged as a T2a, N1, M0, Stage IIA; he received and tolerated four cycles of adjuvant chemotherapy with Carboplatin and Pemtrexed. He is currently being followed-up for targeted therapy harboring EGFR mutation.

Discussion/Conclusion: This case demonstrates the need for cautious interpretation of results for VGCC-Ab which has scope for associations beyond established norms, necessitating further investigation in seemingly asymptomatic presentations. Furthermore, the rare cribriform predominant histology, not currently included in the pulmonary adenocarcinoma classification systems, has been associated with the second highest proliferation rate and worst outcome for disease-free survival among all patterns. Interestingly, this pattern has not been associated with EGFR mutations.

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AN UNUSUAL CAUSE OF SPLENIC INFARCT

Introduction: Thrombotic thrombocytopenic purpura (TTP) is classically characterized by the pentad of fever, thrombocytopenia, hemolytic anemia, acute kidney injury, and neurologic dysfunction with associated decrease in circulating level of ADAMTS 13. We present a case of TTP without the classic pentad but rather with splenic infarct.

Case presentation: A 47 y.o African American woman with a history significant for TTP, SLE, DVT/PE initially presented with 5 days of abdominal pain, nausea and vomiting. She was diagnosed with viral gastroenteritis and treated symptomatically with improvement. Notably, the platelet count decreased from 120K on admission to 50K on day 3 of hospitalization. It subsequently rose to 95K on day 4, just prior to discharge, without intervention. Recurrent TTP was suspected, but it was considered to be unlikely due to rare schistocytes seen on smear. Though the LDH was over 900, the haptoglobin was elevated at 200 s. Troponin was mildly elevated at 0.37. ADAMTS13 level was sent prematurely and returned as <5% post discharge. Patient was advised of results by phone and mail. Unfortunately, patient was lost to follow up.

Patient was readmitted 1 month later with 3 weeks of sharp left upper quadrant abdominal pain with associated vomiting. She was again found to have elevated troponin (0.99) and LDH over 900, but only few schistocytes on smear. Of note, CT abdomen shows evidence of splenic infarct. Given patient’s thrombocytopenia, severe deficiency of ADAMTS 13, and evidence of organ ischemia (heart and spleen), diagnosis of relapsed TTP was then confirmed. Urgent plasmapheresis was initiated. After 6 sessions of plasma exchange and daily corticosteroid treatment, the patient’s clinical symptoms resolved, and both platelet count and the ADAMTS 13 activity normalized.

Discussion: This case illustrates an atypical presentation of TTP with thrombocytopenia but rare schistocytes, and nonspecific GI symptoms related to the splenic infarct. TTP can present with a wide spectrum of symptoms and without “classic pentad,” which makes prompt recognition of the disease challenging. Confirmation of low ADAMTS 13 is not necessary prior to initiating plasma exchange treatment if clinical suspicion is high, as early treatment is crucial for survival. This case highlights splenic infarction and abdominal symptoms as the major presenting symptoms in a patient with TTP.
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Title: When hoofbeats come from zebras: from hypercalcemia to Multiple Endocrine Neoplasia

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Multiple Endocrine Neoplasia I is a rare genetic disorder resulting from an autosomal dominant mutation of the MEN1 gene located on chromosome 11q13. This mutation results in a characteristic constellation of endocrine neoplasms, with the typical distribution involving the parathyroid, pituitary, and pancreas. However, given the range in presentation, diagnosis is often delayed.

Mr. A is a forty-two-year-old male with history of HLD, obesity, GERD, and recurrent non-functioning pituitary macroadenoma, who presented to the emergency department with weakness, fatigue, and muscle spasms. A week prior to admission, he underwent transphenoidal pituitary resection for recurrent pituitary adenoma, which had presented as bitemporal vision loss. Mr. A reported that he never returned to his baseline health, with persistent symptoms of weakness and fatigue, and was noted to appear slightly confused by his partner. His physical exam was notable for diffuse neurofibromas and an obese abdomen, with otherwise normal neurologic exam. Laboratory data was remarkable for calcium of 15. To further explore his hypercalcemia, a PTH level obtained was 457, suggesting primary hyperparathyroidism. The diagnosis was confirmed with a sestamibi scan, which identified three parathyroid adenomas. Upon further questioning, the patient recalled that he had a remote history of parathyroid surgery over 20 years ago, with no subsequent follow-up with Endocrine. During his hospitalization, he endorsed severe reflux, despite being on aggressive suppressive therapy. Given presentation of two of the three classically involved organs of MEN1, a gastrin level and CT scan were ordered. Gastrin resulted at 15,509, with normal levels less than 20, and a large 11 cm pancreatic mass was discovered on CT imaging. A fine needle aspirate of the pancreatic mass was obtained with histologic staining confirming diagnosis of a pancreatic neuroendocrine tumor.

