

Clinical Vignette Symposium 2024

Book of Abstracts

University of Oklahoma –Tulsa School of Community Medicine

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Dear Colleagues,

It is my pleasure to welcome you to the 13th annual Clinical Vignette Symposium (CVS) hosted by the OU School of Community Medicine. At CVS, trainees present interesting and unusual medical case studies in a conference setting. We look forward to learning the stories behind these unique case reports.

In addition to poster and podium presentations at CVS, authors have an opportunity to upload their work to the Open Science Framework (OSF). Posters and podium presentations uploaded to OSF will be more widely disseminated to a global community. Authors will also be able to include these presentations as citations on their Curriculum Vitae. Awards will be given to the top scoring posters. These awards may be used toward travel or publication fee expenses to further disseminate their scholarly work.

I would like to thank the Tulsa County Medical Society for their generous donation to this year's event. I would also like to thank the presenters as well as those who organized and are hosting CVS this year. We hope you enjoy CVS 2024.

Sincerely,

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ORDSA would like to thank all faculty and staff who contributed their time and energy to organizing the 2024 Clinical Vignette Symposium. ORDSA would also like to thank the library for their assistance. Finally, ORDSA would like to thank the Tulsa County Medical Society for sponsoring this event and the OUSCM faculty who provide financial contributions to it.

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Oral Presentations

<u>Abstract #3</u> Nasal Polyps Leading to Newly Diagnosed Cystic Fibrosis in an Adolescent

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Introduction

Cystic Fibrosis (CF) is a genetic disorder characterized by various pathogenic mutations in the CFTR gene, typically diagnosed by newborn screening. We present a case in which two rare CFTR mutations eluded routine newborn screening but were identified during the investigation of multiple nasal polyps, emphasizing the importance of remaining vigilant in the face of atypical clinical presentations.

Case Description

A previously healthy 14-year-old white adolescent presented with congestion refractory to conventional treatment. Physical exam revealed a large, right nasal polyp. The adolescent was referred to ENT and started on dupilumab. After dupilumab failed to improve symptoms, a CT sinus without contrast revealed multiple sinonasal polyps involving the bilateral maxillary sinuses and nasal cavity prompting work up for CF. Newborn metabolic screen revealed an elevated IRT of 204.5 ng/mL with a subsequent reflex genetic panel being negative for common CF mutations, and therefore, sweat chloride testing was deferred at that time. For concern of CF given nasal polyposis with history of abnormal newborn metabolic screen, further CF expanded variant panel analysis was completed and revealed two rare CFTR mutations, p.Ser492Phe and c.2490+1G>A, that have never been reported to be occurring simultaneously in one individual. Sweat chloride values of 76 mmol/L and 90 mmol/L on the right and left arms confirmed the diagnosis of CF. Consequentially, fecal elastase and pulmonary function testing were completed and found to be within normal limits. Her nutritional status was normal and lab testing was unremarkable with only a mild vitamin D deficiency.

Discussion/Conclusion

The presence of nasal polyps in this case prompted a broader diagnostic workup, revealing two rare CFTR mutations. This case highlights the critical role of clinical vigilance beyond routine newborn screening in diagnosing CF, particularly when faced with atypical presentations such as nasal polyposis in the setting of an undetected genetic mutation on newborn screening. The awareness that a negative newborn screen may miss some CF diagnoses underscores the need for a high index of suspicion and thorough evaluation in the presence of persistent or unusual symptoms, enabling timely diagnosis and intervention for improved patient outcomes.

<u>Abstract #71</u> Hyperthermia: A Unique Presentation of Sympathomimetic Toxicity

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Introduction

While sympathomimetic syndromes and hyperthermia are common emergency room visits individually, hyperthermia from sympathomimetic toxicity is far less common. This case highlights the presentation, diagnosis, and treatment of a hyperthermic patient secondary to sympathomimetic toxicity.

Case Description

A 38-year-old male presented to the emergency department via emergency medical services after being found in a car with altered mental status and diffuse tremor like movements. He was able to answer some questions initially but became obtunded just prior to arrival to the ED. EMS noted the patient to be tachycardic with a heart rate of 180 bpm and hypotensive with systolic blood pressure of 70 mmHg. The patient was intubated upon arrival to the ED for airway protection after administration of phenylephrine for blood pressure support. His initial temperature was found to be 42.5 degrees Celsius (108.5 degrees Fahrenheit) with outside temperatures around 50 degrees Fahrenheit. Ice packs were placed around the torso, axilla and groin and the Artic Sun external cooling device was applied. The patient was given two liters of normal saline, Ativan, and dantrolene IV for suspected malignant hyperthermia of unclear etiology. Significant laboratory abnormalities included, creatinine of 2.0 dl/ mL, CK of 6900, troponin of 0.3 ng/mL, venous blood gas showing metabolic/respiratory acidosis, and lactic acid of 8.7. CT head showed no acute abnormality and urine drug screen was positive for amphetamines and cannabinoids. It was later reported the patient had an argument with his significant other and planned to overdose on methamphetamine. The patient was stabilized in the Emergency Department and was admitted to the ICU. After a prolonged hospital course requiring dialysis from renal failure secondary to rhabdomyolysis, the patient returned to his neurological baseline and was discharged home.

Discussion/Conclusion

Sympathomimetic toxidromes are commonly seen in the emergency department, but malignant hyperthermia is a rare presentation. Potentially lethal hyperthermia is thought to be caused by both central and peripheral processes as well as decreasing heat dissipation1. Early recognition is vital to improving patient outcomes and decreasing mortality.

Poster Presentations

Emergency Medicine

<u>Abstract #12</u> Gastroduodenal Artery Stump Bleed Requiring Massive Transfusion in the ER

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Introduction

The Whipple procedure or pancreatoduodenectomy is a surgical procedure used in the treatment of malignant neoplasms of the pancreas. Whipple procedures were initially associated with a high mortality rate, but mortality has improved1 over time. Complications still arise, including pancreatic fistulas, hemorrhage, infection, enteric leaks and delayed gastric emptying. Gastroduodenal stump bleeding is the most concerning hemorrhagic complication.

Case Description

This case concerns an 80-year-old male who had a Whipple procedure. He was admitted several days post procedure for evaluation of an anastomotic leak. After discharge from observation admission, he presented to outlying facility with hematemesis. Hemoglobin fell from 11.9 to 9.3 during transfer to our facility. The patient was resuscitated with matched packed red blood cells and was initially admitted to the medical floor. Hepatobiliary surgery reviewed outside CT and was not immediately concerned for acute bleed. He then developed large volume hematemesis, becoming hypotensive, altered, and tachycardic. Mass transfusion was initiated. The patient received 14 units of blood, was intubated and a Cordis placed. Pressure support was initiated with norepinephrine and phenylephrine. Gastroduodenoscopy was attempted in the ER, but unsuccessful due to massive hemorrhage. Interventional Radiology was consulted, identified a gastroduodenal stump bleed, and successfully placed a coil. Bleeding was controlled with IR intervention. The patient was noted to have favorable anatomical variant for collateral blood distribution. The patient was stabilized and admitted to the ICU. His course improved and he was downgraded to the medical floor and eventually inpatient rehab and discharge.

Discussion/Conclusion

Post operative hemorrhage is a feared complication of most surgical procedures. Providers should not hesitate to initiate care for massive hemorrhage. Mass transfusion is defined as ten or more units of blood in 24 hours. The primary objective of mass transfusion is to prevent fatal outcomes from critical hypoperfusion2. An ABC score is used to drive mass transfusion initiation but should be used in conjunction with clinical judgement. The ABC score accounts for penetrating mechanism, hypotension, tachycardia, and a positive FAST bedside ultrasound study. Admittedly a FAST was not performed in this case due to CT findings already were suggestive of possible bleeding. In this case the patient was hypotensive and altered but did not meet other ABC criteria. Given his history suspicion of a life-threatening exsanguination event was high. So, there was little lag time in ordering mass transfusion given clinical suspicion previously in place.

<u>Abstract #17</u> Buprenorphine Extended-Release Subcutaneous Injection as a Cellulitis and Abscess Mimic

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Introduction

Buprenorphine is a partial mu-opioid agonist used to treat moderate-severe opioid use disorder, now available in an extended-release subcutaneous injection. While several formularies of buprenorphine are available, there are many barriers to adequate treatment for this nationwide epidemic including medication adherence, accessibility, abuse and subtherapeutic dosing particularly with daily or weekly doses. Extended release buprenorphine injection is administered once monthly which may address some of these treatment gaps. Despite being FDA approved in 2017, it is considered to be a novel therapy with adverse reactions that not all healthcare providers may be familiar with.

