BREAKOUT
ROOM
NUMBER FIVE

CV 41 - 50
Moyamoya disease: More than just a “Puff of Smoke”
Hannah M. Conn M.D. and Christopher J. Haas M.D., Ph.D.

Moyamoya is a rare, poorly-understood, life-threatening cause of ischemic and hemorrhagic stroke that requires urgent neurosurgical revascularization for secondary stroke prevention. Here, we present a case of a young female that presented with hemorrhagic Moyamoya disease managed with indirect bypass and functional recovery.

A 35-year-old El Salvadorian female with no medical history presented in the context of obtundation. Vitals were unremarkable save hypertension (211/141) and physical examination notable for a GCS of 3, intact corneal reflexes, preserved pain response, right lower extremity triple flexion, and marked weakness of the left lower extremity. She was emergently intubated and nicardipine drip initiated. CT head demonstrated right-sided intraparenchymal, subarachnoid, and intraventricular hemorrhage, 9.7mm midline shift, and subfalcine, uncal, and cerebellar tonsillar herniation. CTA demonstrated bilateral MCA stenosis with collateralization between posterior and anterior circulations, suggesting Moyamoya vasculopathy. Emergent right-sided decompressive hemicraniectomy, intracerebral hemorrhage evacuation, and ventriculostomy catheter placement was performed. Hospital course was complicated by DVT necessitating IVC filter, PEG/tracheostomy placement, and simple seizure. She eventually underwent right-sided encephalo-duro-arterio-venous (EDAMS) indirect bypass surgery and cranioplasty. She was discharged to rehabilitation with excellent recovery. Repeat cerebral angiogram showed improved revascularization of the right posterior and frontal MCA territories.

Moyamoya is a rare condition characterized by bilateral steno-occlusive changes of the supraclinoid ICA with vessel collateralization that often presents with TIA or ischemic/hemorrhagic stroke. The true incidence and number of asymptomatic Moyamoya cases are unknown. While the underlying pathophysiology has not been fully elucidated, it is hypothesized that ischemic stroke is caused by vessel narrowing due to smooth muscle hyperplasia and luminal thrombosis, whereas hemorrhage is thought to occur secondary to collateral vessel rupture. Moyamoya management necessitates revascularization to stabilize native blood supply while promoting the regression of Moyamoya vessels via neurosurgical interventions - direct external-to-internal bypass, indirect bypass, and combination revascularization. Unfortunately, given the potential asymptomatic nature of the disease and lack of familial inheritance patterns, most Moyamoya cases declare themselves following a sentinel ischemic or hemorrhagic event.

Program Director’s Name: Stephanie Detterline MD

(indicating review of abstract)

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!
A DISSECTION TO REMEMBER: WHEN THE ABSENCE OF PAIN CAUSES DOUBT

Jennifer Rose F. del Castillo MD, Nahar Saleh MD, Rehan Farooqi MD

Aortic dissection (AD) poses an acute life-threatening risk to patients, especially when left unrecognized. We report an unusual presentation of a young patient who was found to have AD.

A 52 year-old female with tobacco, marijuana, and alcohol abuse presented with abdominal discomfort and constipation for two weeks. She attempted bisacodyl suppositories, fleet enemas and consumed four bottles of magnesium citrate without success. On presentation, she was hypertensive, but otherwise hemodynamically stable. Physical examination was notable for decreased bowel sounds over the left lower quadrant. Laboratories were unremarkable with the exception of a positive urine toxicology for cannabis, phenylcyclidine and opiates. Computerized Tomography (CT) of the chest, abdomen and pelvis revealed a type B AD involving the supra-celiac abdominal aorta extending into the distal abdominal aorta, with intramural hematoma extending from the subclavian artery continuing to the upper abdominal aorta. She was transferred to the ICU for blood pressure control with esmolol and diltiazem drips. Repeat CT scan unfortunately demonstrated an increase in the size of the aorta proximal to the diaphragm measuring 5.2 cm. Given the increase in size between scans and risk for rupture, the decision was made to repair with Thoracic Endovascular Aortic Repair (TEVAR).

