ACP American College of Physicians
Leading Internal Medicine, Improving Lives

Utah Chapter

2015

Clinical Vignette Competition

UTAH ACP RESIDENTS & FELLOWS COMMITTEE

Scott C. Woller, MD FACP – Chair
Kencee Graves, MD
Julianna Desmarais, MD

SPRING BANQUET & CLINICAL VIGNETTE PROGRAM
FRIDAY, FEBRUARY 27, 2015

University of Utah | Guest House | Douglas Ballroom

4:45 PM **WELCOME & OPENING REMARKS**
Residents & Fellows Committee

JUDGES
David Fedor, MD
Josh Labrin, MD
Andrea Nelson, MD

4:50 PM **PRESENTATIONS**

5:25 PM **ANNOUNCE RUNNERS-UP AND 1ST PLACE**

5:30 PM **CLOSING COMMENTS – Adjourn for Dinner & Awards Ceremony**
Residents & Fellows Committee

UTAH ACP RESIDENTS & FELLOWS COMMITTEE | MISSION STATEMENT

To Improve the professional and personal lives of Utah Residents and Fellows and encourage participation in the American College of Physicians – American Society of Internal Medicine.

1. **Foster Internal Medicine Resident's interest in the ACP – ASIM.**
   - Encourage ACP associate membership and a lifelong interest in ACP – ASIM.
   - Encourage representation on National and Local ACP subcommittees.
2. **Foster educational Opportunities for Internal Medicine Residents.**
   - Encourage participation in local and national ACP – ASLIM Associates Clinical Vignette and Research opportunities.
   - Organize the local competitions. Provide information on board review courses. Publicize local and national educational opportunities. Work with residency programs to improve residency education.
3. **Identify practice management issues for Internal Medicine Residents.**
   - Provide information for residents as they prepare to enter practice, such as practice opportunities and contract negotiation.
4. **Identify public policy concerns of residents.**
   - Monitor local and national health policy and how it relates to Internal Medicine and residency training.
5. **Encourage an interest in community service.**
   - Identify ways associates can become involved with community service in Utah.

CONTENTS

THE FEVER CONTINUES, A DIAGNOSIS ELUDES | AMANDA BREVIU, MD

CASE PRESENTATION: The patient is a 30-year-old male with prior medical history of type 2 diabetes mellitus and obesity who presents with 4 weeks of persistent fever. Other symptoms include a headache, dry cough and profound fatigue. He had been hospitalized twice during his course of symptoms and both times a definitive diagnosis was not found. Prior infectious work-up including blood cultures, urine cultures and chest x-ray were all negative.

PHYSICAL EXAM ABNORMALITIES: Vital abnormalities included daily fevers of 39˚ Celsius. Pertinent negatives on exam included lack of neck stiffness, lungs clear to auscultation. There was no palpable lymphadenopathy or splenomegaly though exam was limited by body habitus.

Lab/Imaging Results:Lab work was notable for elevated ESR and CRP, 123 and 28 respectively. CBC was notable for normocytic anemia with hemoglobin of 11.7. Abdominal CT revealed enlarged lymph nodes and spleen. PET scan revealed diffuse uptake throughout lymph nodes and spleen.

DIFFERENTIAL DIAGNOSIS: Prior to imaging results, the differential for his fever of unknown origin remained broad and included the categories of infectious, rheumatologic or hematologic/oncologic. Infectious etiology became less likely given persistently negative cultures and other negative infectious testing. Rheumatologic etiologies of fever of unknown origin such as adult Still's were considered. Malignancy remained a significant possibility despite anemia being his only hematologic lab abnormality. Following the PET scan, excisional lymph node biopsy was performed and results revealed a diagnosis of Multifocal Castleman's disease.

DISCUSSION: Multifocal Castleman's disease is a disorder involving proliferation of benign lymphocytes due to inappropriate pro-inflammatory cytokines, particular interleukin-6. Human herpes virus 8 and HIV are both known to be associated with Castleman's disease. Interestingly, this patient was negative for HIV and HHV-8. There is an entity of HIV and HHV-8 negative Multifocal Castleman's disease that is referred to as idiopathic Castleman's.

This patient underwent treatment with weekly rituximab. He was also started on prednisone, initial dosing of 100 mg which has been tapered gradually. He has had resolution of his fevers and improvement in his inflammatory markers.

CONCLUSION: Multifocal Castleman's disease is an uncommon disorder which may manifest as fever of unknown origin. There is an entity of this diagnosis that occurs in HHV-8 and HIV negative individuals, though is still mediated by inappropriate IL-6 signaling. Current treatments focus on blocking this pathway of signaling.

CASE: 67 y/o M with epilepsy and ESRD admitted for seizures and encephalopathy. The patient was found down "seizing." He was taken to the ED where he had a second similar seizure. The seizures were described as shaking and gasping. Patient was markedly confused after each seizure and had incontinence with the second. He was given 4 mg of Ativan in the ED. Wife reported one missed dose of anti-epileptic medications in the last month. Patient had missed HD two days prior. No recent vomiting, diarrhea, fever, headache, or neck stiffness. Patient was afebrile. Respiratory rate was 14-22 bpm. He was somnolent, but moved all limbs after sternal rub. His chemistry was most notable for a CO2 of 43 mmol/L. Most recent lactosamide level was therapeutic. An ABG revealed a pH of 7.48, pCO2 of 61 mmHg, and bicarbonate of 45 mmol/L. Ionized calcium was low at 1.1 mmol/L with a total calcium of 11.1 mg/dL. Spot EEG and CT head were unrevealing. The patient eventually underwent hemodialysis with improvement in his presenting electrolyte abnormalities and mental status.

DISCUSSION: The patient's seizures were initially attributed to uremia. However, the most significant new electrolyte abnormality was the patient's elevated bicarbonate. The patient had no history of vomiting, diarrhea, or changes in diuretic medications. He was volume up and had not received any bicarbonate or citrate containing products. Upon clearing, the patient denied cocaine use – a documented cause of bicarbonate ingestion. However, he reported recently suffering from "heartburn" with therapeutic ingestion of large volumes of "baking soda." Alkalemia secondary to NaHCO3 ingestion is known to precipitate seizures (1). Alkalemia results in deprotination of albumin, increased binding of Ca2+, and ionized hypocalcemia. Calcium plays an integral role in nerve conduction (2). Though this patient's pH was not dramatically elevated, his ABG with iCa2+ was collected > 7 hours after presentation, and his pCO2 suggested an exaggerated respiratory compensation, possibly secondary to multiple doses of Ativan. Patient's pH and ionized calcium were likely higher/lower at time of initial seizure. Treatment of metabolic alkalemia in an HD patient is straightforward -- stop all alkalizing agents and hemodialyze with or without negative bicarbonate dialysate (3). This case reveals the critical importance of a sound history (4) and a rare, but important, cause of seizures that should be considered when working up patients with new or break-through seizures.

1. Fitzgibbons LJ, Snoey ER. Severe metabolic alkalosis due to baking soda ingestion: case reports of two patients with unsuspected antacid overdose. J Emerg Med. 1999;17(1):57-61.
2. Castilla-Guerra L, Fernandez-Moreno MC, Lopez-Chozas JM, Fernandez-Bolanos R. Electrolytes Disturbances and Seizures. Epilepsia. 2006; 47(12):1990-98
3. Solak Y, Turkmen K, Atalat H, Turk S. Baking soda induced severe metabolic alkalosis in a haemodialysis patient. NDT Plus. 2009;2(4):280.
4. Hampton JR, Harrison MJ, Mitchell JR, Prichard JS, Seymour C. Relative contributions of history-taking, physical examination, and laboratory investigation to diagnosis and management of medical outpatients. Br Med J. 1975;2(5969):486-9.

ONCE UPON A TIME IN MEXICO | HARRIS CARMICHAEL, MD [RI]

IDENTIFICATION: 20 YO Hispanic female

CHIEF COMPLAINT: Abdominal Pain

HISTORY: A previously healthy 20 YO female presents to the ED with nausea and vomiting for 1 day. The onset of abdominal pain was atraumatic and acute. She denies diarrhea, constipation, fever, chills, melena, or coffee-ground emesis. Of note patient traveled to Mexico in July for three weeks 2 months prior. She ate a variety of local foods but had no contact with dogs or farm animals. No other family members from the trip have similar abdominal pain.