Case Description

A 35-year-old male with opioid use disorder, on the buprenorphine extended-release injection, Sublocade, presented to the Emergency Department for pain, erythema and swelling at his injection site. The patient received his first Sublocade injection in prison 14 days prior to presentation. Shortly after injection, he developed a firm subcutaneous abdominal mass at the site which worsened over the next few days becoming erythematous and painful. Additional symptoms during this time include chills, nausea and vomiting.

On evaluation, physical exam revealed a 3 cm x 3 cm indurated, erythematous, non-fluctuant, mobile, mid-abdominal mass, lateral to the umbilicus. Additional skin changes were visualized surrounding the mass. Bedside ultrasound was performed to evaluate for abscess as this diagnosis was high on the differential. Soft tissue edema without a drainable fluid collection was visualized. Incision and drainage of the mass was considered but not felt appropriate due to concern for precipitating opioid withdrawal or potential overdose by releasing all contents of the depot. Inflammatory reaction to the medication was felt the most likely diagnosis. Due to concern for developing cellulitis, the patient was discharged with a paper prescription for cephalexin and educated on the signs and symptoms of worsening infection. He was directed to fill the prescription if his symptoms progressed.

Discussion/Conclusion

Studies have shown that the most common side effect seen with buprenorphine extended-release subcutaneous injections, like Sublocade, are injection site reactions. Although typically benign, knowledge of this potential reaction is beneficial to both patient and healthcare providers as the symptoms mimic cellulitis and abscess. Current literature does not identify the potential outcomes of misdiagnosis leading to incision and drainage of the buprenorphine extended-release injection depot. This case study highlights the need for both patient and provider education on adverse reactions to this novel therapy to prevent unnecessary harm.

<u>Abstract #36</u> latrogenic Injury due to Improperly Placed Prehospital Intraosseous Device

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Introduction

Rapidly obtaining vascular access in critically ill patients can be difficult, especially in the prehospital setting. An intraosseous (IO) device allows vascular access by placing a needle through the cortex and into the medulla of a bone. The proximal tibia, proximal humerus, and sternum, among others, are all suitable for IO placement. Success rates range from 40% to 96% across a broad range of devices and practice settings. Adequate training and proper technique is necessary, since improper placement can lead to osteomyelitis, fracture of bone, and extravasation. Extravasation of large amounts of fluids can progress to compartment syndrome3. Here we describe a rare, transient compartment syndrome from iatrogenic IO misplacement.

Case Description

A 70-year-old male presented to the emergency department by emergency medical services (EMS) for a syncopal episode. The paramedic reported that the patient was hypotensive upon arrival with systolic blood pressure 70 mmHg. Intravenous access was unable to be quickly obtained, so an IO was placed in the left lower leg and one liter of normal saline was infused through it. Upon arrival to the emergency department, the left lower leg was significantly more swollen than the right lower leg, suggesting that infiltration had occurred. The IO device was expeditiously evaluated and it was noted to be improperly positioned medial to the tibia so it was immediately removed. The leg was tense to palpation, painful, and cold. Dorsalis pedis pulse was unable to be located by palpation or doppler. Compartment pressure was measured at 74 mmHg. Warm blankets were applied to lower extremities and orthopedic surgery was consulted. Fasciotomy was not felt to be indicated. Patient was hospitalized for evaluation of syncope. Swelling gradually decreased, symptoms improved, and patient was discharged home in stable condition after a two-day hospitalization.

Discussion/Conclusion

This case highlights the importance of recognizing iatrogenic injuries early. Thankfully, the improperly placed IO was identified immediately upon arrival to the emergency department. It was removed before any additional fluid was infused into the soft tissue compartment. Prompt evaluation of pressures and consultation of the appropriate specialist led to a positive outcome without further trauma to the lower extremity. The chief complaint of the patient was shifted briefly as the potential compartment syndrome was more time-sensitive than evaluation of syncope.

Abstract #66 Accidental Cannabis Consumption in a Pediatric Patient

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Introduction

In 2022, 583 cases of accidental cannabis consumption were reported to the Oklahoma Poison Center, most involving edibles and around half in children < 6 years old1. Since many cannabis edibles look and taste like regular consumer products, accidental ingestion is common, especially in children. Patients and parents may also be reluctant to disclose the possibility of cannabis ingestion because of fear of repercussions.

Case Description

Patient is a 7-year-old male with no past medical history who presented to the Emergency Department with mother for acute onset of headache, dizziness, vomiting, ataxic gate and pallor. Patient was well prior to symptom onset. Described as sudden onset with headache and dizziness preceding vomiting. Mom denied fevers, recent illness or possibility of accidental ingestion. Initial vital signs were WNL. Physical exam significant for pale, ill-appearing boy who was slow to answer questions. Neurological findings included horizontal nystagmus, abnormal finger-to-nose and equivocal pronator drift. POC VBG showed mild metabolic acidosis of 7.32 with a lactate of 4. Fluid bolus of 20ml/kg initiated. A CBC, CMP, CRP, blood culture, viral panel, UA, UDS, toxicology level, CT head, and abdominal ultrasound were all negative except positive findings for rhinovirus and enterovirus. Pediatric Neurology consulted and suggested CTA head and neck (also negative) with MRI of brain to follow to rule out posterior stroke. On re-evaluation his exam had not changed. Mom was consented for lumbar puncture. Just prior to procedure, UDS positive for cannabinoids. He was admitted to medical floor for observation. Supportive care led to discharge without lasting effects. It was later discovered relatives who had visited that day had edibles the patient ingested thinking they were a normal consumer product.

Discussion/Conclusion

Cannabis intoxication can present with many signs, from mild symptoms like dry mouth, vertigo, and vomiting to severe symptoms such as respiratory depression leading to intubation2. This patient's symptoms drew concern to clinicians for life threatening illnesses. The job of emergency medicine clinicians to rule out these conditions can lead to an extensive work up and painful procedures. In this case, early collection of urine could have led to less testing. It is important for clinicians to have a high index of suspicion for cannabis ingestion, and to reassure families that a search for cannabis intoxication is medical, not punitive, in nature.

Family and Community Medicine

<u>Abstract #37</u> Misdiagnosed Meningioma Masquerading as Herniated Nucleus Pulposus

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Mr. Mataeo Anderson - OU-TU School of Community Medicine

Dr. Casey Smith - Tulsa Bone and Joint

Dr. Jason Deck - OU-TU School of Community Medicine, Department of Family & Community Medicine

Dr. Sarthak Parikh - Saint Francis Health System

Introduction

Spinal cord meningiomas are tumors arising in the meninges surrounding the spinal cord. There are rare cases of them presenting similarly to herniated discs, which occur when the nucleus pulposus bulges out of the annulus fibrosus. Both meningiomas and herniated discs can present clinically with radiating radicular pain, often with associated paresthesia.

Case Description

A 32-year-old male, with no past medical history, presented to an orthopedic clinic with lower back pain, radiating down his left leg. MRI imaging without contrast demonstrated a left paracentral disc herniation at L5-S1, per the radiologist. Epidural injections were administered three months after his initial clinic visit. Two weeks after the injections, he reported no change in pain. He was seen in clinic three weeks later where it was determined that surgery would be performed the following day. During surgery, a vascular mass above the L5-S1 area had to be removed which pathology determined to be an angiolipoma. As the surgery progressed, no evidence of disc herniation was found, but as tissue was retracted, a tumor was identified. The operating physician performed a full laminectomy to relieve pressure, but the tumor was not resected at that time. Patient followed up two months post-op where he reported minimal pain with activity in his lower back but overall felt better and denied any radiating pain in his legs. An MRI with contrast was ordered one month after the operation but performed two months later. It showed an extramedullary mass, specifically a meningioma with little enhancement or enlargement but also showed an anterior disc bulge and signs consistent with early degenerative disc disease. Two months later, he returned to the clinic for episodic recurrent back pain that lasted three to five minutes and new onset spasms of his left leg. To combat the arthritic pain, the patient was referred to neurosurgery. Upon evaluation, neurosurgery performed an anterior lumbar interbody spinal fusion at L5-S1 to reduce arthritic pain. The patient has not returned to the orthopedic clinic since and there have been no new updates regarding his condition.

Discussion/Conclusion

This case was unique because the presentation was consistent with a herniated disc but was in actuality a meningioma. Often, sequestered herniated discs are misdiagnosed as extramedullary tumors due to fragments of the disc migrating through the epidural space, but this case was the opposite. On the other hand, literature regarding cases of meningiomas mimicking disc herniations is sparse.