AD is an intimal tear creating a false lumen (FL) through the aortic media. The incidence is 30 cases per million a year. Dissection may be classified depending on involvement of the aorta: Stanford Type A, which involves the ascending aorta and requires emergent surgery, and Stanford Type B, involving the descending aorta, and may be medically managed. Pain is the most common symptom in AD. Painless AD has been described only in type A with a prevalence between 3-7%. It presents as syncope, heart failure, or stroke. In general, painless AD patients are older (>65), diabetic, had antecedent aortic aneurysm or prior aortic valve repair. This is the first case of an atypically painless type B AD that presents as constipation. The pathological mechanism may be explained by mal-perfusion caused by the FL resulting to an altered perception of pain as the site or aortic innervation, adventitial layer, is involved by the dissection. This case reminds us that deadly diseases may hide in the guise of benign manifestations and present in unsuspecting patients without risk factors. Being mindful of clinically relevant differentials with keen consideration of the diagnosis regardless of presentation leads to timely diagnosis and prompt management.
MARY JANE – QUEEN OF BROKEN HEARTS: MARIJUANA-INDUCED TAKOTSUBO CARDIOMYOPATHY

Takotsubo cardiomyopathy (TCM) is a non-ischemic heart failure associated with transient reduced systolic function and characterized by regional wall motion abnormalities, specifically apical ballooning. While the exact mechanism remains unclear, a catecholamine surge is hypothesized to result in myocardial stunning. Marijuana (MJ) is a well-recognized and oft-used recreational psychoactive substance that has been associated with numerous cardiopulmonary complications. Recent evidence suggests an association between MJ use and TCM via activation of CB1 receptors.

A 60 year-old woman with a history of HTN, HLD, and COPD presented with lightheadedness of one day duration. Her symptoms were associated with nausea and shortness of breath, but no chest pain, edema, orthopnea, or paroxysmal nocturnal dyspnea. Social history was notable for a 40 pack-year smoking history, intermittent alcohol and MJ use, and no stressors. Vitals were unremarkable. Examination demonstrated bilateral wheeze and diminished air entry, with a normal cardiac examination. Diagnostics demonstrated elevated troponin, elevated lactate acid, and leukocytosis with neutrophil predominance. BMP and pro-BNP were unremarkable. ABG demonstrated combined respiratory and metabolic acidosis with severely reduced PaO2 and oxygen saturation. Urine toxicology was positive for MJ. Imaging was unremarkable. Though initial EKG was normal, she developed new-onset T wave inversions and was initiated on ACS protocol, with concern for evolving cardiogenic shock. Echocardiogram demonstrated reduced EF to 25% with multiple incongruent regional wall motion abnormalities. Catheterization demonstrated abnormal LV motion consistent with apical ballooning, reduced EF, elevated LVEDP, and non-obstructive coronary disease. She was diagnosed with MJ-induced TCM and discharged on optimal medical therapy with continued improvement over the next three months.

TCM is hypothesized to result from a catecholamine surge and it has been theorized that via the actions of THC on CB1 receptors, MJ use can exert a similar effect. CB1 receptors have been found not only within the HPA axis and amygdala, critical neural centers involved in stress responses, but also on cardiac tissues themselves, suggesting both indirect and direct actions on cardiac activity, respectively. Nevertheless, the exact mechanisms as well as individual patient susceptibilities remain to be elucidated.

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BREAKOUT ROOM 5
HUMAN HERPES VIRUS 6 ENCEPHALITIS COMPLICATED BY HEMOLYTIC UREMIC SYNDROME IN AN IMMUNOCOMPETENT PATIENT: Alnabulsi M, MBBS;
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Infections caused by Human Herpesvirus 6 infections (HHV-6) usually manifest as mild febrile illness with a mild rash, particularly in children. After primary infection, the virus can have a life-long latency in the human body. Upon reactivation, HHV-6 can have neurological complications, including febrile seizures, altered mental status, and encephalitis. Manifestations of HHV-6 infections tend to be more severe in immunocompromised hosts.