This case illustrates the diagnostic challenge of identifying an uncommon diagnosis when clinical manifestations are separated in points of time. It is important to continually question clinical presentations and humbly reassess our patients' pathologies. A single unifying diagnosis may manifest.
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Vasculitic Neuropathy Associated with Rheumatoid Arthritis, a case report

Case presentation: A 70-year-old woman with rheumatoid arthritis (RA) presented with bilateral lower limb numbness and left foot drop of 1-day duration. She noted generalized fatigue and small punctate lesions on her shins and forearm. For 20 years, she has been on 20 mg methotrexate and 5 mg daily prednisone. The methotrexate was discontinued 2 years earlier after undergoing a partial colon resection for perforated diverticulitis. Since then she has been on prednisone 10 mg followed by 7.5 mg daily. Her rheumatoid arthritis was considered under control. Her pertinent exam findings included small punctate lesions across the right foot. Small subcutaneous nodules across the left forearm, bilateral ulnar deviation, and mild synovial thickening of MCPs. Left foot dorsiflexion 0/5 strength, and absent left ankle jerk. The sensation was diminished on the dorsum of the left foot and lateral leg. An extensive workup for neuropathy revealed normal electrolytes, CBC, TSH, copper level, cryoglobulins, and vitamin B12. Her Lyme serology and SPEP were negative. MRI spine was unremarkable. Negative immunological studies. Elevated RF 585 IU/mL and C3 133.2 mg/dl. Low C4 8.4 mg/dl. Left Sural nerve biopsy revealed vasculitis, confirming the diagnosis of mononeuritis multiplex due to rheumatoid vasculitis. She was treated with methylprednisone 1 gram daily for 3 days, 60 mg prednisone taper and rituximab infusions 375 mg/m2 times 4. Her left foot dorsiflexion improved to 2/5 strength.

Discussion: The current incidence of rheumatoid arthritis is 1.3 million U.S adults.2 Rheumatoid vasculitis is a rare complication with 1% incidence rate. Most common are cutaneous manifestation; nail fold lesions, palpable purpura, and leg ulcers.3 Vasculitic neuropathy results in mononeuritis multiplex or distal symmetric sensory neuropathy.4 Less common manifestations are scleritis, pericarditis, aortitis, pulmonary vasculitis, and necrotizing glomerulonephritis.5,6,8 The development of extra-articular manifestations were associated with rheumatoid factor seropositivity.9 Seronegative rheumatoid vasculitis is rare.10,11,12 There are no current recommended guidelines for the treatment of rheumatoid vasculitis. One study demonstrated improvement with IV cyclophosphamide plus methylprednisone.13 However, rituximab plus corticosteroids were preferred over cyclophosphamide for neuropathic vasculitis.14,15,16 There were reports of a decrease in rheumatoid vasculitis with the increase use of methotrexate, corticosteroids, and DMARDs.17,18 However, the incidence of rheumatoid vasculitis increased with the use of biologics but decreased when hydroxychloroquine was combined with low dose aspirin.19 Further research is needed to evaluate the impact of immunosuppressive agents on rheumatoid vasculitis incidence.

Conclusion: Vasculitis in the setting of RA can be due to the primary disease or its treatment, especially in this new era of biologic pharmacotherapy.
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Myeloma Outside the Bone: Diagnostic Challenges in an Evolving Disease
Richard Ferraro, MD; Amanda Kagan, MD; Leo Luznik, MD
Multiple myeloma is the neoplastic proliferation of clonal plasma cells, accounting for 1.8% of all malignancies[1]. Myeloma traditionally leads to significant skeletal destruction and is limited to the bone marrow, though can rarely lead to soft tissue plasmacytomas and extramedullary disease, posing unique clinical challenges[2]. Here we present an interesting case of extramedullary disease representing diagnostic and treatment dilemmas for clinicians.

A 67-year-old female with a PMH of breast cancer treated 14 years prior to presentation now in remission, and IgA multiple myeloma status-post multiple lines of therapy (Beginning treatment 6 years prior to presentation) complicated by diffuse lytic bony lesions, now presents with acute-on-chronic severe right arm pain and swelling. The patient had noticed right arm pain for one month prior to presentation following a mechanical fall, with XR in the ED at that time exhibiting evidence of pathologic humeral head fracture. The patient went home and continued to experience right arm pain, with acute worsening of pain and swelling on the day of presentation. Exam was notable for erythema particularly at the shoulder, swelling with hardness to touch, and pain to palpation throughout the right arm. Ulnar and radial pulses were intact on Doppler. Given the patient’s findings, orthopedic and infectious disease teams were consulted for further management. Ultrasound and CT imaging were obtained exhibiting numerous lytic lesions, nondisplaced humeral head fracture, and extensive soft tissue masses of the deltoitd, axilla, and lateral chest wall leading to compression of axillary and proximal brachiocephalic veins. Given atypical growth for myeloma, soft tissue biopsy was obtained demonstrating anaplastic myeloma with extensive necrosis. The patient was subsequently treated with palliative radiation to the right upper extremity with symptomatic improvement.

Extramedullary myeloma remains a rare presentation, though the distribution of atypical growth patterns is rapidly changing as treatments allowing for longer disease courses evolve. It is important to maintain awareness of such presentations of myeloma, and potential complications that may be otherwise attributed to causes more often seen in normative disease progression. As exemplified here, upper extremity pain and swelling thought to be secondary to fall in the context already lytic bone may signify a more involved process. Knowledge of atypical presentations may help clinicians identify

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Non-ST Elevation Myocardial Infarction as a Presentation of Carbon Monoxide Poisoning.

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Carbon monoxide (CO) poisoning is a condition that can be caused by inhaling excessive amounts of CO emissions. Once inhaled, CO binds strongly to multiple molecules including cytochrome oxidase, cardiac myoglobin and hemoglobin, causing global ischemia and multiple organ damage. Cardiotoxicity in moderate to severe CO poisoning has been shown to cause arrhythmias, conduction blocks, and myocardial ischemia leading to infarctions.