Abstract #38 Unusual Radiographic Profile of Meningioma Leads to

Misdiagnosis

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Introduction

A meningioma is a tumor of the meninges that can present with a variety of symptoms, including radiculopathy, back pain, and paresthesia. Extra-dural tumors and disc herniations can be difficult to differentiate as they often present similarly on clinical exam and imaging due to spinal impingement. To accurately make the diagnosis an MRI without contrast is usually sufficient. However, if a tumor is suspected, MRI with contrast is more sensitive and specific. This case report recounts the preoperative misdiagnosis of a patient suspected to have a herniated disc, whose surgical dissection revealed a meningioma.

Case Description

A 64-year-old male presented to an orthopedic spine clinic with low back pain and bilateral lower extremity radicular pain that began several years ago and worsened with time. In clinic he rated his pain at 15 out of 10. An MRI without contrast was ordered three months prior to the visit. The radiologist's impression was a herniated nucleus pulposus at the L3/L4 level causing central and lateral recess stenosis and central canal stenosis. He had failed to gain relief through conservative measures, such as physical therapy and lumbar facet injections. After failing conservative therapy, lumbar diskectomy at the L3/L4 level was recommended. After dissection through the deep fascia to the disc space, there was no evidence of disc herniation. Upon further examination, the dura was observed to be white and thickened along the wall. Neurosurgery was consulted intra-operatively and suggested the pathology resulted from a ventral, dural-based mass, likely a meningioma. Due to the dural thickening and ventral dura-based location of the tumor, no attempt was made to excise the tumor. A full laminectomy at L3/L4 was performed to relieve central stenosis and allow space for the spinal cord posteriorly without impingement. MRI with contrast the following day confirmed meningioma. The patient was seen back in clinic three months post-op with repeat imaging showing no evidence of a residual lesion. The patient also reported significant clinical improvement.

Discussion/Conclusion

This patient is unique because normally if a misdiagnosis is made it is disc herniations being misdiagnosed as tumors, and 75-90% of spinal meningioma's occur in females, not males. It is also interesting that the tumor disappeared on follow up. Surgeons should be cognizant of misdiagnosing tumors as disc herniations or vice versa and consider including both on a differential. Available tools, like MRI with contrast, can allow better differentiation for final diagnosis.

Abstract #42 Neuropsychiatric Manifestations in Systemic Lupus Erythematosus

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Dr. Crysta Chatman - OU-TU School of Community Medicine, Department of Family & Community Medicine

Introduction

Systemic lupus erythematosus (SLE) is a multifactorial, type III hypersensitivity disorder where the immune system mistakenly forms an inflammatory response against the body's healthy tissues. In SLE, immune complexes are deposited on various tissues and organs, leading to diverse clinical manifestations affecting the skin, joints, kidneys, and other organs. Altered mental status in SLE can be multifactorial, stemming from the direct effects of the disease on the central nervous system and the secondary effects of vasculitis.

Case Description

A 23-year-old female with a history of iron deficiency anemia requiring blood transfusions presented with myalgias and heavy vaginal bleeding – a clinical tapestry indicative of systemic SLE. She was worked up for SLE and was positive for dsDNA, low complement levels, and a critically low platelet level of 23. She was admitted to the hospital for idiopathic thrombocytopenia (ITP) and received steroid treatment. Over the next month, she was prescribed hydroxychloroquine for her SLE but did not start it. A subsequent admission for ITP led to a steroid taper; however, the patient left against medical advice. One week later, she presented to the emergency room with altered mental status and thoughts of harming others. Initial labs were unremarkable, and a urine drug screen was positive for marijuana. The toxicology screen and medication non-compliance did not fully coincide with the complexity of her altered mental status. An MRI was performed with no remarkable findings except mild left sinus disease. She was initiated on Solu-Medrol and transitioned to Prednisone 60 milligrams daily. Psychiatry evaluated her and started her on Zyprexa, and her mental state improved. She was discharged on Zyprexa and Prednisone taper and continued to take these medications as prescribed outpatient with a resolution of psychosis symptoms.

Discussion/Conclusion

This case illustrates the underscored association between psychiatric symptoms and immunological disorders. The complex presentation of SLE involves the central nervous system, encompassing a spectrum of psychiatric symptoms ranging from mild cognitive dysfunction to severe manifestations such as psychosis. Autoantibodies, such as anti-nuclear antibodies, and the formation of immune complexes contribute to vascular and inflammatory processes and potentially disrupt normal brain function. The current literature includes abnormal magnetic resonance imaging in patients with SLE. The heterogeneity of these interactions reveals the challenge of unraveling the mechanisms behind lupus-associated psychosis. Clinically, early recognition and intervention are crucial for effectively managing neuropsychiatric symptoms, highlighting the need for a multidisciplinary approach to address this comorbidity.

<u>Abstract #53</u> Diagnosing Superior Mesenteric Artery Syndrome in a Patient with Chronic Gastrointestinal Symptoms

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Introduction

Superior mesenteric artery syndrome (SMAS) is an under diagnosed condition characterized by vascular compression of the duodenum leading to nonspecific gastrointestinal symptoms. It results from a narrowing of the aortomesenteric angle, either congenital or acquired and particularly following sudden, extreme weight loss.

Case Description

A 35-year-old male with a history of scoliosis and previous small intestinal bacterial overgrowth (SIBO) presented to a primary care clinic with chronic bloating and difficulty gaining weight. Four years prior, he was hospitalized for SIBO, protein-calorie malnutrition, and a recorded BMI of 17.04 kg/m2. Despite treatment with antibiotics and follow-up recommendations, the patient developed persistent bloating and difficulty gaining weight only reaching a BMI of 17.39 kg/m2.

Prior documentation showed extensive benign results, including esophagogastroduodenoscopy, colonoscopy, anal manometry, celiac panel, stool tests for ova and parasites, and occult blood. Nuclear medicine gastric emptying study demonstrated gastric retention slightly greater than acceptable limits at one and two hours but within acceptable limits at four. He had no previous surgeries, reported intermittent use of milk of magnesia for bloating and constipation, and did not take any medications. Repeat CBC, CMP, and TSH were unremarkable. A thorough review of systems, past medical, family, and social history did not suggest any autoimmune, social, or psychiatric etiologies.

The patient was instructed to begin a daily dietary log and pause any supplemental intake. Upon his return, the food log revealed a balanced diet, averaging 1750 to 2500 kcals daily, coupled with moderate level of physical activity. Given the persistent nature of his symptoms and extensive rule out of common gastrointestinal disorders, a diagnosis of SMAS was considered. An MRI abdomen revealed Abdominal Aorta to SMA distance of 5-6 mm (normal > 10 mm) and aortomesenteric angle < 25° (normal range 38°-65°), compressing both the patient's duodenum and left renal vein.

Discussion/Conclusion

SMAS is prevalent in individuals with altered spinal anatomy (e.g., scoliosis) or following significant weight loss. As a diagnosis of exclusion, especially in the primary care setting, more common pathologies that lead to difficulty gaining weight should be considered, such as an absorptive disorder (e.g., IBS, Crohn's, and Ulcerative Colitis) or eating disorders. The condition often requires a high index of suspicion and a comprehensive approach involving detailed history, laboratory, imaging studies, and exclusion of more common pathologies. This case underscores the importance of considering SMAS as a differential diagnosis in persistent unexplained gastrointestinal symptoms.

<u>Abstract #67</u> Acute Systemic Lupus Erythematosus: An Exuberant Case in a Spectral Disease

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Introduction

The prevalence of systemic lupus erythematosus (SLE) in the United States is 20 to 150 cases per 100,000, with a complex association of genetic and environmental factors categorizing it as a spectrum from skin lesions to systemic manifestations. The recognition of subtypes of SLE-specific skin lesions can reflect the activity and severity of the disease, being an essential diagnostic tool.