PHYSICAL ABNORMALITIES: Tachycardia. TTP in upper quadrants R>L. Soft, ND, no guarding, +BS

LAB RESULTS: WBC: 18.71; Polymorphonuclear 77; Band 14; Eosinophil 0

CT ABDOMEN: A solitary 8.8 cm uniloculated cystic mass in the right hepatic lobe. Findings are most suggestive of an amoebic abscess. Pyogenic abscess could also have this appearance. Moderate ascites.

DIFFERENTIAL DIAGNOSIS: E. histolytica, Echinococcus, pyogenic/bacteria, and biliary cystadenoma.

CASE PRESENTATION: The patient was septic and after administration of Zosyn and Flagyl patient was noted to have hives that resolved with IV Benadryl. The etiology of the hives were unclear given onset quickly after IV Flagyl. Given the patient's CT demonstration of rupture from the liver cyst it was determined that IR should drain the remainder of the cyst. The yield from the liver drain was negative organisms and all cell types. Echinococcus antibody was noted to be indeterminate. All other labs were normal. Hives resolved after 24 hours (at the same time Flagyl was discontinued) and the patient improved after drain placement and Zosyn.

DISCUSSION: Given the two month timeline after the patient's trip to Mexico and 9cm hepatic abscess it was considered the patient would have sufficient immunity to a theoretical organism causing the abscess. The indeterminate status of Echinococcus antibody and uniloculated appearance of the cyst with no organisms seen on examination of the cystic fluid cast doubt on the diagnosis of Echinococcus. A recheck of Echinococcus antibody was scheduled for 10 days. Stable, the patient was discharged on Unasyn after a second sample of drain fluid was sent for analysis and returned with parasitic organisms with refractile hooklets with marked acute inflammation, morphologically consistent with Echinococcus. Pt was treated with Albendazole and surgical resection of segments 5 and 6 of the liver.

CONCLUSION: Echinococcus.

SO YOU HAVE A NOSE BLEED? | HARRIS CARMICHAEL, MD [RI]

IDENTIFICATION: 30 YO male

CHIEF COMPLAINT: Epistaxis and thrombocytosis

HISTORY: Pt was discharged to inpatient rehab 3 days prior to admission to the STICU. The patient was originally admitted for trauma due to an auto vs. pedestrian. During this admission the patient was administered heparin for DVT prophylaxis. This admission was complicated by HCAP pneumonia requiring the use of Vancomycin and Zosyn. After discharge to inpatient rehab, where he was continued on lovenox for his reduced mobility, the patient was doing well until he developed a fever. Chest CT demonstrated increased right pleural effusion. Given concern for persistent HCAP he was restarted on vancomycin and Zosyn.

The following morning he developed epistaxis. He did not have any other bleeding or new bruising. He did cough up several clots of blood likely secondary to severe epistaxis. CBC revealed platelet count had dropped from 549 to 9 in 24 hours. The patient was transferred to the STICU.

PHYSICAL ABNORMALITIES: Active bleeding from bilateral nares, clotted blood noted in bilateral nares, no clear source. Tachycardic, decreased breath sounds in bilateral lung bases, course rhonchi bilateral bases

LAB RESULTS: Hgb 8.5 (from 10.1 in 24 hrs), Plt 9 (from 549), Fibrinogen 592, LDH 251, HIT antibody negative, Smear: no significant schistocytes to suggest that the recent marked drop in platelet count is related to microangiopathic hemolysis. There are no platelet clumps to indicate pseudothrombocytopenia. There is no evidence for hematologic malignancy.

DIFFERENTIAL DIAGNOSIS: Pseudothrombocytopenia, DIC, HIT, Drug induced immune thrombocytopenia, malignancy, aplastic anemia

CASE PRESENTATION: The patient had no evidence of a consumptive coagulopathy (LDH, Fibrinogen, schistocytes) although a falling anemia and new thrombocytopenia. Given his appropriate timeline of 7 days exposure to Vanc/Zosyn and repeat exposure within 24 hours of epistaxis and thrombocytopenia it was considered that the patient is most likely suffering from drug induced immune thrombocytopenia. Offending antibiotics were changed, he was started on 42 grams IVIG followed by a total of 5 units platelet transfusion and 2 pRBC's for progressing anemia.

DISCUSSION: The patient stabilized, thrombocytopenia, anemia, and pneumonia resolved, and he was discharged. The most likely etiology remains drug induced immune thrombocytopenia and a sample has been sent for laboratory conformation by the Mayo Clinic for Vancomycin/Zosyn induced immune thrombocytopenia.

CONCLUSION: Drug induced immune thrombocytopenia, likely secondary to Vancomycin or Zosyn.

INTRODUCTION: Secondary causes of weight gain are rare compared to primary obesity, but they do warrant consideration. A complete history and physical exam is likely to elucidate the cause, so that directed laboratory evaluation can confirm the diagnosis.

CASE PRESENTATION: An 18-year-old Swedish female presented to her primary care physician with complaint of weight gain, increased body hair, and progressive for 2-3 years. She was particularly concerned that she was beginning to be significantly more overweight and round-faced than her identical twin, despite similar eating and exercise habits. She additionally complained that her menses had become irregular, she had developed a bump on the back of her neck, abdominal stretch marks, excessive sweating, fatigue, and darkening of the skin in her armpits. For the past 2 years, she had been experiencing dull pain in her right flank. Her primary care physician considered the constellation of symptoms and performed abdominal ultrasound, which revealed a mass. Subsequently, a triphasic CT was performed and she was found to have a 4x5cm mass on her right adrenal gland. She was referred to urology and underwent right adrenalectomy without complications. Perioperatively, she was treated with high dose IV hydrocortisone and discharged on 20mg in AM and 10mg in PM and asked to follow up in endocrinology clinic. Review of her initial labs revealed a 24-hour urine cortisol of 550mg. Because of the extreme cortisol excess, the postoperative replacement dose was tapered slowly over several months. Over the next 2-3 months, she experienced gradual, weight loss, decrease in body hair, acne, and sweating. Her “moon face” and “buffalo hump” were much less pronounced.

DISCUSSION: The patient presented with classic features of Cushing’s Syndrome, which may be caused by several conditions including exogenous glucocorticoid, pituitary tumor, other ACTH-secreting tumor, adrenal hyperplasia, and adrenal adenoma. Adrenal adenoma will be responsible for about 10% of Cushing’s syndrome. First-line evaluation for Cushing’s syndrome is cortisol evaluation with either 24-hour urine cortisol or overnight 1mg dexamethasone suppression test. This patient was evaluated in a more roundabout way, likely due to her complaint of flank pain. Unilateral adrenalectomy is the preferred treatment for Cushing’s syndrome caused by adrenal adenoma. Special considerations include increased risk for VTE perioperatively, and high dose glucocorticoid replacement followed by slow taper to allow the contralateral adrenal gland to resume function.

DAZED AND CONFUSED: HOW I LEARNED TO STOP WORRYING AND RATIONALLY ORDER IMAGING FOR SYNCOPE WHEN INDICATED | AARON CROSBY, MD [RI]

IDENTIFICATION AND CHIEF COMPLAINT: Mr. S is a 54 year old otherwise healthy gentleman who presented to the Emergency Department following an unwitnessed fall and loss of unconsciousness at home.

HISTORY: The patient arose the morning of admission in his usual state of health. He lost consciousness while pouring himself a glass of juice, and his wife heard his fall and found him on the kitchen floor breathing but unresponsive for 3-5 minutes. She noted occasional twitching of his legs without rhythmic jerking movements, tongue-biting or bowel or bladder incontinence. Paramedics transferred him to the Emergency Department. Over the course of the hour long ride, his confusion improved, and in the Emergency Department he was fully alert. He denied light-headedness, chest pain, palpitations, déjà vu, or olfactory hallucinations. He noted fatigue and subjective fevers for three days leading up to the episode.

PHYSICAL EXAM: Vital signs were all within normal limits. The patient was awake and alert. Head and neck had no meningsmus or bite-marks on tongue. Heart, lung and neurologic exam were completely normal.

DIFFERENTIAL DIAGNOSIS: The differential diagnosis focused on characterizing the patient's episode as syncope or seizure. The patient had no seizure history, incontinence, or tongue biting, but did have a prolonged period of unconsciousness and confusion following the episode.

LABS, IMAGING, OTHER STUDIES: After consultation with a neurologist, an MRI and EEG were obtained. The EEG was benign, but an MRI of the brain showed edema of the right temporal cortex concerning for encephalitis. A lumbar puncture was significant for 201 WBCs (91% lymphocytes), normal glucose, elevated protein and positive HSV PCR.