An 86-year-old male with history of coronary artery disease (CAD), hypertension, hyperlipidemia, and atrial fibrillation on coumadin was transferred to the Emergency Room (ER) from an outside hospital (OSH) after he was found down next to a running tractor for an estimated time of 2 hours. At the OSH, the patient received cardiopulmonary resuscitation after a ventricular fibrillation arrest. Laboratory results at that time were significant for carboxyhemoglobin of 22.6%, lactate 7.9 mmol/L, and troponin of 0.05ng/mL. On arrival to the ER, the patient was sedated and required hemodynamic support with pressors. He was emergently taken for hyperbaric oxygen therapy. On repeat lab studies, the patient was found to have a rising troponin that peaked at 25.9 ng/mL. Electrocardiogram (EKG) showed atrial fibrillation and poor R wave progression, without evidence of ST elevations, depressions, or Q waves. A transthoracic echocardiogram (TTE) showed newly decreased left ventricular ejection fraction of 20-25%, as well as severely decreased right ventricular systolic function.

During the hospitalization, the patient underwent a cardiac catheterization that showed mild non-obstructive CAD with mildly elevated filling pressures. The patient was diagnosed with a non-ST segment elevation myocardial infarct secondary to CO poisoning and hypoxemia. He was eventually weaned off of pressors and extubated shortly after receiving hyperbaric oxygen therapy. A repeat TTE conducted 6 days later showed similar findings, now thought to be representative of a Takotsubo or stress cardiomyopathy. The patient was subsequently discharged to subacute rehab with appropriate follow up.

CO poisoning can cause global ischemia and can lead to myocardial infarctions. Early suspicion and active surveillance of cardiac injury can lead to quick identification of ischemic events; and use of standardized cardiac testing and may improve overall mortality and quality of life.
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Back to the basics: using the evidence-base and an unclothed exam to make a difficult diagnosis

A 21 y/o man with atopic dermatitis presented to the ED with chest pain and cough. Two months prior to admission he began experiencing burning, 7/10 chest pain at rest, exacerbated by coughing. A month prior he noted a non-productive cough with dyspnea and wheezing but no B symptoms. He was seen for a scheduled well visit with his pediatrician; exam was documented as normal save for sinus arrhythmia with plan for GERD treatment. When symptoms persisted his primary doctor scheduled a chest x-ray. He was instructed to go to the ED for evaluation of a right chest mass. In the ED he was afibrile, HR 140, RR 16, BP 141/90 and O2 sat 99% on RA. WBC was 11.8 and lactate 3.5. CTA revealed a large anterior mediastinal mass, multiple nodal masses in the upper and lower mediastinum and along the anterior cardiac surface, and postobstructive right upper lobe consolidation. An unclothed exam on admission revealed tachycardia, summation gallop, double impulse on PMI, diffuse bounding carotid arteries, right chest wall tenderness to palpation, and diminished breath sounds in the RLL. He disclosed an extensive family history of lymphoma. Core biopsy and flow cytometry revealed primary mediastinal (thymic) large B-cell lymphoma. He was discharged with oncology follow up with plan for genetic work-up.

Discussion:
Per CHEST guidelines, the workup for non-infectious subacute cough includes investigation of the most common etiologies. As such appropriateness criteria discourage routine CXR without clinical concern for a cardiopulmonary process, concordant with the PCP's initial evaluation and triage. When the cough persisted for a month and clinical suspicion heightened, he appropriately ordered a CXR instrumental to the final diagnosis. It is hard to know how the exam changed between encounters but a gown exam with careful evaluation of PMI and chest wall is a critical step in evaluation of a presumed cardiopulmonary process in the outpatient setting. A hyperkinetic apical impulse alone increases the probability of a mediastinal abnormality.

In the outpatient setting it is important to utilize evidence-based guidelines as a means for appropriate triage especially in relatively rare disease processes. Additionally a thoughtful, unclothed physical exam with attention to the chest wall and palpation of the point of maximal impulse is critical component of any cardiopulmonary exam, in this case assisting with a rare, but important diagnosis.

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ACUTE ENCEPHALOPATHY FROM HUMAN METAPNEUMOVIRUS
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Background
Human metapneumovirus (hMPV) usually causes an infection of the nasopharyngeal tract. It has rarely been associated with encephalitis which has mostly been reported in young children, elderly and those who are immunocompromised. However, studies show that there is extensive exposure to human metapneumovirus in the global population, with variation in both clinical symptoms as well as severity. This case report provides evidence hMPV can lead to encephalitis.

Case Presentation
A 52yr old man presented with altered mental status. He had cough, rhinorrhea, and chest congestion for the previous four days and back pain on the morning before admission. He had just finished a 3-day course of ciprofloxacin which did not improve his symptoms. He had no recent travel history. During the diagnostic evaluation, hMPV was detected from a nasal swab. Cerebrospinal fluid (CSF) analysis showed the absence of pleocytosis and marginally elevated glucose and protein. No organisms were seen on gram stain and culture of the CSF. Both midazolam and dexmedetomidine were used to control his agitation. By day 5, his neurological status improved without any sequelae, and he was discharged home.

Discussion
Case reports of hMPV causing encephalitis are infrequent, likely due to the lack of diagnostic capabilities. Due to the potential severity of infection with hMPV, availability of treatment options is crucial. So far, studies have shown that neutralizing human monoclonal antibodies may be an ideal treatment for those with severe disease such as encephalitis.

Conclusion
hMPV is a cause of acute upper and lower respiratory infection. However, it may cause encephalitis in adults as well as children.

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A Rare Malignancy in Neurofibromatosis Type 1

Introduction: Neurofibromatosis type 1 (NF-1) is known to be associated with increased risk of malignancy by at least fourfold. Most commonly seen are nervous system tumors like optic gliomas, neurofibromyosarcomas, and leukemia. Malignant lymphomas are extremely rare in adults. Hereby we present a rare case of diffuse B cell lymphoma in a patient with NF-1 since childhood.