Case Description

A 42-year-old woman with a history of hypertension presented to the emergency department with recurrent oral and vulvovaginal candidiasis, oral ulcers, skin rash, and right-sided chest pain. She reported a month-long illness and recent hospitalization for pneumonia. During this last admission, she was treated with Levofloxacin and developed skin and oral mucosal lesions, being diagnosed with Stevens-Johnson Syndrome. Discharged on prednisone, her symptoms improved, but upon cessation of medication, skin lesions flared up, and oral and vulvovaginal candidiasis started. The patient was admitted to our service due to reduced oral intake resulting from painful swallowing and concerns for candida esophagitis. Physical examination revealed a malar rash, sharply bordered erythematous patches on both eyebrows, multiple small circular plaques and macules on the chest, and oral ulcers. Laboratory workup showed anemia, lymphopenia, low C3 and C4 complement levels, positive ANA, and anti-dsDNA antibodies. Chest x-ray showed bilateral pleural effusion. TTE showed pericardial effusion. Renal function tests showed increased serum creatinine and nephrotic proteinuria. Renal biopsy revealed lupus nephritis. The patient was diagnosed with SLE based on the 2019 European League Against Rheumatism/American College of Rheumatology (EULAR/ACR) criteria, leading to the initiation of treatment with hydroxychloroquine 200mg and IV Diflucan 200mg daily.

Discussion/Conclusion

The admitting team initially suspected SLE due to the symmetry and exuberance of the lesions in photo-exposed areas. This case emphasizes the importance of performing a thorough history and physical exam. It illustrates how primary care physicians are pivotal in identifying and screening severe and often misdiagnosed medical conditions before major complications are encountered and providing adequate treatment.

Internal Medicine

<u>Abstract #8</u> Steering In The Wrong Direction: Magnesium Stearate, Offending Agent For Alpha-Gal Allergy

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Introduction

Alpha-gal is an IgE-mediated allergy to galactose-alpha-1,3-galactose, a carbohydrate expressed on non-primate mammalian proteins. This allergy can induce allergic responses ranging from hives, angioedema, or anaphylaxis. Meat by-products can be found in many medication formulations including the commercially available levothyroxine. Inactive ingredients such as magnesium stearate can be procured from mammalian sources and have detectable amounts of alpha-gal. Our objective was to report a patient in whom there were concerns regarding formulation of levothyroxine due to a history of anaphylactic reaction related to alpha-gal allergy, and then to describe the therapeutic approach taken.

Case Description

A 65-year-old-female with hypothyroidism and alpha-gal allergy presented with lip swelling and angioedema. We discovered that the inactive ingredient magnesium stearate is derived from a non-primate source and is commonly found in the formulation of levothyroxine. The diagnostic studies included IgE immunoassays for alpha-gal and thyroid function testing to personalize thyroid replacement therapy in a patient with hypothyroidism and alpha-gal allergy. After consulting Allergists and Immunologists, drug companies, and compounding pharmacies, she was prescribed a plant-based compounded preparation of levothyroxine with good clinical results.

Discussion/Conclusion

This case emphasizes the significance of identifying the variable risk factors, etiologies, and common medications that may be associated with alpha-gal allergy which in turn affect other endocrinopathies. A multidisciplinary collaboration is necessary to best tailor medications in patients with alpha-gal allergies and comorbid conditions. Gaining a better interpretation of the prevalence, diagnosis, and the therapeutic approach of alpha-gal allergy would assist in refining and standardizing medication formulation and prescription selections.

<u>Abstract #9</u> Acquired Idiopathic Generalized Anhidrosis In A Patient With Cholinergic Urticaria

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Introduction

Acquired idiopathic generalized anhidrosis (AIGA) is a rare disorder characterized by the insidious onset of the inability to sweat involving >25% of body surface area with the exclusion of other neurological and sweat gland dysfunctions. Symptoms may include cholinergic urticaria, facial or full-body sweating, cutaneous pain, pruritus, hyperthermia, nausea, vomiting, and more. We describe a case of AIGA affecting a Caucasian man with signs of anhidrosis and hyperthermia. Although there is no sufficient evidence for the effectiveness of systemic steroids, they are the recommended treatment for early stages of AIGA. However, our patient's symptoms were well-controlled on high-dose antihistamines. It is important for clinicians to recognize this rare entity and to initiate timely intervention to prevent serious consequences of hyperpyrexia.

Case Description

An otherwise healthy, active 20-year-old white man was referred for sudden onset of inability to sweat which he first noticed with strenuous physical activity. He had not noticed a lack of sweating performing his normal daily activities in the weeks prior. He experienced hives over his back, chest, and face with exertion and overheating. Diagnostic studies included total IgE levels and extensive urticaria, autoimmune, and inflammatory workup which in turn were largely unremarkable. He was trialed on Cetirizine 20mg twice daily with significant improvement in his symptoms.

Discussion/Conclusion

The importance of accurately recognizing and promptly treating AIGA cannot be overstated as hyperpyrexia can lead to seizures, coma, and even death. Prompt intervention and patient education prevented serious consequences from hyperthermia in our patient.

<u>Abstract #10</u> Mimicking Ischemic Strokes: Familial Cerebral Cavernous Malformations

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Introduction

Cerebral cavernous malformations or "cavernomas" consist of bundles of low-flow, thin-walled capillaries in the brain predisposing patients to intracerebral hemorrhage causing focal neurological deficits and seizures. The majority of these lesions occur sporadically and remain asymptomatic. Familial Cerebral Cavernous Malformations (FCCM) is a genetic cause making up approximately 20% of cases. Patients with FCCM develop numerous cavernomas and carry much greater risk of developing symptoms. We introduce a case involving a patient with probable FCCM, who exhibited acute, localized neurological impairments that closely resembled the symptoms of an ischemic stroke.

Case Description

A 77 year old female with a history of right hemispheric intracranial hemorrhage, pontine cavernous malformation treated with stereotactic radiotherapy, and rheumatoid arthritis presented to the emergency department with a 2 day history of fever, dysarthria, right facial droop, and acute worsening of chronic left-sided weakness preventing her from ambulating independently. Her daughter also has cavernomas suggesting the strong probability of a genetic component. The only change in this patient's medical care preceding this presentation was the initiation of Humira (adalimumab) therapy for rheumatoid arthritis. CTA of the head and neck was negative while CT of the head revealed a 1.5cm left temporal hyperdensity concerning for a subacute hemorrhagic cavernoma. An MRI revealed diffuse, innumerable cavernous malformations involving the brainstem, cerebellum, subcortex, and cortex, as well as evidence of a subacute left temporal hematoma secondary to a cavernoma. Paradoxically, the patients presenting focal neurologic disabilities did not correlate with imaging findings. The patient was managed with supportive care consisting of physical therapy and blood pressure optimization. Her strength subsequently improved to her baseline. It was recommended that she consider alternative immunotherapy as a similar event happened years ago causing fatigue, fever, and weakness when she was previously given TNF alpha inhibitor.

Discussion/Conclusion

With a reported prevalence of 1/5000, familial cerebral cavernous malformations represent a rare cause of focal neurological deficits, seizures, or intracranial hemorrhage. Current treatment options are limited due to the highly variable nature of the disease with surgical removal being indicated in select cases. Further research is necessary in order to better understand the pathophysiology of cavernoma formation and to decrease the risk of neurologic injury. There has not previously been a reported case of exacerbation of hemorrhagic cavernomas as an adverse reaction to TNF-a immunotherapy. This case represents an example of the unique presentation of cavernomas and the overlap in their presentation with ischemic stroke.

Abstract #26 An Oral Presentation of an Esophageal Disease

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Introduction

Eosinophilic Esophagitis (EoE) is an increasingly recognized disease distinguished by eosinophilic inflammation causing non-specific symptoms such as epigastric pain, esophageal dysphagia, non-cardiac chest pain, and food impaction.¹ The pathophysiology of the disease is believed to be secondary to environmental factors disrupting the epithelial layer of the esophagus through the production of cytokines by T regulatory and T helper cells, causing eosinophilic inflammation.2 EoE is diagnosed by the presence of ≥15 eosinophils per high-power field of esophageal biopsy tissue,3 and current therapies for EoE include proton pump inhibitors, topical steroids, and dietary modification.⁴

Case Description

A 62-year-old male presented to the clinic for evaluation of recurrent oral ulcers of uncertain etiology. Patient reported lesions had been present for 8 months. Most noticeable symptoms included difficulty swallowing and odynophagia. Differential diagnosis at that time was broad and included Sjogren's disease, Behcet's disease, Crohn's disease. He had been treated with colchicine, oral antifungal medications, lidocaine mouthwash and oral prednisone without resolution of ulcers. He noted around 30-pound weight loss since ulcers first presented. The patient was referred to a gastroenterologist and underwent an EGD with biopsy consistent with Eosinophilic Esophagitis. The patient was started on a proton pump inhibitor and an inhaled corticosteroid for treatment of EoE. An ENT referral was also considered for a biopsy of the patient's oral lesions. However, the patient's oral lesions rapidly resolved with treatment of EoE before an oral biopsy could be obtained. It was hypothesized that the oral lesions were eosinophilic in nature given the prompt resolution in treatment of the EoE.