Hemolytic-uremic syndrome (HUS) and thrombotic thrombocytopenic purpura (TTP) are diseases associated with microangiopathic hemolytic anemia. HUS is divided into typical and atypical presentations; typical HUS is associated with infections from Shiga toxin-producing Escherichia coli O157:H7. While there have been case reports about unusual manifestations of HHV-6 infection, there have been no cases in the literature of HHV-6 primary or reactivation of a latent-infection associated with TTP or HUS.

A 79-year-old immunocompetent man with hypertension and hyperlipidemia was transferred to our facility for management of multiorgan failure. He initially presented with abdominal symptoms and was found to have pan-colitis on abdominal computer tomography. He was started on appropriate antibiotics. However, he continued to deteriorate clinically, and his hospital course was complicated by acute renal failure, thrombocytopenia, worsening anemia, altered mental status and seizures. He eventually required intubation for airway protection and was transferred to our facility. His brain imaging studies were negative for any acute processes. His lumbar puncture was positive for HHV-6. Additionally, his peripheral blood smear showed schistocytes. Hematology was consulted for concern of HUS/TTP and they recommended initiation of plasma exchange (PLEX) and steroids. Additionally, he was started on hemodialysis (HD) and Ganciclovir for HHV6 encephalitis. However, given the patient’s overall poor prognosis, the family expressed that they would like to pursue comfort care. The patient was extubated, HD, PLEX, and antivirals were stopped, and the patient expired shortly thereafter.

HHV-6 reactivation sequelae are not uncommon in the immunocompromised patient. However, it should be considered in immunocompetent patients as well. It has a wide variety of complications; some are more common such as encephalitis; others are highly unusual such as HUS.

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(indicating review of abstract)
ATYPICAL PRESENTATION OF H. PYLORI GASTRITIS IN PATIENT WITH CHEST PAIN. Iqbal T, MD. The University of Maryland School of Medicine and VA Medical Center, Baltimore, MD.

Chest pain is a frequent reason for admission to a hospital. The differential diagnosis is broad and includes cardiac, pulmonary, gastrointestinal, and psychiatric etiologies. The history and physical are usually able to delineate which etiology the chest pain originates from but it is sometimes difficult to ascertain.

A 57 year old man with history of human immunodeficiency virus (HIV) (stable, on antiretroviral therapy), essential thrombocytosis (chronic, on aspirin 325mg daily), hypertension, hyperlipidemia, and type 2 diabetes mellitus, presented to the emergency department with mid-sternal, non-radiating, exertional chest pressure. It had been intermittent for 2-3 days prior to presentation but worsened on day of presentation. On day of presentation, the patient described significant dyspnea on exertion with mid-sternal chest pressure when going up the stairs in front of his home, which was a change from his baseline. He is normally physically active and able to exert himself without limitations. In the prior week, he had episodes of diaphoresis with dyspnea, worsened by lying down flat. He had a chronic cough but no acute worsening. He reported occasional nausea. He had one episode of emesis with associated mild umbilical pain a few days prior to admission after taking medication, but the emesis resolved. He denied fevers, weight loss, paroxysmal nocturnal dyspnea, sick contacts, or recent travel. He was taking his medications as prescribed. He smoked cigarettes intermittently and smoked 3 joints of marijuana per day.

He was admitted to rule out acute coronary syndrome. EKG on admission showed normal sinus rhythm with T wave inversions in leads V5-V6, which were chronic. Serial troponins were negative (<0.02 ng/mL) times three. On examination, the patient became diffusely diaphoretic if he reclined less than a 45-degree angle. Cardiovascular exam was otherwise normal. D-dimer was elevated, but subsequent computed tomographic angiography of the chest was negative. Differential diagnosis also included gastroesophageal reflux. The patient's symptoms completely resolved after being given a dose of pantoprazole 40mg orally. H. Pylori stool antigen test was positive.