Case presentation: A 75-year-old male with NF-1 presented complaining of generalized weakness, nausea and vomiting for 3 months. Review of systems was positive for unspecified weight loss, anorexia, chest pain due to recent trauma and progressive increase in the number of neurofibromas on the anterior and posterior trunk. Pertinent physical examination findings included: a soft mass on the umbilical area which was tender to palpation, with clear margins, and measured about 8x4 cm. In addition, he had multiple, small sized, flesh colored dome shaped fibromas clustered on the trunk, abdomen and upper extremity. Laboratory demonstrated WBC count 20.0 × 103/μL with predominant neutrophils, calcium 16.2 mg/dl, PTH 9.5pg/ml, PTH related protein 2.6pmol/ml and vitamin D level 15.9ng/ml. A computed tomography (CT) scan of abdomen showed a dominant oval shaped soft tissue density in the middle of upper abdomen quadrant superficial to the abdominal aorta. CT-guided biopsy of a retroperitoneal lymph node was consistent with diffuse large B-cell lymphoma (DLBCL). Bone marrow aspirate was negative. He was started on treatment with chemotherapy (CHOP). He tolerated the first cycle well with no major side effect and was discharged to rehabilitation program.

Discussion: NF-1 is known to be a significant risk factor for malignancy, mainly the peripheral nerve sheath tumors, gliomas and leukemia. The occurrence of malignant lymphoma is a rare phenomenon. In a literature review, we found only 16 cases where NF-1 co-existed with lymphomas, three cases were B-cell type, the rest were T cell type. This was an unexpected presentation as patient was asymptomatic and such association is rare as stated above. This case served as a reminder to always keep a broad differential diagnosis, independent of common associations and age groups.

Conclusion: This case emphasizes the importance of having a high index of suspicion for malignancies during follow up of patients with a history of NF-1. It is also important that during follow up, a multisystem approach should be implemented for diagnosing malignancies early. Lymphomas when diagnosed early carry better prognosis than carcinomas and sarcomas.

Indicate your participation in research process (4 sentences or less):
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SEBACEOUS ADENOMAS AND COLORECTAL ADENOCARCINOMAS; A RARE CASE OF MUIR-TORRE SYNDROME
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Introduction: Although 140,000 new cases of colon cancer are diagnosed annually in the United States, only 5-10% are suspected to be due to heritable mutations. Muir-Torre syndrome (MTS), a subtype of Lynch Syndrome, is a rare presentation of a heritable cancer syndrome involving cutaneous and visceral malignancies. First discovered in 1967, MTS is now known to involve loss of the mismatch repair genes. Inheritance is autosomal dominant, but spontaneous presentations exist. Here we report a unusual case of Muir-Torre syndrome in a patient without any family history of malignancy.

Case: A 49-year-old man without prior medical history or family history of malignancy presented to his primary care physician due to a new skin growth in the right groin. Biopsy revealed a sebaceous adenoma. A year later, the patient presented with symptoms of early satiety, 10lb weight loss, and new onset bright red blood per rectum. Colonoscopy revealed a 2.5cm rectal adenocarcinoma and esophagogastroduodenoscopy demonstrated a poorly differentiated adenocarcinoma extending from the distal esophagus to the gastric cardia. Neo-adjuvant cisplatin, 5-flourouracil, and radiotherapy were given, and the patient subsequently underwent surgical resection of both masses. Screening computerized tomography of the abdomen at age 51 revealed a suspicious liver lesion, and biopsy demonstrated metastatic adenocarcinoma. The patient received radioablative therapy and did well. At age 56 he was diagnosed with a right colon adenocarcinoma and underwent right hemicolectomy. At age 64, the patient was diagnosed with 2 new sebaceous adenocarcinomas and at age 65 the patient suffered a large bowel obstruction due to a T3N1 adenocarcinoma at the site of his colonic anastomosis. Immunohistochemistry for mismatch repair protein was performed and demonstrated loss of expression of mutS homolog 2 (MSH2) and 6(MSH6), carcinogenic mutations responsible for MTS.

Discussion: Muir-Torre syndrome is a rare cancer syndrome that predisposes patients to visceral and cutaneous malignancies. MTS requires clinical vigilance for sebaceous adenomas, and when found should prompt screening for defective mismatch repair genes. Patients should be referred to genetic counselling once MTS is diagnosed. Long term survival remains good as many of these malignancies associated with MTS may be less aggressive and may respond well to treatment.
A Case of Myasthenia Gravis and Myocarditis after Pembrolizumab Treatment

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Introduction: Checkpoint inhibitors are a class of drugs that modulate the immune system to help fight cancer cells and are used to treat a growing list of cancers including melanoma, non-small cell lung cancer, bladder cancer, and Hodgkin’s lymphoma. Common side effects include fatigue, loss of appetite, and skin rash. Rarely, these drugs can also cause side effects that mimic autoimmune conditions such as hypothyroidism, myasthenia gravis, and type 1 diabetes. The prompt diagnosis and management of these immune-related adverse events are essential to prevent morbidity and mortality.

Case Description: A 79-year-old man on pembrolizumab (checkpoint inhibitor) as second-line treatment for his metastatic bladder cancer presented to the hospital for 1 week of diplopia, ptosis, hypophonia, dysphagia, and proximal weakness. He underwent a head CT and MRI with no evidence of CNS metastasis or stroke. On exam, his ptosis was fatigable. Antibodies (AChR, MUSK) and repetitive nerve stimulation were negative. However, single-fiber EMG was consistent with myasthenia gravis. CT imaging of the chest did not demonstrate a thymoma. Modified barium swallow study showed impaired swallow and he was kept NPO. He was diagnosed with immune checkpoint-inhibitor (ICI) related myasthenia gravis and treated with IVIG and pyridostigmine. His symptoms improved except for dysphagia and a PEG-tube was placed.