Discussion/Conclusion

Traditionally, oral eosinophilic ulcers will present as a single, non-recurring ulcer that resolve following a biopsy. Recently, a small number of case reports document eosinophilic ulcers that are chronic, recur, and are not affected by biopsy. 5 This case highlights a similar chronic oral ulcer and illustrates a potential uncommon presentation of EoE. Clinicians should consider EoE on the differential for patients with recurrent ulcers. EoE is a disease that often goes on for years before eventual diagnosis and treatment with a median diagnostic delay time of 6 years. 6 Earlier recognition and diagnosis is important as progression of the disease can lead to strictures and recurrent ulcers. Diagnosis is achieved with biopsy, and current treatment guidelines recommend proton pump inhibitors, topical steroids, dietary modification, and most recently dupilumab, a monoclonal antibody which was FDA approved in 2022.

<u>Abstract #27</u> Immune Reconstitution Inflammatory Syndrome (IRIS) Unmasking A Bartonella Henselae Neuroretinitis

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Introduction

Cat scratch disease (CSD) is a self-limited, systemic zoonotic infectious process that primarily manifests as a regional lymphadenitis, caused by Bartonella henselae. However, humans can develop extra-nodal dissemination with life-/organ-threatening complications, especially in immunocompromised hosts. Bartonella species are a family of small, fastidious, pleomorphic gramnegative intracellular bacilli that are typically acquired through inoculation from penetrating trauma from the claws or teeth of felines; but arthropod-related transmission of non-henselae Bartonella species has also been described. The most common ocular manifestations are neuro-retinitis and focal retinochoroiditis with retinal infiltrates.

Case Description

A 38-year-old male with treatment-experienced chronic HIV-1 infection (diagnosed ~ 13 years prior to presentation), has a history non-adherence to various ART regimens, with resultant AIDS. He was previously diagnosed with neuro-/ocular syphilis and received 14 days of parenteral penicillin G (diagnosed ~ 8 years prior to presentation). He initiated TAF/FTC/BIC 3-4 months prior to presentation. He presented to a local tertiary care center reporting 1 week of abrupt onset rightsided diplopia and discomfort with extraocular movements. Ophthalmology consulted urgently and diagnosed an acute anterior uveitis with features of vitriitis, and concerns for posterior involvement. A presumptive diagnosis of ocular syphilis was made given his the reactive treponemal serology, with an RPR titer of 1:256. MRI orbits showed "irregularity, abnormal thickening and abnormal enhancement involving the right posterior retina...altered signal involving the right globe vitreous humor." Bartonella serology was performed, that revealed an IgG titer of 1:2048, with an IgM of 1:10. The molecular testing of plasma for Bartonella species was below the level of detection. Given the multi-compartmental involvement of the orbit (including retina), and the strong serologic response despite the lymphopenia (CD4: 136 cells/μL), patient was initiated on antimicrobial therapy for Bartonella neuro-retinitis with rifampin and doxycycline. He was also treated with corticosteroids for suspected immune reconstitution inflammatory syndrome (IRIS), and as an adjunctive therapy for Bartonella neuro-retinitis.

Discussion/Conclusion

IRIS unmasking an infectious process after initiation of ART requires purposeful monitoring and early detection. Management of IRIS may require immunomodulation in the setting of a life-/organ-threatening disease process. The use of antimicrobials that achieve high intracellular concentrations are needed, but high-quality studies are lacking. The diagnosis of Bartonella-related ocular disease requires a high index of suspicion and access to serologic testing. Bartonella chemoprophylaxis is not routinely performed, but macrolides have demonstrated protective efficacy.

Abstract #29 A Rare Case of Ecthyma Gangrenosum

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Introduction

Ecthyma gangrenosum (EG) is a skin condition that has been extensively linked with Pseudomonas bacteremia in immunocompromised individuals. It typically presents in intertriginous areas as a black/gray lesion with a central necrotic base surrounded by erythematous border. When EG is suspected, empiric antibiotic therapy against Pseudomonal organisms should be promptly started while awaiting blood cultures. Aggressive EG lesions require surgical debridement. Histology and culture of EG can show gram negative bacteria, inflammatory cell infiltration and vascular proliferation. There are very few cases described in literature linking E coli bacteremia to EG. We present a rare case of that to emphasize the importance of considering non-Pseudomonal organisms when EG lesions are observed.

Case Description

A 62-year-old female with a history of glioblastoma presented to the emergency department with a two-day history of fever, chills, nausea, vomiting, and generalized weakness. Five days prior, she had completed a six-week therapy of temozolomide and radiation treatment. She was found to have neutropenic fever and thrombocytopenia. Upon admission, she also reported painful lesions in both underarms. On exam, she had two distinct lesions – bilateral chronic appearing nodular lesions as well as multiple tender black and gray ulcerative lesions with necrotic cores, which, at first, were thought to be hidradenitis suppurativa, but input from infectious disease revealed the lesions to be more consistent with EG. She was initially started on Zosyn for Pseudomonal coverage. Blood cultures revealed E coli with susceptibility to ceftriaxone and antibiotics were narrowed. She later developed right thigh cellulitis that progressed to a necrotic lesion resembling EG. Antibiotics were broadened to Vancomycin and Zosyn and the patient underwent surgical debridement. Although the right thigh lesion wasn't sent for pathology, it was highly suspicious for EG. Subsequently, she developed a necrotic oral lesion. Pathology of the oral lesion was negative for acid-fast bacilli and fungal organisms. The lesion was not tested for gram negative bacteria.

Discussion/Conclusion

This case illustrates that it is important to avoid anchoring to Pseudomonas bacteremia when EG lesions are observed. Although rare, cases of EG and E. Coli and even fungal bacteremia have been discovered. These lesions often require surgical debridement to get source control of the underlying infection, therefore identifying EG correctly can lead to improved patient care. Additionally, this case highlights the importance of thorough dermatological examinations to accurately diagnose and treat skin conditions, and to better equip medical professionals with a strong dermatology foundation.

Abstract #30 TMVR for High-Risk Valve Revision with Percutaneous PVL Closure

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Introduction

Transcatheter Mitral Valve Replacement (TMVR) is a new technology that allows for full repair of damaged mitral valves. TMVR can be used for complete replacement of the mitral valve and is currently reserved for patients that are not good candidates for open surgical procedures. TMVR can be done on a valve that has previously been surgically replaced. Below is a case that walks through a TMVR being used in the setting of a previously replaced valve and the additional challenges.

Case Description

A 71-year-old presented with a history of mitral valve endocarditis repaired in 2008, with bioprosthetic redo repair in 2014. The original mitral valve was replaced due to valve destruction from endocarditis. At the time of presentation, she is following up with the outpatient cardiology clinic for her worsening shortness of breath.

After the second valve revision she was found to have a mild paravalvular leak (PVL) on transesophageal echocardiogram (TEE) in 2016. Subsequently she was found to have a moderate to severe paravalvular leak with thickened leaflets with moderate stenosis in 2021. Repeat in 2022 showed regurgitation with progression to severe stenosis. She was considered to be a high-risk candidate for redo-redo sternotomy for any further repair. After discussion with the multidisciplinary heart team, it was decided that percutaneous PVL closure followed by TMVR was her best option.

She was taken to the Cath lab where a transseptal puncture was accomplished using TEE guidance to gain access to the left ventricle. A ventricular septal defect (VSD) occluder was successfully used for closure of her paravalvular leak followed two months later where she was taken for revision of the bioprosthetic mitral valve via TMVR. TEE guidance was then used for atrial septostomy, and the valve was placed in position under fluoroscopic guidance. Bidirectional shunting was noted following valve deployment, and the atrial septum was closed using an atrial septal defect (ASD) occluder. Her symptoms improved with the repair of her severe bioprosthetic mitral valve stenosis and PVL.

Discussion/Conclusion

TMVR is becoming a more common procedure in the field of cardiology as additional techniques and advancement in technology continue to improve. Having these kinds of procedures at a cardiologist's disposal can allow for more successful revisions of valvular disease and decrease the risk of severe outcomes that come with open procedures. This case exhibits how some of the technological advances help provide better outcomes for high-risk patients.

Abstract #31 Duodenal Perforation from Secondary Syphilis

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Introduction

Syphilis is an infection caused by the spirochete Treponema pallidum. Most cases of human infection are sexually transmitted, but congenital spread has been well described. The incidence of syphilis is on the rise in the United States, despite readily available diagnostics, and highly efficacious therapeutic options. Syphilis has been implicated as a pathogen in nearly every organ system. The following case describes a spontaneous duodenal perforation that occurred in a patient receiving therapy for neurosyphilis with suspected syphilitic myelitis.