While patients with characteristic coronary artery risk factors (i.e. smoking, hypertension, hyperlipidemia, diabetes and HIV) should be ruled out for cardiac etiologies of chest pain, atypical presentations should also increase suspicion for alternative etiologies.
Takotsubo Cardiomyopathy after Influenza Infection: A Rare Case

Introduction: Takotsubo cardiomyopathy, also called "broken heart" syndrome or apical ballooning syndrome, is a reversible cardiomyopathy characterized by left ventricular dysfunction and ballooning of the left ventricular apex on imaging during systole. Although influenza is a known cause of myopericarditis, it has only been implicated in a few case reports as the cause of takotsubo cardiomyopathy.

Case Presentation: A 67-year-old woman with past medical history of diabetes mellitus, hypertension, depression, anxiety, and bipolar disorder initially presented with cough, body aches and back pain. At that time, her examination revealed decreased breath sounds bilaterally and bilateral flank tenderness. She was diagnosed with influenza with a positive rapid influenza A test, and urinary tract infection with a positive urinalysis. She was then discharged home on cefdinir and oseltamivir.

Two weeks later, she experienced squeezing chest pain that was associated with walking and relieved by rest. She denied palpitations, cough, or sputum production. There is a remote history of intravenous drug use 14 years ago. She smokes 1.5 packs of cigarettes per day and has 2 alcoholic drinks per week. Physical examination showed a distressed female moaning in pain. Except for BP of 184/117 mm Hg, remainder of the examination was unremarkable.

Clinical Course: Initial EKG showed new T wave inversion and loss of R waves in leads V1-V3. Initial Troponin was 0.599 ng/mL, which up-trended to 1.40 ng/mL. CTA chest and abdomen were negative for aortic dissection, pulmonary embolism, or pericardial effusion, but it did show hepatic venous congestion. She was started on heparin infusion per ACS protocol. A transesophageal echocardiogram (TEE) revealed normal LV size with severely decreased systolic function, global hypokinesis with sparing of the inferolateral wall, and left ventricular ejection fraction of 30%. A left heart catheterization the following day revealed minimal non-obstructive CAD, but severe apical hypokinesis and hyperdynamic basal segments, consistent with stress-induced cardiomyopathy. Heparin was stopped in favor of high dose atorvastatin, lisinopril, and metoprolol, with subsequent improvement in her chest pain.

Conclusion: Takotsubo cardiomyopathy can be a rare complication of influenza infection. It should be considered in patients presenting with signs and symptoms of acute coronary syndrome in the setting of influenza infection.
NEW STEPS: REGAINING AMBULATION AND IMPROVED PREPARMENT STATUS FOLLOWING DIAGNOSIS AND TREATMENT OF PARANEOPLASTIC MYASTHENIA GRAVIS
Stephen Njau MD, Patrick Bagley DO, Jeffery Gray, MD.
National Capital Consortium/WRNMMC

INTRODUCTION: Paraneoplastic syndromes (PNS) affect 6% of cancer patients, often in small cell lung cancer. PNS with prostate cancer typically include cerebellar degeneration, subacute sensory neuropathy, and encephalomyelitis. These syndromes are the first signs of progression to castrate resistant disease in 20% of cases, yet they are often underappreciated.

CASE: A 68-year-old African American man with metastatic castrate-resistant prostate cancer status-post six cycles of docetaxel three years prior to presentation was admitted from oncology clinic for malaise. Despite living one block from the clinic, the patient missed appointments due to difficulty ambulating. He was deemed to have a poor performance status and was not a candidate for further treatments. Upon admission, he had severe lumbago and electrolyte abnormalities. Spinal myelopathies were ruled out with CT and MRI, and his pain was controlled, but he was still unable to ambulate. Neurology consultants elicited length-dependent sensorimotor polyneuropathy and proximal predominant weakness. EMG was performed which demonstrated decrement on repetitive stimulation without evidence of myositis. These findings were concerning for a post-ganglionic neuromuscular junction deficit consistent with myasthenia gravis; however, anticholinergic receptor binding antibodies and striated muscle antibodies were negative. He was diagnosed with seronegative myasthenia gravis. His symptoms markedly improved with initiation of pyridostigmine, as he regained the ability to ambulate and participate in physical rehabilitation. His performance status improved to the point of reconsidering treatment options going forward.