On admission, he was also noted to have elevated troponin and a new right bundle branch block on ECG. He had no cardiac history and denied cardiac symptoms. TTE was unrevealing with a normal EF, but a subsequent cardiac MRI showed subendocardial delayed enhancement at the apex, consistent with ICI-associated myocarditis. He received high dose steroids followed by a taper. His cardiac biomarkers downtrended and he had no evidence of other arrhythmias.

Discussion: This patient was found to have not one but two rare side effects related to his cancer treatment with a checkpoint inhibitor. With prompt recognition and treatment, he avoided the life threatening complications of MG (respiratory failure) and myocarditis (arrhythmia). Unfortunately, he still had dysphagia reminding us that although rare, the side effects of these drugs can lead to significant morbidity. As the use of immune checkpoint inhibitors like pembrolizumab become more prevalent in cancer treatment, we must be aware that any symptoms occurring during or after treatment may be a potential side effect.

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!
AORTIC ROOT ABSCESS WITH AORTA TO RIGHT ATRIUM FISTULA DUE TO VANCOMYCIN-INTERMEDIATE STAPHYLOCOCCUS AUREUS (VISA). Hankinson, S MD, Sultanik, E MD. University of Maryland Medical Center and Baltimore VA Medical Center.

Aortic valve fistula (AVF) is found in 1.8% of cases of native valve aortic valve endocarditis (AVE) and 3.5% of prosthetic valve AVE. The overall mortality rate of AVF due to endocarditis is 41%.

A 74-year old man with a history of below the knee amputation (BKA) of the left leg due to methicillin-resistant Staphylococcus aureus (MRSA) and subsequent coronary artery bypass graft, aortic valve replacement (AVR), and mitral valve replacement 5 days after limb amputation presents with MRSA bacteremia. Of note, post-operative course was complicated by a prolonged course of vancomycin for a left stump surgical site infection. Transesophageal echocardiogram (TEE) at that time was negative for AVE. He was treated with a 6-week course of vancomycin; however, he again presented 5 days after completing the vancomycin course with VISA bacteremia. Transesophageal echocardiogram (TEE) at that time showed an aortic root abscess, left to right shunting, and a mobile 2.4 cm x 1.9 cm echodensity projecting into right atrium. He was diagnosed with an AVF, treated with daptomycin, ceftioroline, and rifampin, and underwent redo AVR and patch repair of aorta to right atrial fistula.

Upon reviewing the literature this appears to be the first case to discuss AVF formation due to VISA. VISA is a relatively rare infection that occurs in the setting of heavy prior use of vancomycin. It is difficult to identify exactly when the bioprosthetic aortic valve was seeded. Perhaps the valve annulus was seeded during implantation of the bioprosthetic aortic valve or perhaps the BKA stump cellulitis lead to hematogenous seeding of the bioprosthetic aortic valve. This case also demonstrates that TEE is the superior imaging modality since it allows for the detection of almost all fistulas and allows for optimal characterization of each fistula tract.

AVF formation and VISA bacteremia are both relatively rare conditions. Per review of the literature, this is the first case to discuss AVF formation due to VISA. TEE, as demonstrated by this case, continues to be the preferred imaging modality to diagnose fistulas. Clinical suspicion for AVF should be high in patients presenting with new murmurs, persistent bacteremia, and hemodynamic collapse. Moreover, the emergence of multidrug resistant organisms, like VISA, makes medical management of AVE increasingly difficult and must be considered in patients with a prolonged course of antibiotics.

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Case of Late Onset Systemic Lupus Erythematosus

Introduction: Systemic lupus erythematosus (SLE) is a chronic autoimmune disease. It is typically seen and diagnosed in younger patients; specifically females aged 15-45 years. SLE is uncommon after the age of 50. We present a case of SLE diagnosed in an elderly female.

Case: A 61-year-old Hispanic female with Raynaud syndrome for 1 year (not on any medication) and post-surgical hypothyroidism presented with dizziness, diaphoresis and more prolonged bluish discoloration of the digits upon exposure to cold (as compared to her baseline) for 2 days. She complained of dry eye for last 2 years. She denied dry mouth, rash, pleuritic chest pain, photosensitivity and any joint swelling. No history of miscarriage or thromboembolism. Physical examination revealed normal vital signs and unremarkable cardiovascular, respiratory, gastrointestinal and neurological systems. The digits in her hands were severely cyanotic, she had livedo reticularis in her hands and feet; radial and pedal pulses were intact. Laboratory workup showed hemoglobin 19.3 g/dl and anti-nuclear antibody titer of 1:320 with speckled pattern. At that point a diagnosis of Raynaud syndrome secondary to SLE with Sjögren syndrome made her. She was started on high dose prednisone, nifedipine and hydroxychloroquine. Her clinical condition remarkably improved and she was discharged. During outpatient follow up she was doing well, she was using heavy gloves to protect her hands and the cyanotic changes in her fingertips were in the process of healing. She was transitioned to mycophenolate mofetil as a steroid sparing agent.