Case Description

A 37-year-old male with a history of inhalational methamphetamine use, presented to a rural hospital with one week of progressive lower back pain, urinary incontinence, and lower extremity weakness. MR imaging showed diffuse cord signal of the cervicothoracic territories compatible with myelitis. Lumbar puncture revealed a culture-negative lymphocytic pleocytosis with elevated protein consistent with aseptic meningitis. Based on a reactive treponemal EIA screen, a nontreponemal RPR titer of 1:256, and a compatible clinical syndrome, treatment was initiated for neurosyphilis with parenteral penicillin G. VDRL from the CSF was later found to be positive with a titer of 1:8. The hospital course was further complicated by a spontaneous duodenal perforation, resulting in a septic peritonitis with multi-organ dysfunction syndrome requiring emergency laparotomy.

Upon transfer to tertiary care facility, antimicrobials were transitioned to ceftriaxone and metronidazole. Repeat laparotomy with revision of the duodenal perforation was performed. Immunofluorescent staining of the biopsied perforation site revealed the presence of spirochetes. Postoperative course was complicated by feculent output from surgical drains requiring repeat laparotomy. Exam revealed a gangrenous and necrotic right colon necessitating hemicolectomy. Repeat immunohistochemical stains demonstrated the presence of spirochetes within the colon, suggesting the bowel perforations may have been the result of overwhelming treponemal infiltration. Unfortunately, the patient continued to suffer nosocomial complications including a ventilator-associated pneumonia with a resultant pseudomonal bloodstream infection. After weeks of aggressive measures, the patient succumbed to the multitude of infectious and metabolic insults on hospital day 25.

Discussion/Conclusion

This complex case demonstrates this organism's capacity for dissemination, and the tropism for various tissues. Its moniker as "The Great Imitator" has been well-deserved after the initial descriptions of this pathogen were described more than 500 years ago. Although rarely described enteric involvement by these treponemes may be underrecognized as a clinical entity. Given the rising prevalence of this disease, enteric manifestations should be considered in acute and non-acute settings in those with an active syphilis diagnosis.

Abstract #33 Secondary Hypercalcemia from Pulmonary Histoplasmosis

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Introduction

Pulmonary histoplasmosis is a fungal lung infection caused by the organism Histoplasma capsulatum that can be acquired through the inhalation of fungal spores in endemic areas. The disease can range from mild and even asymptomatic to more severe and debilitating particularly in an immunocompromised host.

Case Description

Here we present a 54-year-old male with a history of multiple sclerosis on immunosuppressive therapy who was hospitalized initially for acute encephalopathy and sepsis related to a complicated UTI, renal failure with hypercalcemia (Ca corrected: 14.9), and multifocal bacterial pneumonia that later progressed to septic shock resulting in a protracted ICU stay. His hypercalcemia was initially attributed to acute renal failure and responded appropriately to fluids with recovery of renal function; however, he later developed hypercalcemia again on hospital day 18. This was attributed to hypercalcemia of immobility and was treated again with fluids, but the hypercalcemia again returned on day 26. An extensive work up for secondary causes of hypercalcemia demonstrated an elevated 1,25-OH vitamin D level, elevated 1,3-beta-D-glucan (Fungitell test for invasive fungal infections), and eventually fungal serologies that were suggestive of pulmonary histoplasmosis. The patient was treated for his bacterial and fungal infections and responded somewhat to treatment with the help of multiple consultants including nephrology, infectious disease, and critical care, among others. Given the severe nature of his presentation in collaboration with the patient and family, palliative care was involved and after a prolonged hospital stay exceeding 70 days the decision was made to pursue hospice and the patient was discharged to home where he later expired.

Discussion/Conclusion

This case highlights the importance of early initiation of secondary hypercalcemia workup in immunocompromised patients particularly when other risk factors for opportunistic infections exist.

Abstract #41 Hepatic Involvement in Systemic Mastocytosis

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Introduction

Systemic mastocytosis is an aggressive disorder characterized by the hyperproliferation of excessively active mast cells, which can manifest as anaphylaxis, skin flushing, neuropsychiatric complaints, and nonspecific GI complaints. The pathophysiology of this disease is thought to be due to mutations of KIT, the gene responsible for encoding CD117 transmembrane tyrosine kinase which plays a role in the growth, survival, and migration of mast cells. Diagnosis of systemic mastocytosis is accomplished with bone marrow biopsy, which can reveal multifocal dense infiltrates of mast cells (> 15 mast cells in an aggregate), mast cells with atypical morphology, or detection of KIT point mutations in the bone marrow. In rarer cases, malignant ascites can be one of the presenting features of the disease.

Case Description

A 68-year-old male was referred to oncology for unintentional weight loss, fatigue, leukocytosis, elevated alkaline phosphatase, retroperitoneal lymphadenopathy, and newly found hepatosplenomegaly. The initial differential included infection, autoimmunity, metastatic disease, and primary bone marrow related pathology. A PET CT scan revealed osteoblastic metastasis in the axial and appendicular skeleton. This, in addition to labs positive for IgM kappa monoclonal protein raised concern for a clonal hematopoietic process, so a bone marrow biopsy was taken which revealed atypical mast cell aggregates which were CD 117+ and KIT D816V positive, consistent with systemic mastocytosis. The patient then developed recurrent ascites, and underwent multiple sessions of paracentesis, with initial fluid studies revealing malignant cells. Gastroenterology was consulted for evaluation of the recurrent ascites, and liver elastography revealed a shear wave elastography average of 18.8 kPa, which fell under the F4 category and was indicative of cirrhosis. The patient was placed on diuretics for the recurrent ascites, and the oncology team initiated treatment of the mastocytosis with midostaurin, a tyrosine kinase inhibitor. Following initiation of this treatment, the patient has shown improvement in labs and has not required another paracentesis, and liver biopsy has been deferred.

Discussion/Conclusion

This case highlights the need for clinicians to be familiar with mastocytosis and its potential for uncommon systemic manifestations such as malignant ascites. Earlier diagnosis is important as progression of mastocytosis can lead to neuropsychiatric complaints, anaphylaxis, decreased appetite, significant weight reduction, and widespread metastasis. Diagnosis is achieved through bone marrow biopsy, which can reveal mast cell abnormalities and/or KIT mutations. Treatment is highly individualized, and can include small molecule kinase inhibitors that target the KIT receptor, stem cell transplantation, and symptom-directed therapy.

<u>Abstract #43</u> From Hamburgers to Hospitalization: A Significant Presentation of Alpha Gal Syndrome

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Introduction

Galactose- α -1,3-galactose is a carbohydrate molecule found in non-primate mammals and their animal products including pork, beef, lamb, dairy, and gelatin. Alpha-gal syndrome (AGS) refers to an IgE antibody-directed hypersensitivity reaction following an exposure to certain tick species, including the Lone Star Tick (Amblyomma americanum). Although not fully understood, some evidence shows that the saliva of the Lone Star Tick has epitopes that react with alpha-gal antibodies leading to increased levels of alpha-gal specific IgE when exposed to certain meat causing clinical symptoms ranging from pruritis to anaphylaxis. AGS is most common in the southeastern United States and is increasing in prevalence. From 2010 to 2018, over 34,000 individuals tested positive for alpha-gal IgE.

Case Description

A 52-year-old female with a past medical history including hereditary hemochromatosis and hypothyroidism presented to the ED after becoming hypotensive during an in-patient sleep study. She also experienced hives on her upper body, pruritus, throat tightening, and a decline in mental status. Patient was found to have systolic blood pressure readings in the 40's-50's, which improved with fluids and vasopressor support. Labs of note on admission include an IgE total of 69.0, C1 Esterase Inhibitor 25.0, and CRP 20.3. Over the previous 4 months, she had several similar, but milder episodes which resolved with over-the-counter antihistamines. The patient denied any new exposure to medicines or food.

Additionally, the patient reported frequently visiting her father who lives in a rural, wooded area. She denied any known tick bites. Her brother, who also visited their father frequently, had been diagnosed with AGS. The patient recalled consuming a hamburger a few hours prior to her sleep study. An alpha-gal IgE titer was measured and found to be 28.90 IU/mL (positive >0.1IU/mL) suggesting alpha-gal as the source of her anaphylaxis. Patient was encouraged to follow up with an allergist.