DIAGNOSIS: Herpes Simplex Virus Encephalitis

TREATMENT: The patient underwent two weeks of IV acyclovir and was placed on levetiracetam for three months.

DISCUSSION: One of the questionable practices the ACP highlights in its list for the Choosing Wisely campaign is the use of brain imaging for patients with LOC. It is true that the majority of patients who present with syncope should not receive a brain MRI. However, in this case, the history and physical led to a rational use of neuroimaging that ultimately resulted in the diagnosis and treatment of a potentially fatal condition. This illustrates the importance of using fundamental clinical skills in the delivery of high value care.

DON'T SWEAT THE SMALL STUFF | DANIEL ERHARDT, MD/MPH [RI]

CASE PRESENTATION: The patient is a 47 year old gentleman with no significant past medical history who presented to the ED with a three month history of fatigue, weakness, fever, myalgias, night sweats, chills, and 30 pound weight loss. He had initially presented to his primary care doctor after noting similar symptoms two months prior. He was diagnosed with mononucleosis and given reassurance. His symptoms persisted, prompting the patient to seek further evaluation.

In the ED, he was febrile to 39.8C with significant pancytopenia. Abdominal CT on admission was notable for significant splenomegaly and multiple enlarged abdominal lymph nodes. He was admitted to the hematology service for further evaluation and management of suspected lymphoma.

PHYSICAL EXAM ABNORMALITIES: Exam was noted for febrile and cachectic gentleman. He had significant hepatosplenomegaly. Peripheral lymph node exam was benign. Follow-up PET/CT imaging demonstrated systemic evidence of disease including bulky, disseminated hypermetabolic adenopathy, splenic hypermetabolism, and innumerable skeletal hypermetabolic foci.

DIFFERENTIAL DIAGNOSIS: Concurrent multisystem infection was of concern as the patient had met sepsis criteria during admission. A diagnosis of Hodgkin lymphoma was confirmed by retroperitoneal lymph node biopsy. A bone marrow biopsy was performed and found to have evidence of hemophagocytosis, and the patient met six out of eight criteria for Hemophagocytic lymphohistiocytosis (HLH), which included ferritin level >3000, and soluble IL-2a receptor >24,000 copies. Of note, infectious workup was incidentally positive for EBV infection with > 40,000 copies/mL.

TREATMENT: The patient's pancytopenia was managed supportively with blood product transfusions. He was started on dexamethasone, and ABVD chemotherapy was initiated for Hodgkin lymphoma. By treating the underlying malignancy, the immune system derangement would presumably be corrected and his HLH would resolve.

CONCLUSION: HLH is a rare life-threatening condition that results in aberrant activation of the immune system. Untreated patients have a survival of months, and so early diagnosis is critical. HLH can occur due to an immunologic trigger such as infection, malignancy, or autoimmune condition. It is diagnosed by involvement of at least five out of eight criteria. Differential diagnosis is that of several multisystem organ illnesses characterized by fever, liver failure, and neurologic symptoms. When the etiology is known, treating the underlying condition is considered to be an effective method to resolving the HLH. Interestingly, either the patient's lymphoma or EBV infection could have theoretically triggered his HLH, and he coincidentally suffered from both illnesses simultaneously.

HISTORY: The patient is a 58 year old male in good health. He awoke one morning with sudden onset cough and fever to 40 degrees with rigors, body aches, and malaise. Several hours later he went to the ED, where he was found to be hypoxic and tachypneic.

PHYSICAL ABNORMALITIES: On exam the patient was alert and oriented but ill appearing. He had dry mucous membranes and no oral/pharyngeal abscess. His neck had no tenderness, masses or lymphadenopathy. Lungs auscultation revealed bilateral crackles. His abdomen was soft, non-tender to palpation with normal bowel sounds.

LAB/IMAGING RESULTS: On admission, his chemistries showed a BUN of 20 and creatinine of 1.42. His WBC on admission was 9.5 with 70% neutrophils. His S. pneumonia urine antigen was positive and blood cultures were drawn. His rapid flu antigen was negative. Chest x-ray revealed a moderate multifocal pneumonia, predominantly involving the left lower lobe.

HOSPITAL COURSE: The patient's presentation appeared classic for community acquired pneumonia, and he was started on ceftriaxone. However, after several days he remained febrile and hypoxic and developed a WBC and hemoptysis. Chest CT did not reveal an empyema or loculation. Drug resistance infections, such as MRSA, or alternative sources of infections, such as endocarditis, crept higher on the differential. However, on hospital day four, his blood culture grew out fusobacterium species.

DISCUSSION: Fusobacterium is a gram negative anaerobic rod that is part of oral and bowel flora. The bacteria can cause Lemierre's Syndrome, a complication of fusarium pharyngitis or dental infections. If the infection extends into the parapharyngeal space, the bacteria can cause jugular vein thrombophlebitis. This often results in septic emboli to the lungs and can mimic a multi-lobar pneumonia. The syndrome is diagnosed with positive blood cultures and CT of neck. Treatment is 2 weeks of antibiotics with anaerobic coverage, such as Unasyn, Zosyn, or a carbapenem. In addition, fusobacterium bacteremia can result from abdominal abscess.

CONCLUSION: As the patient had no concerning oral or neck exam findings, no neck imaging was performed. He had CT abdomen that did not show an abdominal abscess. He was switched to ertapenem and after 3 days his fever, hemoptysis, hypoxia, and malaise resolved. He was switched to oral Levaquin and Flagyl for two weeks with plan for repeat chest CT in two weeks to ensure resolution.

AN ATYPICAL "COLD" | DAVID GILL, MD [RI]

CASE PRESENTATION: 29 year old healthy female presented to her PCP in August for evaluation of cough, sore throat, rhinorrhea, & subjective fevers of 2 weeks duration. At that appointment, she was anemic (Hgb 7.5) with elevated total bilirubin (11) and was prescribed azithromycin. After 3 days of treatment, her symptoms worsened and she presented to ED where she was found to have Hgb 5.3, WBC 38, Tbili 6.6, and a mild transaminitis.

On presentation, she endorsed dark urine correlating with the onset of symptoms. Denied recent travel, but she did note that both her husband and son had recently been sick with URI-type symptoms. CXR and urinalysis were unremarkable, and she was admitted for further workup.

PHYSICAL ABNORMALITIES: Exam was notable for jaundice with mild icterus and pale mucus membranes. No pharyngeal exudates, rash, splenomegaly, or lymphadenopathy. No increased work of breathing or respiratory distress. No murmurs.

LAB RESULTS: WBC 38 (PMN 58%, bands 17%, lymphocytes 19%) | Hgb 5.3 (8% reticulocytes) | Platelets 618 | AST 73, ALT 95, Tbili 6.6 | Viral Hepatitis Panel (-) | LDH 1160, Haptoglobin 10 | Mycoplasma PCR (+) | IgG-C3 (+) | Cold auto-antibody anti-I (+)

DIFFERENTIAL DIAGNOSIS: Hemolytic anemia was clear on presentation with elevations in LDH/bilirubin and decreased haptoglobin, but the underlying etiology required further workup. She had no history of hereditary causes (G6PD, sickle cell, hereditary spherocytosis), autoimmune disease, heart valve abnormality, or liver disease. With her viral URI prodrome, mild transaminitis, and marked leukocytosis, hemolytic anemia secondary to Mycoplasma cold agglutination was the presumed diagnosis. Confirmation was obtained with cold auto-antibodies and Mycoplasma PCR.

TREATMENT: In addition to azithromycin for 5 days, she was started on prednisone 100mg daily. She experienced rapid resolution of symptoms. Prednisone was tapered and discontinued two weeks after discharge.

CONCLUSION: Extra-pulmonary manifestations of Mycoplasma including hemolytic anemia, hepatitis, arthritis, myocarditis, and pericarditis are common. Hemolytic anemia typically occurs in the extremities due to slightly decreased temperatures leading to complement-mediated hemolysis. This is named cold agglutinin disease.

She displayed typical cold auto-antibodies to Mycoplasma (anti-I), but very atypically her antibodies were IgG. Typically IgM antibodies are expected as IgG is associated with warm agglutinin disease. As in this patient, marked leukocytosis can be present and is associated with underlying hemolysis.

Treatment includes supportive care, blood transfusion, cold avoidance, and corticosteroids. Immunosuppressants and splenectomy are sometimes necessary for refractory cases.