Discussion: SLE is a severe, complex, multi-system autoimmune rheumatic disease. Late-onset SLE is defined as SLE diagnosed at or over the age of 50. There has been a lot of debate regarding whether late-onset SLE is associated with a more benign disease course and better prognosis than early-onset SLE. Pulmonary involvement and serositis are more frequently observed in patients with late-onset SLE, whereas malar rash, photosensitivity, arthritis, and nephropathy occur less commonly. Primary Sjögren syndrome usually affects patients older than those with SLE, with a mean age at onset of 52.7 ± 0.85 years and has a lower frequency of renal involvement, lymphadenopathy, and thrombocytopenia but has a higher frequency of Raynaud phenomenon. A study by Boodaert et al., performed from 1980 to 2000, reported that none of their 47 late-onset SLE patients died from SLE flares. Late onset SLE patients are more often positive for rheumatoid factor and negative for anti-RNP antibodies, anti-Smith antibodies, and low CH50 complement. Deaths secondary to the medications used to treat SLE are rarely mentioned in the literature. Age at SLE onset was previously reported as a risk factor for death. Despite the lower rate of major organ involvement and more benign course, late-onset SLE has poorer prognosis because older patients more frequently have significant comorbidities and higher organ damage due to aging and longer exposure to traditional vascular risk factors.

Conclusion: SLE is less prevalent in the elderly. SLE should be suspected in elderly patients who have atypical features of lupus. Multiple studies suggest late-onset SLE is not benign, reduced survival has been observed when compared to younger SLE patients.
HENOCH-SCHÖNLEIN PURPURA IN A PATIENT WITH CROHN DISEASE. Kathari Y, MD. The University of Maryland School of Medicine and VA Medical Center, Baltimore, MD.

Henoch-Schönlein purpura (HSP) is an IgA-mediated small vessel vasculitis that can involve the skin, gastrointestinal tract, joints, kidneys, and lungs. Classic presentations include erythematous macular lesions that progress to palpable purpura, especially in the lower extremities. HSP can also present with abdominal pain, bloody diarrhea, arthralgias, joint swelling, glomerular lesions, and rarely pancreatitis. Typically, a pediatric disease, HSP can be rarely seen in adults.

A 23 year-old female with past medical history of Crohn disease on adalimumab (until three weeks prior to presentation) presented with symptoms of odynophagia, worsening abdominal pain, bloody diarrhea and fevers for seven weeks. Three weeks prior to presentation, she was diagnosed with acute streptococcal pharyngitis by her outpatient provider and was prescribed a course of amoxicillin. At that time, adalimumab was held in the setting of fevers and active infection. On presentation, she was tachycardic, febrile, and hypotensive. She was started empirically on piperacillin-tazobactam and vancomycin. On physical exam, she had multiple oropharyngeal ulcers, diffuse abdominal tenderness, and small raised pruritic papules over her lower extremities bilaterally.

Her blood, throat and stool cultures were negative. Laboratory studies were negative for Streptococcus, Giardia, Clostridium difficile, Cytomegalovirus (CMV), and Herpes simplex virus (HSV). Respiratory viral panel was positive for rhinovirus and enterovirus. Computed tomography (CT) abdomen and pelvis showed acute inflammatory changes related to Crohn disease. An esophagogastroduodenoscopy (EGD) showed non-bleeding esophageal ulcers and erythematous gastric mucosa. Biopsies were taken, which were negative for CMV and HSV. Pathology showed small vessel vasculitis with fibrinoid necrosis and IgA testing is pending. Antibiotics were discontinued, and she received IV methylprednisolone for three days and subsequently transitioned to oral prednisone. Her symptoms, vital signs, and skin changes improved with steroid therapy.

HSP is a rare diagnosis in adults, however is important to include in the differential in a patient with abdominal pain, bloody diarrhea, and a new rash over the lower extremities. This case also demonstrates that patients with a pre-existing inflammatory disorder (Crohn disease), can have another concomitant inflammatory disorder (in this case, HSP).
A CASE OF CHLAMYDIAL CULTURE-NEGATIVE ENDOCARDITIS. Braunfeld J, MD. The University of Maryland School of Medicine and VA Medical Center, Baltimore, MD.

The exact incidence of culture-negative endocarditis is uncertain, although it may represent up to 50% of infective endocarditis cases. Most commonly caused by Bartonella and Coxiella species, cases of endocarditis due to Chlamydia species are far less frequently reported.

A 58-year-old male with history of atrial fibrillation, pulmonary embolism, and end-stage renal disease on hemodialysis presented with chest pain and shortness of breath for one week. The pain was primarily right-sided, sharp, and worsened with deep inspiration and the shortness of breath worsened on exertion. On admission, the patient was afebrile and his physical exam, was notable only for a mid-systolic murmur. Transthoracic echocardiogram (TTE) revealed a mobile echodensity on the mitral valve leaflet concerning for a vegetation. Blood cultures drawn on admission grew coagulase-negative Staphylococcus in 1 out of 2 bottles which was not speciated. Several repeat blood cultures, including one more set prior to commencement of intravenous antibiotics, were drawn throughout admission and all remained negative.

Transesophageal echocardiogram (TEE) on hospital day 4 revealed clear masses on the mitral and aortic valves that were interpreted as likely vegetations. After the procedure, the patient was noted to be febrile however he defervesced without intervention. He was again febrile the next day and chest X-ray revealed new opacities concerning for developing pneumonia after a suspected aspiration event during TEE the previous day. Despite adequate antibiotic coverage of Staphylococcus and aspiration pneumonia with vancomycin and piperacillin/tazobactam, respectively, the patient became febrile for a third consecutive day and his white blood cell count (WBC), which had climbed daily throughout admission, reached its peak of 38,200/µL.

Serologies for causes of culture-negative endocarditis revealed a C. pneumoniae IgG titer of 1:512 and a C. trachomatis IgG titer of 1:64 with negative IgM titers. Doxycycline was commenced on hospital day 9. Repeat TTE 2 weeks after discharge showed no evidence of vegetations and the patient was symptom-free at his clinic follow-up 1 week later. The patient received 8 weeks of Doxycycline for presumed culture-negative endocarditis from a Chlamydia species.