Discussion/Conclusion

The diagnosis of AGS can be difficult given the non-specific and often 2-6 hour delay of symptoms after consumption of the offending agent. Because the patient was being monitored during a sleep study, she was able to get immediate medical care. With the growing incidence of AGS being seen in the population, it is important for clinicians to consider AGS when determining the source of an allergic reaction. Follow-up with an allergist is essential as dietary restriction and antihistamine regimens remain the current treatment. Future studies to consider include finding potential ways to reverse AGS.

Abstract #46 A Backup of Bile; Knowing When to Escalate Care

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Introduction

Progressive familial intrahepatic cholestasis (PFIC) is a rare autosomal recessive disorder with defects in intrahepatic transportation of bile, resulting in cholestasis (1). Symptoms usually present in infancy and include jaundice, pruritus and hepatomegaly. Long term complications include portal hypertension, cirrhosis, and increased risk of hepatocellular carcinoma (2). Reported incidence varies from 1/50,000 -100,000 live births with no indication of increased incidence related to gender or specific populations. This disease often presents in childhood, but one subtype can have a delayed presentation (3). PFIC types 1 and 2 present earlier in childhood, while type 3 presents later in infancy or into adulthood. (3). Diverting biliary techniques are often utilized to delay or avoid liver transplantation, but one study mentioned transplantation rates up to 87% in patients with PFIC 2 (2).

Case Description

A 37 year old male with a history of PFIC presented with one week of severe pruritus, hyperbilirubinemia, abdominal distention, and decreased biliary drainage. He reported similar episodes since infancy. He was evaluated for transplant at age 18 but was eventually removed from the transplant list at age 25 due to clinical improvement. Shortly after that, he underwent partial external biliary diversion at the University of Nebraska, a regional leader in gastroenterology and GI surgery. The procedure involved everting his gallbladder, bringing it to the abdominal wall, and creating a stoma that drains his gallbladder contents into an ostomy bag.

At time of presentation to the hospital, he reported around a 50% reduction in biliary ostomy output and a 10 pound weight loss. Notable labs included alkaline phosphatase 195, total bilirubin 30.6, direct bilirubin 22.3. CT abdomen was significant for partial small bowel obstruction and a percutaneous cholecystostomy fistula. General Surgery was consulted and recommended immediate transfer to the University of Nebraska for evaluation, which occurred within 8 hours.

Discussion/Conclusion

PFIC is one of several rare conditions that Health Care professionals may encounter. Providers often lack previous experience and standardized guidelines on management can be difficult to find. Rare conditions, like PFIC, may require care at quaternary care centers. Clinicians need to be able to gauge the severity and acuity of the illness at time of presentation. This often can be done by a basic laboratory workup and interdepartmental collaboration, as demonstrated by this case. This allowed the patient to be transferred to the appropriate facility, thus avoiding a delay in care.

<u>Abstract #48</u> A Man with Debilitating Dystonia: Case Report of Stiff Person Syndrome

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Introduction

Stiff Person Syndrome (SPS) is a neuroimmunological syndrome characterized by painful muscle spasms and rigidity due to autoantibodies. SPS is an extremely rare condition, with an estimated prevalence of 1 out of every 1.25 million people. The syndrome can present in varying regions of the body and severity levels. In this study, we present the case of a man with progressively worsening dystonia who was diagnosed with SPS.

Case Description

A 45-year-old male presented to the neurology clinic for follow-up of progressive dystonia symptoms he was experiencing for more than 17 years. His symptoms began with spastic episodes in his distal upper extremities. As the years progressed, he began to experience increased severity, pain, frequency, and body involvement of the spastic episodes, to the point of constant diffuse myalgias and muscle spasms. His spasms are present to a greater extent in limbs compared to torso, particularly in the legs. Patient also reports involvement of the face, eyes, throat, and respiratory muscles. On physical exam, the patient presents with slight hyperreflexia (3/4 in all locations) and decreased pinprick, vibratory, and proprioceptive sensation in all extremities. Strength is 5/5 in all muscle groups. Intermittent spasms occur throughout the encounter. Previous imaging and muscle biopsy over the years were all unremarkable. GAD65 antibody testing was negative. He has an unknown family history and had no neurological history prior to the beginning of his spasms. Diagnosis of SPS was made after exclusion of other potential diagnoses. Primary medication for symptomatic control is dantrolene. The patient reports his 24 year old son has also begun to experience the same painful muscle spasms, indicating a condition with a likely autosomal dominant inheritance pattern.

Discussion/Conclusion

Diagnosis of SPS is primarily made with testing for the GAD65 autoantibody. However, only 70-85% of patients with the condition are reported to be positive for this specific antibody, which could explain why this patient was negative for it. The classic presentation of SPS involves the whole body and is the most common type (70%). Clinical manifestations are commonly seen in the torso first and then the limbs. Symptoms include hyperlordosis, rigidity, muscle spasms, pain, gait spasticity, and hyperreflexia. Genetic predisposition to the condition has been identified in several cases. Currently, no definitive treatments exist for SPS, and the disease ultimately results in severe morbidity or even mortality. Current treatments focus on three areas: symptom relief, immunomodulation, and physical therapy/rehabilitation.

Abstract #49 Pantoea Agglomerans, Not Always a Contaminant

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Introduction

Pantoea agglomerans is a nonencapsulated, non-spore-forming anaerobic Gram-negative bacillus belonging to the Enterobacteriaceae family. It is a rare cause of opportunistic infections caused by wound infection with plant material or as a hospital-acquired infection mostly in immunocompromised individuals, affecting premature infants to seniors.

Case Description

A 63-year-old with past medical history of antiphospholipid syndrome, SMA thrombus s/p resection with resultant short gut syndrome on chronic TPN, GERD, paroxysmal atrial fibrillation, and diastolic heart failure Presents with nausea/vomiting, headache, and fatigue. On admission, vitals were unremarkable and lactic acid and white blood cell counts were normal. Chest x-ray and urinalysis were without acute process. On physical exam, his "right chest portacath was without erythema or swelling". Abdominal XR and CT showed a partial small bowel obstruction at the anastomosis site. Patient developed a fever up to 39.5°C and continued to be febrile despite broad-coverage antibiotics (vancomycin, piperacillin-tazobactam). Initial blood cultures showed no growth, but culture was repeated given a continued fevering without an obvious source. Two-out-of-two blood cultures resulted with Pantoea agglomerans and were marked as presumed contaminant by lab. Infectious disease was consulted and theorized a GI source of infection with possible enteric translocation of bacteria resulting in recurrent infections. The lab was called to request susceptibilities. Ultrasound revealed mild subcutaneous edema adjacent to the catheter site and the catheter tip was then exchanged and cultured, which grew P. agglomerans. P. agglomerans bacteremia was diagnosed. Patient was treated with prolonged oral doxycycline 100 mg twice daily (30 -days with option to extend therapy). The patient was successfully treated and recovered fully without any complication.

Discussion/Conclusion

Possible routes of transmission in this patient include contamination of his intravascular catheter, contamination of his parental nutrition, or gastrointestinal translocation in the presence of gastroesophageal mucosal lesions (given this patient's history of an anastomosis site and gastroesophageal reflux disease). Outcomes seem favorable with the institution of appropriate antibiotics even in immunocompromised patients. In a cohort study of spontaneous bacteremia, 100% of isolates were susceptible to cefotaxime, ceftazidime, piperacillintazobactam, imipenem, ciprofloxacin, gentamicin, and amikacin. *Pantoea* agglomerans is a pathogen of low virulence even in an immunocompromised adult host, causing diverse clinical picture, and can be treated successfully with proper antibiotic use.

<u>Abstract #50</u> More Swollen than Swole: Anabolic Steroid-Associated Multi-Organ Failure

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Introduction

Anabolic steroid use among weightlifters and bodybuilders has long been associated with a wide range of potential health complications largely as a consequence of endocrine dysregulation. We present a case of multi-organ dysfunction as a result of such steroid use.

Case Description

A 44-year-old man presented to the emergency department for shortness of breath and intermittent chest pain of 6-8 weeks duration. He endorsed orthopnea and lower extremity edema which coincided with the onset of his symptoms. Notably, on admission he admitted to a history of anabolic steroid use for the past 3 months and had been using a combination of testosterone, methandienone, and medroxyprogesterone. Physical exam revealed severe hypertension, sinus tachycardia, and 2+ pitting edema of bilateral lower extremities. Lab results were significant for mild elevations of troponin and BNP, hypoalbuminemia, and a normal GFR. Echocardiogram revealed severe left ventricular hypertrophy with global hypokinesis and a left ventricular ejection fraction of 45%. Further workup for his hypoalbuminemia included measurement of a urine protein/creatinine ratio which was significant for nephrotic range proteinuria. He ultimately received a renal biopsy which demonstrated focal segmental glomerulonephritis with crescent formation. The patient's hypertension and edema improved with antihypertensive therapy and guideline-directed medical therapy for new-onset heart failure and he was counseled on cessation of anabolic steroid use.