DISCUSSION: This case highlights the importance of appreciating PNS in metastatic prostate cancer. This case exemplifies diagnosis of a rare, albeit treatable, condition that significantly improved this patient’s quality of life and potentially enabled to him to receive additional chemotherapy going forward. Further, neuroendocrine prostate has similar morphology to small cell cancer and is increasingly being associated with ADT resistant metastatic cancer.

CV 47

BREAKOUT ROOM 5
MEETING THE MINOCA QUOTA

Introduction: Acute coronary syndrome is one of the most common causes of inpatient admissions. Myocardial infarction with non-obstructive coronary arteries (MINOCA), however, is a relatively uncommon cause of chest pain with distinct management strategies.

Case Description: A 53-year-old woman with a history of migraines and hypertension presented with acute, substernal chest pain that radiated to her left arm. Her pain was associated with nausea, dizziness, and diaphoresis. She denied fever, cough and shortness of breath, but she was experiencing a migraine at the time. She had no history of tobacco, alcohol, or drug use. She had a family history of sudden cardiac death. Her vital signs were stable, and she had no evidence of volume overload on exam. Her complete blood count, metabolic panel, and lipid panel were normal. Troponins peaked at 5.6 ng/mL and Pro-BNP was 1144 pg/mL. Her electrocardiogram (EKG) demonstrated ST-segment elevations in the anterior leads with reciprocal ST-segment depressions. She received aspirin 325 mg and a loading dose of ticagrelor, and she was started on a heparin drip in the Emergency Department. She also received sublingual nitroglycerin which quickly relieved her chest pain. She was urgently taken for a left heart catheterization, which did not show any significant coronary artery disease (CAD). A trans-thoracic echocardiogram demonstrated a normal ejection fraction with no wall-motion abnormalities. She was discharged on aspirin, ticagrelor, atorvastatin, and ezetimibe, and she was continued on her home amiodipine. She was scheduled for close cardiac follow up and cardiac rehabilitation.

Discussion: This patient was diagnosed with MINOCA, given clear evidence of myocardial infarction on laboratory and EKG testing in the absence of CAD on angiography. There are numerous underlying causes of MINOCA, with coronary spasm being the most likely cause for this patient. She met two out of three criteria for vasospastic angina: her chest pain was nitrate-responsive and she had localized EKG changes during her episode of chest pain, but she did not have angiographic evidence of spasm. She also had a history of migraines – another vasospastic disease. Other common causes of MINOCA, such as Cardiac Syndrome X and Takotsubo cardiomyopathy were less likely due to a significant troponin elevation and lack of echocardiogram findings on admission, respectively. Since additional causes of MINOCA, especially acute thrombosis, could not be ruled out, aggressive risk-reduction measures were taken on discharge.

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!
Importance of Immunosuppressive therapy for Managing Insulin Resistance type B
Samuel Amankwah M.D., Artit Silpasuvan M.D.

Background: Autoimmune Antibodies against insulin receptors leading to refractory hyperglycemia is known as Type B insulin resistance. In addition to insulin management, immunosuppressive therapy appears to be an essential part for successful management.

Clinical Case: A 20 year old African American woman with no significant past medical history presented to the Emergency Department with five days of worsening nausea, vomiting and shortness of breath. She admitted to polyuria, unintentional 15 lb weight loss over six months, decreased appetite, and hyper-pigmented rashes predominantly on her back. She was not on any medications and had no known drug allergies. Family history was significant for grandmother with Type 2 Diabetes Mellitus and Lupus on father's side of family. The patient denied use of alcohol, tobacco or illicit drugs.