Chlamydia endocarditis is difficult to diagnose as Chlamydia serological assays have been shown to falsely result positive in the presence of antibodies to Bartonella species. However, given the absence of reactivity in all Bartonella assays performed, the diagnosis of Chlamydia infection in this case is much more secure.

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2019 Mulholland Mohler Resident Meeting

An atypical cause of back pain in a 31-year-old man
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Skeletal tuberculosis (or Pott’s disease) accounts for only about 2% of all tuberculosis cases in the United States. Pott’s disease is an uncommon cause of back pain in young adults.

A 31-year-old man presented to the emergency room with acute back pain after playing basketball. He was otherwise asymptomatic. He denied fever, chills, cough, hemoptysis, dyspnea, night sweats, abdominal pain, numbness, tingling, weight loss. Physical exam was unremarkable with clear breath sounds, no abdominal tenderness, negative psoas sign. No focal spinal or paraspinal tenderness. Back pain improved with cyclobenzaprine and ibuprofen, so he was subsequently discharged home from the ED. Just prior to discharge, he had a chest radiograph, which showed a possible consolidation in the right upper lobe. He was called back to the emergency room the following day where CT chest demonstrated a cavitory lesion in the right upper lobe eroding into the vertebral bodies with erosion/sclerosis concerning for osteomyelitis. He was born and raised in Baltimore and denied any travel outside of the city. Social history notable for 1 month incarceration approximately 10 years ago. No history of homelessness or housing insecurities.

CT of the abdomen and pelvis showed multiple retroperitoneal, pelvic, and gluteal abscesses. MRI spine confirmed multi-level vertebral osteomyelitis of the thoracic/lumbosacral spine in addition to paraspinal abscess formation. He underwent percutaneous drainage of the two largest collections in the iliopsoas and gluteal regions. This fluid was sent for mycobacterial culture and was later found to be positive for mycobacterium tuberculosis complex. He had three induced sputum samples that were AFB negative but positive on GeneXpert (NAAT probe assay) without rifampin resistance. He was started on RIPE therapy with pyridoxine and enrolled in directly observed therapy with the Health Department. Later transitioned to isoniazid and rifampin with plan for 1 year of total therapy given vertebral tuberculosis.

This case illustrates an atypical presentation of Pott’s tuberculosis in a young patient whose only risk factor was 1 month of incarceration over a decade ago. Additionally, he did not have any localizing signs or symptoms on history or exam. This case is a reminder for us to keep an open mind and a broad differential when evaluating our patients.

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CARDIAC MYXOMATOUS TUMOR WITH CONCERN FOR CARNEY COMPLEX. Khan Z, MD, Alkhatib H, MD, Ramani GV, MD, Gammie JS, MD. University of Maryland School of Medicine and VA Medical Center, Baltimore, Maryland.

Intracardiac tumors are rare entities, but amongst the most common are myxomas (~50% of tumors), which are gelatinous pedunculated tumors comprised of stellate or globular cells within a mucopolysaccharide stroma. Up to 75-80% of myxomas originate within the left atrium and can result in emboli, features of heart failure, or constitutional symptoms. Myxomas can be familial (~10%) in cases where facial freckles or other adnexomas are concurrently present.

Carney complex is an autosomal dominant condition where an inactivating mutation of the PRKAR1A gene responsible for protein kinase A (PKA) regulatory alpha-1 subunit results in multiple neoplasias (cutaneous, endocrine, cardiac) and facial nevi/spotty pigmentation. Compared to sporadic cases, such myxomas are diagnosed in earlier age and frequently recur.

A 54-year-old otherwise healthy man presented to his doctor with a two year history of non-productive cough and worsening dyspnea on exertion in recent months. Physical exam demonstrated multiple xanthomas and skin tags (ear and arms) and 2/6 blowing diastolic murmur at the left lower sternal border, but was not revealing of peripheral extremity edema, jugular venous distention, or crackles/rales on lung exam. A chest x-ray was ordered for further evaluation, which demonstrated concern from radiology of an intracardiac density. A computer tomography (CT) scan of the chest was ordered for further evaluation, which confirmed the finding of an intracardiac mass. A transthoracic echocardiogram revealed a 5cm x 1cm mass in an enlarged left atrium actively prolapsing into the left ventricle, as well as severe pulmonary hypertension. Electrocardiogram (EKG) was unremarkable apart from notched p-waves in lead II and inverted p-waves in lead V1; consistent with left atrial enlargement. A transesophageal echocardiogram was performed for further evaluation of the intracardiac mass, as well as corroboration of elevated right heart pressures. Patient underwent surgical excision of mass with subsequent recovery.

Following surgery, a genetics consult was ordered and genetic testing for PRKAR1A gene mutation (most common variant) was negative. Patient now remains asymptomatic post-excision of mass.

Cardiac tumors can often be incidental findings in asymptomatic patients, however can be familiar in others. There should be a high index of suspicion for workup of Carney complex in patients that have early identification of myxoma, cutaneous manifestations, or presence of extracardiac neoplasias.
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Thyrotoxic Periodic Paralysis in an African American Male

Introduction: Thyrotoxic periodic paralysis (TPP) is a channelopathy that manifests with muscle weakness. Thyrotoxic periodic paralysis is a sporadic form of hypokalemic period paralysis that is most common in Asian males. Hereby, we present a case of TPP in an African American male.