DO NOT JUST GO WITH THE FLOW | ALEXIS HARRISON, MD

CASE: A 49 year-old woman who was infrequently seen in the Hereditary Hemorrhagic Telangiectasia (HHT) clinic presented with shortness of breath, lower extremity edema, 50lb weight gain, renal failure, and liver dysfunction. She had been having severe epistaxis requiring transfusions for years. She had tried septal dermoplasty and been in a clinical trial of a topical therapy all without epistaxis relief. She had chronic shortness of breath attributed to profound anemia. Recently, she had developed volume retention. She was confusingly prescribed ambrisentan for post-capillary pulmonary hypertension and had been on increasing diuretics and ultimately dialysis for volume control and cardiorenal syndrome.

She has HHT (mutation in ACVRL1) which she and a sister inherited from her mother. She had no brain AVMs, but did have a pulmonary AVM for which she had coil embolization. Screening for liver AVMs is not commonly done and had not been done prior to this admission.

PHYSICAL EXAM AND LABORATORY ABNORMALITIES: Her exam was notable for her nares filled with blood-soaked cotton, a JVP elevated to the angle of her jaw, marked 4+ pitting bilateral edema, and multiple typical telangiectasias on her fingers, hands, and face. CBC showed anemia (Hbg of 7.2) and ferritin was 34. CMP showed hyponatremia, kidney injury (BUN 47, Cr 4.93), and an elevated alkaline phosphotase/bilibrubin (248 and 8.0) without a transaminitis. BNP was 1920.

DIAGNOSIS: Presumed diagnosis was high-output heart failure. An echocardiogram showed hyperdynamic LV function, enlarged RV, and an elevated RVSP of 58 mmHg. Noninvasive cardiac output was 8.7 L/min. Swan-Ganz catherization confirmed high-output HF with a high cardiac index and a low SVR. An abdominal ultrasound was suspicious for hepatic AVMs.

TREATMENT: High-output HF from the combination of anemia and hepatic AVMs resulting in volume overload, hepatic congestion, and cardiorenal syndrome. Our patient underwent a Young's procedure, a surgery that closes the nasal cavity, protecting the nasal mucosa and thus preventing epistaxis. With improved epistaxis but continued HF symptoms, she was started on intravenous bevacizumab. She had dramatic improvement - currently euvolemic, off dialysis, and on maintenance diuretics and intermittent bevacizumab.

CONCLUSION: HHT is a disorder of angiogenesis characterized by mucocutaneous and visceral telangiectasia. One of the rare, concerning, and often missed complications of hepatic HHT is high-output HF. Missing high-output HF can lead to misguided treatments from a failure to understand the driving pathophysiology. Bevacizumab, an anti-VEGF Ab, has recently been very beneficial in treating HHT-related high-output HF.

IN THE FACE OF CLINICAL OBSCURITY, REMEMBER THE *COMMANDMENTS* | SAMMY HAYES, MD [RIIII]

CASE PRESENTATION: CK is a 21-year-old female with history of myelomeningocele status-post repair at birth with residual lower extremity paralysis, shunted hydrocephalus with multiple revisions, neurogenic bladder status-post augmentation with Monti, and neurogenic bowel status-post MACE procedure who presents to the ED with acute abdominal pain and distention starting the morning of presentation. Pain was associated with non-bloody, non-bilious emesis and poor appetite. Last stool was normal the morning of presentation. Catheterization of her urine appeared normal and did not improve pain. She denies fevers or chills.

In the ED, patient was afebrile with tachycardia in the 150s. Given patient's chief complaint and distended abdomen, nasogastric decompression was performed followed by imaging with CT abdomen and pelvis. CT showed large bladder calculus [2.2 x 2.5 x 2.8 cm] with bladder wall thickening, ascites with VP shunt in place, non-obstructing left renal calculus, and multiple cysts involving right ovary. Subsequent two-view KUB prior to urine catheterization showed no evidence of contrast extravasation. Given result, Urology stated bladder stone was unlikely responsible for pain. Patient was admitted to hospitalist service for further work-up.

PHYSICAL ABNORMALITIES: Physical examination significant for tachycardia despite fluid resuscitation and IV antibiotics received in ED. Heart rate remained in 130s with brisk capillary refill time. Heart and lung exams were otherwise normal. Abdomen was distended with hypoactive bowel sounds, multiple surgical scars, and a well-appearing stoma sites. Neurologic examination was significant for an awake, alert patient and a baseline sensation deficit below the level of the umbilicus.

LAB RESULTS: Presenting labs showed WBC 5.6 [with bandemia of 39%], normal chemistries and lactic acid, and a capillary blood gas of pH 7.31/pCO2 43/pO2 39/HCO3 22/BD 4. Catheterized urine was positive for small leukocyte esterase, 1 WBC, 1 RBC and negative for bacteria.

DIFFERENTIAL DIAGNOSIS: Pyelonephritis complicated by renal/bladder calculi, viral gastroenteritis, ovarian cyst rupture with torsion, bowel obstruction, bladder extravasation, and Budd-Chiari Syndrome.

DISCUSSION: This case serves as a disturbing lesson reinforcing two truths in medicine: never ignore abnormal vital signs and always investigate unexplained bodily fluids. Though she was said not to be a surgical case, why did it take the treatment team nearly 36 hours before performing a paracentesis? Did host complexity obscure evident medical clues? Retrospection is often unforgiving. The obtained fluid grew bacteria within two hours and exposed the underlying diagnosis: bladder extravasation with secondary intraperitoneal and shunt infections.

CASE PRESENTATION: Patient is a 25-year-old female who presents with progressive fatigue, weakness, and low-grade fevers since delivery of her second child three weeks ago. Patient had a history of MVA and splenic laceration in 2006. Years after, she developed splenomegaly, leukopenia, anemia, and transaminitis. The patient was seen by both a gastroenterologist and hematologist in the year prior to her presentation. Bone marrow and liver biopsies were non-diagnostic. Mother brought patient to the emergency department when she spiked repeated fevers as high as 40 °C for two days straight.

PHYSICAL EXAM: Patient was jaundiced, lethargic and ill-appearing. RUQ was tender with liver edge palpable 2cm below the costal margin. Scattered ecchymoses over bilateral anterior shins.

CBC was significant for WBC 1.7 K/uL, Hgb 9.7, Plts 104K, and ANC 1.0. LFTs were significant for Alb 2.6, Tbili 3.0, AP 1128, ALT 178, AST 458. INR 1.7. LDH was 1722.

Liver, needle core biopsy: highly atypical lymphoid infiltrate, with a small population of clonal T-cells in the background of reactive oligoclonal T-cells. Some slides with hemophagocytosis.

EBV PCR: 972,000 copies

EBV VCA IgM: negative

EBV VCA IgG: positive

Interleukin-2 Receptor: 3173

Ferritin 39036

Fibrinogen 149 mg/dL.

DIAGNOSIS: The findings are overall consistent with systemic EBV positive T-cell lymphoproliferative disorder of childhood. The patient also met clinical and laboratory criteria for diagnosis of Hemophagocytic Lymphohistiocytosis (HLH).

DISCUSSION: Systemic EBV positive T-LPD of childhood is a clonal proliferation of EBV positive T-cells (or NK cells) with an activated cytotoxic phenotype that mainly affects children and young adults. It may follow acute infection with systemic EBV or evolve out of chronic active EBV infection (CAEBV). In this case, clonality was not definitively demonstrated but the degree of cytologic atypia and abnormal phenotype together supported a clonal process. Most prevalent in Southeast Asia, the etiology of this process is unknown, but association with EBV and racial predisposition strongly suggest genetic immune defect. Patient notably had two family members who “died of blood disorders”.

TREATMENT: Systemic EBV+ T-cell LPD of childhood is an aggressive disorder; some therapeutic success utilizing stem cell transplantation has been reported. Most patients have a fulminant course, with death in days or weeks. This patient developed HLH passed within six weeks of presentation. The mainstays of treatment for HLH is dexamethasone, cyclosporine, and etoposide.

EXERCISE CAN KILL YOU | STEPHEN JENKINS, MD [RI]

HISTORY: A previously healthy 18-year-old man was brought to the emergency department after loss of consciousness and possible cardiac arrest. He had been running on a treadmill at the gym when he suddenly felt lightheaded. He turned off the machine and then collapsed. A witness told paramedics that the patient had seizure-like convulsions. The witness felt a fast, thready pulse, and then couldn't feel a pulse. The patient turned ashen gray. The witness performed CPR for approximately one minute before a pulse was felt again. The patient regained consciousness shortly thereafter and was transported by ambulance. He denied any chest pain, palpitations, vision changes or dyspnea before he fell. He denied any family history of heart disease, sudden death, or drownings. He had recently enlisted in the Air Force. He denied any tobacco, alcohol or illicit drug use.