The patient was afebrile, normotensive but had sinus tachycardia. Upon initial work up, she was found to have new onset diabetes with HbA1c of 9.6% and a massive pericardial effusion on echocardiogram for which she underwent pericardiocentesis. Due to developing complaints of intermittent cyanosis of fingertips and intermittent joint pain of fingers, rheumatology work up was ordered and returned positive for mixed connective tissue disease. Patient was treated with Cellcept 1500 mg BID, Plaquinil Sulfate 300 mg daily, and prednisone 12.5 mg BID.

Hospital course was complicated due to hyperglycemia in 300s-400mg/dL requiring high amounts of insulin.

Diabetes work up was negative for GAd-65 and islet cell antibodies. C-peptide was elevated at 16.1 ng/mL (0.8-3.1 ng/mL) and anti-insulin antibodies were 20uU/mL. Patient was euthyroid.

Hyperglycemia persisted despite increasing doses of long and short acting insulin subcutaneously. The patient eventually required insulin intravenously at 50 units/hour plus 50 units of short acting insulin every 4 hours subcutaneously. The patient was transferred to a facility to begin combined immunosuppressive therapy plus insulin to manage hyperglycemia due to insulin resistance. After six months of rituximab, pulsed steroids, cyclophosphamide, and insulin therapy, her glycemic index improved with HbA1c reduced to 5.6%.

Conclusion: Combined immunosuppressive therapy in addition to insulin management of refractory hyperglycemia due to type B insulin resistance has been shown to not only be effective in controlling refractory hyperglycemia but preventing against recurrences as well.
Spontaneous Coronary Artery Dissection in an Active Duty Female

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**Introduction:** Spontaneous coronary artery dissection (SCAD) is an important cause of myocardial infarction (MI) in patients without typical coronary risk factors. Pathology of SCAD involves the formation of a false lumen within the artery secondary to disruption of the layers of the artery wall. This disruption leads to the formation of an intramural hematoma, occlusion of the true lumen, and potential infarction. Predisposing factors include vasculopathies and hormonal influences. First line treatment remains medical management.

**Case Report:** A 43 year old female with a past medical history notable only for a ten pack year smoking history presented to the emergency department with exertional chest pain and left arm discomfort that was relieved by rest for 72 hours. Physical exam was unremarkable, but EKG was notable for Wellen’s T-waves in V\textsubscript{2} and V\textsubscript{3} with T wave inversions in V\textsubscript{4}-V\textsubscript{6}. Lab values revealed high sensitivity troponin of 69.4 ng/L and leukocytosis of 11.1 x 10\textsuperscript{9}/m\textsuperscript{L}. She was admitted and diagnosed with non-ST segment elevation MI (NSTEMI). Coronary catheterization revealed a linear filling defect in the mid left anterior descending (LAD) artery with thrombolysis in MI (TIMI) 2 flow distally, consistent with dissection. She was treated medically with dual antiplatelet therapy, low dose beta blockers, anti-anginal therapy and was recommended for further evaluation of underlying conditions that may have contributed to her presentation.

**Discussion:** SCAD is a cause of MI seen at higher rates in women younger than 50 who lack typical coronary risk factors. Presentation is clinically similar to MI caused by atherosclerotic plaque disruption, with chest pain and EKG findings suggestive of infarction. Treatment currently favors medical management due to increased risks associated with percutaneous intervention, notably propagation of coronary dissection. However, stent placement may be indicated in certain clinical scenarios, such as patients with SCAD in the setting of ST elevated MI (STEMI) or hemodynamic compromise. Potential infarction size after SCAD correlates with factors such as whether or not the infarct resulted in STEMI, TIMI flow value, and number of vessels involved. Suspicion for SCAD should be heightened in female patients ≤50 who present with STEMI/NSTEMI, without typical risk factors, such as the young active duty service members that comprise a large portion of our patient population at Walter Reed Nation Military Medical Center.