Case: A 31-year-old African American male presents with generalized muscle weakness more prominent in the proximal muscles in the upper and lower extremities. He was diagnosed with hyperthyroidism 6 months prior but was lost to follow-up. After one week at a bowling tournament he developed progressive, ascending muscle weakness that started at the calves and progressed to involve the proximal thigh muscles to the point that he was unable to stand from sitting position. The muscle weakness progressed to involve his upper extremities. He denied any other neurologic symptoms. Physical examination he had lower and upper extremity weakness 4/5. Laboratory investigations revealed 2.1mEq/L, TSH 0.005 mIU/L and FT4 4.96 4.96mg/dl. He was started on a beta blocker, methimazole 20mg daily and potassium was replaced. By the late afternoon on the first day of admission his symptoms had resolved, muscle strength had returned to 5/5 in all extremities after potassium repletion.

Discussion: TPP is most common in young Asian males with an estimated incidence of around 2%; the incidence of thyrotoxic period paralysis in non-Asian males is estimated to be 0.1-0.2%. Thyroid hormone promotes the activity of the sodium-potassium ATPase in skeletal muscle driving potassium into the cell, hyperpolarizing it and theoretically making the cell less excitable. Beta-agonists and insulin have a similar effect on sodium-potassium ATPase. In TPP a loss of function mutation in potassium channels in individuals with untreated or under-treated hyperthyroidism predisposes them to bouts of paralysis with excessive exercise and/or carbohydrate loads. Common laboratory findings in TPP include hypophosphatemia and hypomagnesemia; these findings are more common in TPP when compared to familial forms of hypokalemic periodic paralysis.

Conclusions: Physicians should be aware of this rare disease in all patients presenting with profound hypokalemia and laboratory evidence of hyperthyroidism. As demonstrated by this case, although rare, TPP can occur in individual without Asian decent.

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A RARE COMBINATION OF HEMOGLOBIN SC TRAIT AND SILENT THALASSEMIA CARRIER IN A PATIENT WITH SPLENIC AUTO-INFARCTION

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Introduction: Hemoglobin SC (HbSC) has a milder clinical disease course as compared to homozygous hemoglobin SS (HbSS). Co-inheritance of heterozygous alpha-thalassemia with HbSC, decreases intracellular hemoglobin S polymerization and dramatically reduces the risk of acute splenic sequestration, leading to prolonged survival with improved clinical course and resulting in delayed diagnosis of disease. Here we present a rare case of incidental finding of splenic auto-infarction leading to a diagnosis of HbSC trait with silent alpha thalassemia carrier state in a patient presenting with pneumonia.

Case: A 58-year-old male with previous history of pulmonary embolism presented with one month history of progressive dyspnea and cough. Significant laboratory findings included a leukocytosis of 37 K/MM3, thrombocytosis of 1272 K/MM3, and microcytic anemia of 9.4 G/DL. Computed tomography (CT) imaging of the chest revealed a right upper lobe consolidation. The patient was diagnosed with community acquired pneumonia and started on antibiotics. Abdominal CT done to rule out other sources of infection revealed cholelithiasis and asplenia. Target cells were seen on the peripheral smear and hemoglobin electrophoresis demonstrated hemoglobin S, hemoglobin C, and an elevated amount of hemoglobin A2. Further genetic analysis subsequently revealed HbSC disease with silent alpha thalassemia carrier. Flow cytometry to rule out myeloproliferative disorders was negative. On discharge he was administered the appropriate immunizations including pneumococcal 13, meningococcal and Haemophilus influenza vaccines.

Discussion: HbSC disease occurs in the 9% of the African American populations, of which 20% meets criteria for silent alpha thalassemia carrier. Patients with HbSC disease carry the same litany of complications as HbSS disease, but at a reduced frequency and severity of disease. Surprisingly patients with auto-infarction of spleen usually have HbSC with thalassemia trait rather than HbSS. Those with HbSS have a fibrotic, atrophic spleen and are relatively protected from splenic infarction. The platelet hyperactivity in these patients reflects an increased circulating young metabolically active platelets resulting from an auto-splenectomy. Thrombocytosis and asplenia leads to twice as much risk of thromboembolism in HbSC than HbSS.
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DIGGING DEEPER: A CASE OF ASYMPTOMATIC URTICARIA

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Urticaria is characterized by a well-demarcated eryhematosus rash driven by histamine-mediated vasodilation. Acute urticaria lasts for less than 6 weeks and usually represents a cutaneous reaction. Chronic urticaria persists beyond 6 weeks and warrants more comprehensive evaluation.

Mr. S was an 82-year-old man presented with subacute functional decline, global weakness, and one-month history of diffuse rash involving his back, legs, and arms.

Exam was notable for low-grade fever and diffuse raised eryhematosus blanching papules and plaques involving bilateral flanks and lower extremities as well as dependent palpable purpura. Labs revealed hyponatremia, acute kidney injury, microcytic anemia, thrombocytosis, and elevated C-reactive protein. Further evaluation revealed the presence of antineutrophil cytoplasmic antibodies (perinuclear and cytoplasmic) and decreased complement levels (c4). Skin biopsy revealed urticaria, with nuclear debris and increased cellularity raising the possibility of resolving urticarial vasculitis. Cetirizine was empirically started. Hospital course was complicated by MSSA bacteremia, which was treated with intravenous antibiotics. Despite adequate treatment with negative subsequent blood cultures, fevers persisted prompting imaging. Abdominal CT revealed diffuse lymphadenopathy. Lymph node biopsy was consistent with nodal marginal zone lymphoma. Oncology recommended observation given the indolent nature of marginal zone lymphoma. He was discharged with close follow-up.

Mr. S presented with functional decline and urticarial rash in the context of marginal zone lymphoma. As demonstrated here, an urticarial rash may be a harbinger of underlying systemic processes such as autoimmunity, infection, and malignancy. This is a reminder to maintain a high index of suspicion in the right clinical context.


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