PHYSICAL EXAM, LABS, IMAGING: Exam showed a 1 cm laceration of the lower lip. The patient had a regular rate and rhythm. There were no murmurs with standing or Valsalva. Neurologic exam was non-focal. Laboratory work-up was unremarkable except for a troponin-I of 0.16. Toxicology screen was negative. An ECG was notable for deep Q waves in leads II, III, V5 and V6 with abnormal ST and T wave changes in V1-V4. An echocardiogram showed normal left ventricular function with upper normal left ventricular wall thickness and no valvular abnormalities. Exercise stress test was normal.

DIFFERENTIAL DIAGNOSIS: Causes of exertional syncope and cardiac arrest in a young man include hypertrophic cardiomyopathy, Brugada syndrome, long QT syndrome and arrhythmogenic right ventricular cardiomyopathy. We also considered seizure and neurocardiogenic syncope.

DIAGNOSIS AND MANAGEMENT: We were most suspicious of hypertrophic cardiomyopathy. Cardiac MRI was obtained, showing asymmetrical hypertrophy of the septum and basal to mid anterior wall. There was no evidence of systolic anterior motion of the mitral valve. Electrophysiology recommended placement of a subcutaneous ICD, which was performed without complications.

DISCUSSION: Hypertrophic cardiomyopathy should be considered in any young person with exertional syncope. Loss of consciousness can occur secondary to left ventricular outflow obstruction and from ventricular arrhythmias in the setting of myocardial fibrosis. ICD placement is recommended for patients with risk factors for sudden cardiac death, including prior ventricular fibrillation arrest, sustained ventricular tachycardia, family history of sudden death, unexplained syncope, LV wall thickness greater than 30 mm and heart failure.

A TRANSFER FROM AFAR | RUSSELL JOHNSON, MD MSC

CASE PRESENTATION: Patient is a 39-year-old female who presented to the emergency department with three months of abdominal pain. Patient was recently granted refugee status in the United States and had arrived overnight from Malaysia on a commercial plane arranged by the International Rescue Committee (IRC). Patient was born in Myanmar but had worked in Malaysia for several years prior to transfer. Three years prior to presentation patient was kicked in the back during an assault resulting in paraplegia. She received sporadic medical care in Malaysia, of which no record was available on admission.

PHYSICAL EXAM: Weight 36 kg, BMI 15.64. Extremely cachectic. Diffusely edematous. Protuberant abdomen with shifting dullness and fluid wave. 3x3 cm left buttock wound draining purulent fluid. 2x2 cm sternal eschar. Sacral ulcer. No movement in lower extremities.

LABORATORY RESULTS: WBC 41.4, 98% Neutrophils, Hgb 7.4,Plt 680, Cr 2.6, Phos 7.1 ALP 511, INR 1.5, ESR 57, Prealbumin <3.0. | Multiple wound cultures: Mycobacterium tuberculosis. | CSF, gastric AFB cultures: negative | MTB rifampin resistance: negative | Tracheal aspirate AFB x 1: negative

IMAGING: CT Abd/pelvis with bony destructive changes at T12-L1 with severe spinal canal narrowing. Asymmetric iliopsoas fluid collections tracking to gluteal muscles bilaterally and dermally on the left. Bilateral renal calcifications. Dilated small bowel. Moderate ascites. CT Chest with multiple pleural-based calcifications. Loculated left mid-lung effusion. No cavitary lesions within the apices.

DIAGNOSIS: Disseminated tuberculosis and Pott's disease with associated small bowel obstruction, severe malnutrition, and renal failure.

TREATMENT: Infectious diseases was consulted and recommended renally dosed IV rifampin, linezolid, and moxifloxacin to treat Mycobacterium tuberculosis in a patient with questionable absorptive capacity. Isoniazid was crushed and given through a nasogastric tube.

COURSE: Ultimately patient developed encephalopathy and tense ascites. She suffered cardiac arrest for unclear reasons on hospital day seven and passed.

DISCUSSION: Myanmar is one of 22 countries in the world considered to have a high tuberculosis burden. Prevalence is three times the global average and multidrug-resistant tuberculosis is on the rise. There are an estimated 180,000 new cases each year in Myanmar compared to the 23,000 new or relapsed cases that occur yearly in Malaysia. Whether this patient contracted tuberculosis in Myanmar or Malaysia is unknown. Although one sputum AFB smear was negative, this patient's transfer on a commercial airplane raises ethical questions and warrants investigation into the screening practices employed by the International Rescue Committee.

APPENDICITIS AND UPPER EXTREMITY THROMBOSIS FOLLOWING ART: A SINGLE OR SERIES OF UNFORTUNATE EVENTS? | TREVOR JONES, MD [RI]

CASE PRESENTATION: L.S. was a 25 yo women, 10 weeks pregnant with twins, sent to us for further evaluation of a left upper extremity DVT (UEDVT). She originally sought medical care for 2 days of left neck "muscle cramps" with ipsilateral arm swelling and sharp chest pain. An ultra sound at an outside E.D. showed left subclavian and internal jugular vein clots. Mrs. S had no chronic medical conditions, but had an appendectomy 12 days prior and conceived with ART. She remembered that the surgeon commented on her "large ovaries." The patient provided no history of repetitive left arm movement or neck vein catheterization.

PHYSICAL EXAMINATION: L.S.'s physical exam revealed skin darkening and mild non pitting edema of her left upper extremity. Pulses and neurologic exam were normal. She also had mild bilateral lower quadrant tenderness.

DIFFERENTIAL DIAGNOSIS: A secondary thrombophilia due to ART, ovarian hyper stimulation syndrome (OHSS), or pregnancy most likely caused the clot. Paget-Schroetter(PSS) was a possibility, but, in the setting of pregnancy, we did not perform a contrast venogram for definitive diagnosis. Malignancy, autoimmune disease, and inherited thrombophilia were considered less likely with her normal history and labs.

TREATMENT: The patient received low molecular weight heparin until 6 weeks post-partum for presumed secondary thrombophilia. As the diagnosis of PSS was equivocal, costoclavicular decompression was not pursued. The treatment of OHSS is fluids and supportive care. The patient had no signs of third spacing or electrolyte abnormalities

CONCLUSION: A single diagnosis could explain this presentation. A history of ART, "enlarged ovaries," clinical "appendicitis", and UEDVT are all consistent with OHSS. Pregnancy precludes abdominal CT and, as of this writing, the chart provides no surgical pathology for the appendectomy. Upper extremity DVT without history of central venous catheter or PSS is very rare. Pregnancy increases baseline risk but does not change clot distribution. ART, and more so OHSS, are associated with UEDVT. In-spite of all this, the timing would be abnormal. Her appendicitis occurred sometime during week 6 of pregnancy. OHSS starts between 0-3 weeks and lasts 1-2 weeks. Given the nature of secondary thrombophilia, a definitive diagnosis may not be possible. However this case is more than a series of seemingly disparate and unfortunate events.

CASE PRESENTATION: This is a 52-year-old male, previously healthy until ~9 months ago, who presented as a transfer from an OSH with seizures. He had suffered three generalized, tonic-clonic seizures and was initially able to respond appropriately to yes/no questions. He was admitted to the NCC. Work-up previous to transfer included an MRI brain, demonstrating 2 ring-enhancing lesions: at the L frontal lobe and the R basal ganglia. He underwent an LP; results were bland.

Previous to hospitalization, he had suffered a severe cognitive and functional decline with symptoms including: paranoia, delusions, memory loss, gait disturbances, weight loss and, more recently, non-productive cough and fevers. He had been residing at a care facility before the seizures had started. CT chest demonstrated diffuse, nodular "tree-in-bud opacities." Five days into his admission, he became increasingly encephalopathic, prompting intubation for airway protection. Bronchoscopy with BAL was performed.

PHYSICAL EXAM ABNORMALITIES: Upon transfer to the MICU, he was non-responsive to command with a low GCS. He was cachectic and exhibited L-sided hypertonia with hyperreflexia. He was recently pancytopenic, though WBC count had normalized. A new diagnosis of HIV was confirmed by Western blot. BAL revealed positive PJP PCR. CD4 count was 22. Serum testing for multiple infectious etiologies was negative, including Toxoplasma PCR of CSF.

DIFFERENTIAL DIAGNOSIS: As multiple new diagnoses were present—including encephalopathy, seizures, HIV/AIDS, PJP and progressive supranuclear palsy—the primary concern was discovery of a unifying diagnosis. The DDx for ring-enhancing lesions, especially in a patient with HIV/AIDS, includes Toxoplasmosis and CNS lymphoma. Work-up for an occult malignancy was negative.

TREATMENT: Keppra load and maintenance for seizures. IV Dexamethasone for brain edema. Empiric TMP/SMX for possible Toxoplasmosis, later continued for PJP. ~2 weeks into his admission, stereotactic brain biopsy was performed, demonstrating a B-cell lymphoma and HIV encephalitis. He was later started on HAART: Emtricitabine/Tenofovir and Raltegravir.

Unfortunately, ~1 month into his hospitalization, recurrence of seizures was noted, prompting treatment with Fosphenytoin. He suffered hypotension during infusion, devolving into PEA arrest, with ROSC after CPR. In the context of his marked deterioration, the decision was made to transition the patient to comfort care. He quickly expired after removal of life-sustaining treatments.

CONCLUSION: Primary CNS lymphoma (PCL) most often occurs in the setting of immunodeficiency and is an AIDS-defining illness. Symptoms may include focal neurologic deficits (70%), neuropsychiatric symptoms (43%), signs of increased ICP (33%), seizures (14%) and ocular dysfunction (4%)1. Clinical manifestations are insidious, requiring a high index of suspicion for diagnosis. Unfortunately, PCL is usually rapidly fatal. However, chemotherapy, most often with high-dose methotrexate-based regimens, may improve survival.

[1]*Bataille B, et al. Primary intracerebral malignant lymphoma: report of 248 cases. J Neurosurg 2000; 92:261.*

BOUNCING EYES AND TREMULOUS LIMBS | UYEN T. LAM, MD [RIII]

IDENTIFICATION: Patient is a 57 year old with controlled diabetes and hypertension who presented with vertiginous symptoms and tremors.

CHIEF COMPLAINT: Dizziness and tremors

HISTORY: Mr. X is a 57 year old with diabetes/hypertension who presented with several months of vision instability and constant horizontal shifting of his gaze causing significant nausea/vomiting. Symptoms relieved with Zofran but then developed gait instability requiring assistance of a cane for ambulation. Evaluated by ENT and diagnosed with vestibular neuropathy. Neurology agreed symptoms were secondary to vestibular nerve dysfunction and unlikely to be central given normal MRI brain and MRA head/neck. A few months later he developed diffuse tremors involving head and extremities. Symptoms were exacerbated by emotional distress and ataxia continued to worsen requiring assistance from his wife. Mr. X then developed visual field deficits, seeing a hair in his left eye that moved with eye movements but told by Ophthalmologist that eyes appeared normal. Repeat evaluation by ENT with vestibular nerve testing revealed 40% reduction in the left VOR and referred to the University for evaluation of possible spinal cord stroke.

PHYSICAL ABNORMALITIES: Alert and oriented middle-aged gentleman with circular nystagmus bilaterally; CN and extra ocular muscles intact, no diplopia. Baseline moderate titubation involving head, neck and upper body. Strength, sensation and reflexes normal bilaterally. Wide based gait, worse with tandem walking and required at least 1 person assist to perform functions. Remainder of exam normal with no lymphadenopathy.

LAB RESULTS: CBC and CMP unremarkable. Extensive testing for viral/bacterial/parasitic infection in both serum and CSF all negative. Vitamin deficiency, toxicity screening and endocrine workup normal. Imaging remarkable for anterior cervical spine lymphadenopathy concerning for metastatic disease.

DIFFERENTIAL DIAGNOSIS: Spinal cord stroke, paraneoplastic syndrome, infection such as HIV/HSV/VZV/West Nile.

CASE PRESENTATION: Patient subsequently had workup for paraneoplastic syndrome. ENT consulted, flexible laryngoscopy showed leukoplakia of right base of the tongue and right supraglottis. Preliminary results of FNA biopsy showed small cell versus nasopharyngeal carcinoma.

DISCUSSION: Adult onset opsoclonus-myoclonus syndrome is quite rare. Paraneoplastic and parainfectious causes always on the differential and most people achieve complete remission with treatment of underlying cause or immunotherapy in case of postinfectious causes.

CONCLUSION: Mr. X ultimately diagnosed with stage IVa, T1N2bMo squamous cell carcinoma of the base of the tongue, HPV positive. He was treated to neoadjuvant cisplatin followed by radiation with negative post-treatment PET-CT. Since then his vertiginous symptoms and ataxia have improved.

BRUISING FOR THE WORST | NATE LAMBERT [MSIII]

IDENTIFICATION: 23-month old with 6 month history of anemia and easy bruising

CHIEF COMPLAINT: "Easy bruising"

CASE PRESENTATION: 18-month old female presented to PCP in December with complaint of easy bruising. Bruising had occurred in unusual places such as abdomen. Since she had previously been taking iron supplementation for presumed iron deficiency anemia, her Hb was checked and was reportedly normal. Additionally, she had showed a steep decline in growth curve. Given new complaint of easy bruising, parents were given lab slip to check CBC. CBC was not obtained until 6mos later, and showed anemia, thrombocytopenia, and probable blasts. During this time, parents denied decrease in energy, lymphadenopathy, night sweats, or fevers without a source. After these findings, patient was admitted to PCMC for further evaluation. Upon admission, the following physical abnormalities and labs were noted:

PHYSICAL ABNORMALITIES: Abd: questionable palpable spleen, no hepatomegaly. Skin: Multiple scattered bruised over trunk and extremities, including some bruising over pubic, abdomen and thighs.

LAB RESULTS: Uric acid 4.6 | LDH 2134 | WBC 16.4 | Hct 26.7 | Plt 11 | 18% myeloid blasts | Positive markers: CD4, CD13, CD33 | Bone marrow aspirate: hypocellular, hemodilute marrow with 11% blasts | Chromosome analysis: t(1:22) translocation

DIAGNOSIS: Initially differential diagnosis was AML or ALL after peripheral smear showed blast cells. After flow cytometry and chromosomal analysis a definite diagnosis of AML M7 (Acute megakaryoblastic leukemia) was made.

DISCUSSION: Acute megakaryoblastic leukemia (AML-M7) is a rare form of AML. It accounts for 1% of all childhood leukemias with a yearly incidence of 0.5 per million. However, AML-M7 is the most common form of AML in children with Down syndrome (1). This was a case of acute leukemia presenting with initial symptoms of easy bruising. This was a unique presentation of acute megakaryoblastic leukemia t(1:22) in a young child without Down syndrome. The prognosis in patients with Down syndrome is good, with survival roughly around 70%. The prognosis for patients with AML M7 in children without Down syndrome has not been fully established due to its rarity. However, one source estimated survival rates to be as low as 35% (1), with the average survival of 10.4 months (2). Of note, this patient went on to receive 6 months of intense chemotherapy and has been in remission for 4 years.

REFERENCES:

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2. Cuneo, A et al., Acute megakaryoblastic leukemia. Atlas of Genetics and Cytogenetics in Oncology and Hematology. 2004;8(1):29-30

CURIOUS CASE OF DANCING EYES & FEET | JOSEPH MERRIMAN, MD [RII]

CASE PRESENTATION: A 57-year-old man with diabetes mellitus and hypertension presented with a constellation of symptoms progressing over a three-month period - blurry vision, tremors, nausea/vomiting, and weight loss. Three months prior to presentation, the patient experienced an acute onset of generalized malaise and vertigo. He sought care at an outside hospital with work-up including MRI of the brain, MRA of the head and neck, and an EEG yielding no significant findings. He was given a diagnosis of vestibular neuropathy. What brought him to our ED was the development of double vision with unusual eye and head movements to the point where he could no longer read or drive, and he required the use of a cane for ambulation due to ataxia.

PHYSICAL EXAM: Physical exam revealed revealed bilateral chaotic, rapid, omnidirectional eye movements (opsoclonus) in addition to head and extremity tremors at rest (ocular video available). Cranial nerves II-XII were otherwise intact. His gait was ataxic that worsened with tandem walking. His plantar reflex was downgoing and there was no lower extremity clonus. Speech remained intact and was AAOx3. Remainder of the physical exam was unremarkable.

DIFFERENTIAL DIAGNOSIS: CNS infection, seizure activity, paraneoplastic syndrome secondary to malignancy

LAB RESULTS: Laboratory data was within normal limits (CBC, chemistry, CSF studies). Pertinents included: paraneoplastic serologies pertinent for positive anti-glutamic acid (GAD) antibodies. MRI of the C and T-spine demonstrated enlarged cervical chain lymph nodes. PET-CT confirmed two right-sided enlarged hypermetabolic cervical lymph nodes in addition to uptake on the right base of the tongue. Biopsy of a right cervical lymph node demonstrated HPV-16 positive poorly differentiated squamous cell carcinoma.

TREATMENT: Initial treatment included high dose corticosteroids and IVIG; however, his symptoms progressed. Subsequent treatment for his underlying malignancy, with chemoradiation and cisplatin, led to a significant, symptomatic improvement.

CONCLUSION: This is the case of a 57-year-old previously healthy gentleman that presented with opsoclonus myoclonus syndrome (OMS). Diagnostic testing in this patient revealed a head and neck squamous cell cancer with a suspected primary tumor at the base of the tongue suggesting a paraneoplastic syndrome. Paraneoplastic syndromes are rare, but this would be the first case with a head and neck cancer associated with OMS. The patient was treated with corticosteroids and IVIG that did not alleviate his symptoms; however, targeted therapy with cisplatin and chemoradiation significantly improved his neurological symptoms and returned him to his daily routine supporting a paraneoplastic syndrome secondary to occult malignancy.

WHY IS THIS GUY'S HEAD FULL OF BLOOD? | ROSS SMITH, MD JD [RI]

CASE PRESENTATION: A 61 year-old male was brought into the ED by ambulance after his family hadn't heard from him in days. He was discovered sitting in his chair, confused and having urinated on himself. Head CT showed 1.3cm acute-on-chronic left-sided subdural hematoma. He was admitted to the neurosurgery service. Given the blood of different ages on CT, the question became why the patient was repeatedly falling. His family insisted that the patient had not consumed alcohol in decades, his toxicology screen was negative, and he had no history of trauma. Interestingly, the patient's mother had been wheelchair-bound the last years of her life.

Initial GCS was 13 so it was elected to observe the patient overnight and electively drain him in the morning. He deteriorated around 0200 so he was emergently drained in the OR. There were no complications and he was able to provide more history and a graded neurologic examination on post-operative day 1.

PHYSICAL EXAM ABNORMALITIES: Neurologic exam on post-operative day 1 showed the patient was diffusely weak. Most muscle groups were 4 to 4+ except triceps, finger flexion, and hip flexion were 2-3/5. There was some lower extremity muscle wasting. The patient also had bilateral cataracts and male-pattern baldness.

DIFFERENTIAL DIAGNOSIS: The patient endorsed difficulties with walking down stairs and getting out of chairs. L-spine MRI was performed which showed a normal spine but an incidental unsecured 8cm AAA. Neuromuscular tentatively diagnosed the patient with Type 2 Myotonic Dystrophy. The patient's mother actually had a muscle biopsy in 1987 at our institution and the slides were still archived. It appeared she had been misdiagnosed with limb-girdle muscular dystrophy. Genetic testing was pending.

TREATMENT: The patient had an uncomplicated post-operative course and gradually improved from his subdural. Treatment for myotonic dystrophy is supportive with aggressive PT, OT, and input from PM&R. With his recent subdural, EVAR was deferred a month until he could be anticoagulated. CTA showed the aneurysm was not leaking but did show a transition point in the colon suspicious for malignancy. Type 2 myotonic dystrophy does have an increased risk of colon cancer.

CONCLUSION: Acute-on-chronic subdural hematomas suggest that a patient has had repetitive head trauma over time. While alcoholism is often the culprit, other causes for falling should also be considered. The patient's family history and pattern of weakness strongly suggested a muscular dystrophy, in this case Type 2 Myotonic Dystrophy.

THE EYES SEE WHAT THE MIND KNOWS | TIFFANY A. VOLDEN, MD [RI]

CASE PRESENTATION: Mr. History: Patient is a 64 year old female with a history of depression, HTN, thymoma s/p resection, and breast cancer s/p lumpectomy and radiation who presented with a 1 week history of malaise and diffuse muscle pain. The patient described her symptoms as "coming down with the flu." She also complained of chest pain. She was found to have an elevated troponin and an EKG with Q waves in V1-3. She was admitted to cardiology for ACS rule-out.

PHYSICAL EXAM: No abnormal findings were found on physical exam, with the exception of a flat affect.

LAB RESULTS: Troponin 1.59, CK 1378, CK-MB 75 | Na 134, K 3.5, Cl 96, bicarb 29, BUN 8, Cr 0.9, Ca 9.9 | AST 131, ALT 158, alb 3.3, pro 6.8 | TSH 1.38 | ESR 7, CRP 1.6

DIFFERENTIAL: Presumed diagnosis was viral myositis although ACS remained high on the differential. A slightly elevated corrected calcium (10.5) brought malignancy into the differential; however, the patient was 5 years out from breast cancer and nearly 15 years from thymoma resection.

CASE PROGRESSION: CTA of the coronary arteries was performed and revealed nearly clean coronary arteries and two masses in the anterior and middle mediastinum. After being seen by rheumatology, the diagnosis of viral myositis was solidified as the patient showed significant improvement without intervention. She was discharged with oncology follow-up for further evaluation of the mediastinal masses. The patient returned to the hospital a few days later and was admitted to neurology for progressive muscle weakness. She received the eventual diagnosis of recurrent thymoma after biopsy of a mass. Muscle biopsy and antibody testing showed both myositis and myasthenia gravis, both paraneoplastic syndromes associated with thymomas.

TREATMENT: The patient is undergoing radiation for thymoma recurrence. The myasthenia led to intubation and subsequent tracheostomy. She is currently undergoing extensive rehab and slowly improving with aggressive treatment for myasthenia gravis.

CONCLUSION: Initially, the thymoma history was not known, and the patient continued down the workup and management for ACS. Just prior to CT, the case was re-examined and suspicion was raised for an unknown diagnosis. The findings on CT and deeper history-taking revealed thymoma as the likely unifying diagnosis. It is important to keep a wide and open differential instead of having narrow vision, especially if something does not quite fit. The mind will solve the puzzle, if only the eyes see the pieces.

CONGRATULATIONS, YOUR HGBA1C IS GREAT! | DEVIN WEST, MD [RI]

CASE PRESENTATION: The patient is a 37 year old male without past medical history who presented to the emergency department with double vision, weakness, and confusion that started the previous evening. The patient was busy with family obligations that day and had not eaten prior to developing symptoms. He initially had double vision, and his mother found him confused, clothed, and sitting in the bathtub. He woke the next morning still feeling weak, dizzy, and disoriented. He ate without symptom improvement and presented to the hospital. There, finger stick blood glucose was 27. The patient received 1 amp of D50, and his symptoms resolved. On presentation, he denied ingesting insulin or any medication except for ibuprofen. He was admitted to medicine for further evaluation and treatment.

WORKUP: Vital signs and physical exam were normal. Besides above hypoglycemia, initial labs were fairly unremarkable. CBC was normal, and BMP was significant only for mild hypokalemia. HgbA1c was extremely low at 4.4%. Beta-hydroxybutyrate was low. We then pursued targeted, synchronized, fasting studies. Glucose was low at 20. Insulin was at the upper-limit of normal at 22.9. C-peptide was elevated at 4.4. Proinsulin was elevated at 58.4. MRI was negative for pancreatic mass.

DIFFERENTIAL DIAGNOSIS: Differential diagnosis was limited in this non-diabetic patient with hypoglycemia and hyperinsulinemia. Suspected diagnosis was insulinoma. Other possibilities included noninsulinoma pancreatogenous hypoglycemia syndrome (NIPHS), persistent hyperinsulinemic hypoglycemia of infancy (PHHI), insulin autoimmune hypoglycemia, and sulfonylurea-induced hypoglycemia. As symptoms occurred predominately in fasting state, NIPHS was unlikely. The patient had no symptoms as a child making congenital disease also unlikely. Insulin antibodies and sulfonylurea screen was both negative.

TREATMENT: Additional imaging with triple-phase pancreatic CT revealed a 1.2 cm early arterial-enhancing nodule in the uncinate process. Biopsy via endoscopic ultrasound revealed pathology consistent with neuroendocrine tumor, confirming diagnosis of insulinoma. Patient was initially started on diazoxide and octreotide for insulin suppression. We consulted oncologic surgery, and he has surgery planned for the spring.

CONCLUSION: In 1991, a Mayo Clinic study calculated the incidence of insulinoma at approximately 4 cases per million per year. That same study noted that 87% of these were single, benign tumors. Surgical resection is the preferred treatment and resulted in 87.5% cure rate. Most insulinomas are sporadic, but some are associated with MEN 1 syndrome. It is wise to review diagnosis and treatment of this rare disease so as to provide optimum care when it does occur.

CASE PRESENTATION: EMS was called to the aid of a thirty-seven year-old man with a history of epilepsy and heart failure after his family noticed him breathing very rapidly. When EMS arrived, they saw a man in acute respiratory distress, tripodding with oxygen saturations in the 70s. Patient reported a three day history of productive cough, fever, pleuritic chest pain and stated he had been diagnosed with pneumonia. He denied weight gain, orthopnea, PND. In the ED, he was hypotensive, tachycardic and requiring 15L high flow oxygen. He had leukocytosis, acute renal failure, lactic acidosis, and a right upper lobe opacity was noted on imaging. He was diagnosed with severe sepsis secondary to pneumonia and was admitted to the intensive care unit.

PHYSICAL EXAM/DATA: Patient was confused, distressed, afebrile. He had decreased breath sounds and crackles in the right upper lobe, but lungs were otherwise clear. Cardiac exam was positive for JVP elevation to the jaw, hepatojugular reflux, palpable RV heave, loud split P2 and a systolic murmur at the left lower sternal border which increased upon inspiration. Unilateral lower extremity edema was noted, and venous duplex revealed extensive acute thrombus. Echocardiogram revealed normal left ventricular function and an enlarged, thickened right ventricle with RVSP of 107mmHg.

DIFFERENTIAL DIAGNOSIS: At this point, the pneumonia diagnosis was discarded and evaluation for pulmonary hypertension begun. His HIV, anti-SLC70, anti-centromere, anti-dsDNA were negative. ANA was positive. Liver tests and abdominal ultrasound revealed RV overload. Right heart catheterization demonstrated normal wedge pressure, severely elevated PA pressures (PAP 91/38) and reduced cardiac index. CTPA revealed chronic embolic disease and a right upper lobe pulmonary infarct. With confirmation by V/Q scan, the diagnosis of chronic thromboembolic pulmonary hypertension (CTEPH) was made. Thrombotic testing suggested anti-phospholipid antibody syndrome with 'triple positivity' (positive lupus anticoagulant, anti-beta-2-glycoprotein and anti-cardiolipin antibody).

CONCLUSION: On pointed questioning, he endorsed symptoms for one year, but his family reported symptoms for much longer. An echocardiogram several years earlier noted severe pulmonary hypertension and requested urgent follow up; however the patient lacked insurance and did not return.

CTEPH can be treated medically with agents such as sildenafil; however surgical intervention is the ultimate therapy. This patient's hospital course was complicated by the development of HIT. Currently he is on warfarin and sildenafil with plans for pulmonary thromboendarterectomy at some future date.

It was 3 am on a clear January morning during my third year of medical school. The moon was bright and shining in the patient, GW's, room. I was more worried about this patient than the many others I had worked with before. She had a long history of multiple sclerosis and had been in and out of the hospital many times in the past 10 years. The previous day, however, GW commented that she had not felt this well in months.

When I arrived in the ED, she was struggling to breathe. She had a non-rebreather oxygen mask—maxed out on oxygen flow—and still her saturations were only in the 70's. Her lips and fingers were blue indicating poor perfusion, but had a negative TTE and TEE. She also had a negative brain MRI/CT. She was positive for leukocytosis, and gram-positive cocci, on her blood stain. As I reviewed her vitals, labs, and imaging, I struggled to recall the criteria for SEPSIS that I had learned and been tested on ad nauseam for the past six months.

"Why can't I remember this," I thought. It wasn't that I had forgotten, but my mind was clouded with emotion. Struggling to recall I remembered, "leukocytosis, and fever qualifies her for SIRS, then she has a source of her infection; that's Sepsis." "Lactic acid of 10—, ok that is Severe Sepsis." "With three liters IV fluid, and pressures still dropping, currently 80/40— oh no, she's in Severe Sepsis."

GW was transferred to the ICU where the attending, a 30-year veteran, at one of the best hospitals in SLC came in to discuss how sick she was. Even with her symptoms, he was confident that they had isolated the cause of her issues and with the specified course of treatment he felt she would overcome her illness, though it would take time.

GW was now intubated on the highest settings, however her oxygen saturations were only in the low 90's. She was starting to swell from the sepsis, fluids, and copious amounts of norepinephrine they were pumping into her body.

I had seen people in the ICU with similar issues. Why was this different? Why was my brain so clouded? It was because the patient, GW, was Genell Wells, my mom.

After the attending visited with us about the course of treatment, I told my dad to go home and get some sleep, as it was going to be a long recovery. I took my mom's cold hand into mine and prepared myself for the sleepless night ahead.

I read the names of the drugs on the numerous pumps infusing clear liquid into my mothers' central line. Vancomycin, epinephrine, norepinephrine, D10, dopamine, LR. I felt a surge of adrenaline course through my body every time the monitors would beep. Why was I so hypocritically anxious? I had told patient's families countless times not to worry about these sounds, they were harmless, and they would quickly silence on their own. I read the EKG tracing, looking for any fluctuation from baseline. "Was that VTACH or a PVC? No, still Sinus Rhythm." I racked my brain. "How could she have been having fun bowling with her family the day before, and now be lying in a drug induced coma not able to breathe on her own?" I was exhausted. I kissed my mom's head, whispered how much I loved her, and reclined in the chair looking out to the moon and city light's as my eyelids closed.

No sooner had they closed than the monitor's alarms didn't silence, and shocked me back to consciousness. The lights were flipped on, and chaos ensued. The crash cart was brought in, and drugs and orders were given. I had seen a few codes during my training—and if I am honest—they were my

favorite part of medicine. The excitement of those previous experiences was contagious. I remember secretly hoping that I would be called on to give chest compressions or start an IV.

This time there was no excitement. Nausea, panic, and helplessness were the only feelings I experienced. After a couple of minutes she stabilized and people filtered out. The feelings however, did not leave.

I overheard the physician contemplating aloud with the resident why this was happening. They came to no conclusion. Then they left.

What had happened? I saw the monitors and read the numbers, but my training hadn't prepared me for this.

This continued hourly. The crowd and chaos would come and go without explanation as to why her pressures were bottoming out. More IV pumps were added, more drugs infused, more boluses and more antibiotics; but less answers, less sense, less understanding.

By 7 am the doctor told me to get my father in because my mother was not doing well. As my family arrived they began to ask the questions I had all night.

"What had changed?"

"Matt, why can't they give her something—some magic potion—to cure her?"

"Matt, you know what is wrong why don't you give them some advice on what they need to be doing."

As I am the only person in the medical field they relied on me to explain, as well as to accomplish the unachievable task of saving my mother.

The stress and anxiety were overwhelming; I took a walk to the bathroom. Prayers were said on the dirty bathroom floor between episodes of vomiting.

"Pull yourself together. You have seen people die before."

As I was huddled over the toilet, I thought about what the doctor would say when he updated us this morning. My memory flashed back to a month ago when I stood behind the resident while she explained to a family why Mr. C was not going to survive after a massive stroke to his median artery. I remembered how eloquently the resident gave bad news. I remember how I envied that she was able to be so articulate while giving the news to the patient's family. How they had no questions after...

I pulled myself together, wiped the vomit from my mouth, and walked back into the ICU room. The doctor was in the middle of explaining the overnight digression of my mom's health to my dad. I saw the doctor's mouth move and I knew words were coming out, but my brain wasn't processing it. Instead, I was selectively anticipating the inevitable bad news—it came. "She won't last through the morning."

Many times we, as medical students and physicians, get so caught up in the excitement and cognitive satisfaction of learning and practicing something new by working with critically ill patients. Yet, life has a way of teaching us what is really important. This unfortunate experience has helped me understand, first hand, how people feel during traumatic times in the hospital. At the same time, I also realize how important it is for me to be the doctor who is capable of giving the best care, without regret, and during difficult times, being empathetic to those who have to go through life changing experiences.

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