Discussion/Conclusion

Several cases of dilated cardiomyopathy as a consequence of anabolic steroid use have been reported. Cardiac myocytes express androgen receptors and dysregulation secondary to exogenous androgens is thought to cause adverse remodeling.1 The association between anabolic steroid abuse and FSGS is less well understood; pathogenesis is thought to be multifactorial with androgen dysregulation, direct toxicity, and oxidative stress playing potential roles.2

This case highlights the potential for rapid onset multi-organ failure as a consequence of anabolic steroid abuse and the importance of thorough history in the workup of new onset heart failure. Furthermore, this patient's concomitant diagnoses of heart failure and nephrotic syndrome demonstrate the need to consider multiple underlying etiologies in the presentation of volume overload.

<u>Abstract #54</u> Diffuse Dermal Angiomatosis of the Breast in the Setting of Pregnancy

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Introduction

Diffuse Dermal Angiomatosis, or DDA, is a rare angioproliferative disorder that predominantly affects limbs but can occur on the breasts. Typical manifestation is violaceous patches and plaques on the breasts, often with ulceration. The etiology of DDA of the breast is currently hypothesized as a reaction to tissue ischemia, possibly driven by VEGF, vascular endothelial growth factor. Currently associated risk factors include smoking, macromastia, obesity, and systemic blood vessel disease. Reported treatments have included isotretinoin, breast reduction surgery, and lifestyle changes. To the authors' current knowledge, only one case of DDA of the breast occurring in pregnancy has been reported.

Case Description

A 31-year-old woman at 9 weeks gestation presented with non-itchy, burning bilateral erythematous plaques on the breasts of approximately 2 weeks duration. No ulceration was present. She had previously been diagnosed with mastitis but did not respond to oral antibiotic treatment and felt the rash was continuing to spread. Due to typical treatments being contraindicated in pregnancy, the patient was started on clindamycin 1% lotion and petroleum jelly. A first trimester biopsy was non-specific and found ichthyosiform dermatitis suggestive of atopic dermatitis. Continuing treatment, the patient noted improvement over the next 3 months, but given the thickened nature of her plaques still had concerns about inflammatory breast cancer so repeat biopsies were taken in her second trimester, which suggested DDA of the breast. Given her improvement, the patient continued on clindamycin 1% lotion and petroleum jelly and has remained stable.

Discussion/Conclusion

DDA of the breast is an increasingly recognized condition, but there is still limited evidence regarding the underlying mechanism and optimal treatments, especially in the setting of pregnancy. Breast tissue changes in the first trimester of pregnancy are related to estrogen resulting in proliferation of the ductal tissues, and this increase in tissue mass may increase risk for development of DDA. Additionally, progestin responsive cells in the decidua have been shown to result in increased VEGF-A expression, and DDA in the setting of pregnancy may represent an aberration of this process. The response of our patient to emollients and topical clindamycin provides a safer alternative therapy when treating pregnant patients. Although a rare disease, our patient's journey highlights that DDA of the breast is an entity that can occur in pregnancy and may mimic mastitis or inflammatory breast cancer, and that multiple biopsies may be required to reach the right diagnosis.

Abstract #58 Atypical Hyperpigmentation with Overuse of Setmelanotide

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Introduction

Bardet-Biedl Syndrome (BBS) is a disorder due to a mutation in over 24 genes that play a role in primary cilia function. Evidence has shown that a dysfunctional BBS protein impairs trafficking of the leptin receptor (LEPR) in hypothalamic proopiomelanocortin (POMC) neurons which reduces the activation of melanocortin-4 receptor (MC4R). Due to the role of BBS genes on cilia function and LEPR trafficking, BBS is associated with early-onset obesity, retinopathy, intellectual disability, polydactyly, and renal abnormalities.1 Setmelanotide, an MC4R agonist, is a pharmacological treatment for BBS with a side effect of hyperpigmentation due to agonism of melanocyte receptors in the skin.2 Here, we report a case of severe hyperpigmentation in a patient with BBS from an accidental setmelanotide overdose.

Case Description

A 26-year-old female with past medical history of obesity, asthma, anxiety, obstructive sleep apnea, and gastroesophageal reflux disease presents with BBS (BBS9 and MKKS mutations). She has experienced weight issues, night vision difficulties, and learning disabilities. This met BBS criteria and she was started on setmelanotide (10mg/mL). She accidentally overdosed, taking 1mL injection daily rather than 1mg, resulting in sun-exposed hyperpigmentation. The patient showed no other side effects aside from mild nausea.

Discussion/Conclusion

Ciliary dysfunction in POMC neurons is thought to be the cause of obesity in BBS patients. POMC neurons release melanocyte-stimulating hormone (MSH) in response to LEPR activation. MSH binds to and activates the MC4R on MC4R-expressing neurons that stimulates a cascade of signaling leading to suppression of hunger. Thus, setmelanotide, a MC4R agonist allows for improved weight management and reduced hyperphagia.3

Skin hyperpigmentation is a commonly reported side effect of setmelanotide with a study reporting 78% of patients developing hyperpigmentation. This occurs because melanocortin receptors are expressed on melanocytes, and activation of this receptor leads to accumulation of melanin. Other common side effects of setmelanotide include injection site reaction, nausea, headache, and diarrhea.4

To our knowledge, this is the first reported case of severe hyperpigmentation with overuse of setmelanotide. This was complemented by the fact that the patient did not experience other side effects. Her BBS gene mutations of BBS9 and MKKS may play a role in this patient's unique response to 10x the FDA labeled dosing of setmelanotide. Further research is needed to understand if these mutations played a role in the development of hyperpigmentation without other common side effects.

Abstract #61 Anorgasmia Following Bariatric Surgery

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Introduction

Bariatric surgical procedures are an increasingly common form of management of class II and III obesity. These operations involve alterations in gastric and intestinal anatomy which result in changes in gastric emptying and intestinal transit times, decreased interaction with digestive substances, increased stomach pH, and decreased absorption surface area in the small intestine. The impact on nutritional absorption following bariatric surgery has been well-documented, but there is less information available on the effects of medication absorption.

Case Description

A 41-year-old man with class III obesity, type II diabetes, and depression presented for follow up in bariatric medicine clinic six weeks after undergoing vertical sleeve gastrectomy. He was successfully losing weight at an expected rate, achieving adequate fluid and nutritional intake, and taking bariatric vitamins as prescribed. He reported inability to achieve an orgasm despite no change in sexual desire or ability to achieve and maintain an erection. Further review revealed worsening symptoms of depression following surgery. For the past two years, his depression was stable on 20 mg escitalopram in combination with 150 mg extended-release bupropion for SSRI-induced sexual dysfunction. His worsening depression and anorgasmia was attributed to a combination of SSRI side effect and impaired absorption of extended-release bupropion. He was placed on an equivalent dose of bupropion sustained release twice daily with plans for close follow-up.

Discussion/Conclusion

A common side effect of serotonin-specific reuptake inhibitors is sexual dysfunction. Bupropion is an antidepressant that is used off-label to counteract these side effects. Bupropion is rapidly absorbed by the intestinal tract, so traditional immediate release forms are prescribed two to four times per day. Formulations were developed to include 12-hour sustained release and 24-hour extended-release tablets to decrease the number of daily administrations. Many medications have similar formulations that alter the release period of active substances, and others have narrow therapeutic windows. It is important to understand and monitor these changes in bariatric surgery patients.

The anatomic alterations of bariatric surgery lead to physiologic changes in factors that can effect absorption of medications. This case highlights the importance of close follow-up post-operatively for monitoring the effectiveness and side effects of medications.

<u>Abstract #72</u> Gaining Momentum with Appetite Reduction from Anti-Obesity Medications: Is it Time to Hit the Brakes?

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Introduction

Obesity is a widely prevalent condition that affects greater than 40 percent of the population with annual medical costs of approximately 173 billion dollars in the United States. Tirzepatide is a dual glucagon-like peptide-1 (GLP-1) and glucose-dependent insulinotropic peptide (GIP) receptor agonist. Setmelanotide is a melanocortin 4 receptor (MC4R) agonist used to treat patients with genetic deficiencies in the appetite inhibitors leptin or pro-opiomelanocortin (POMC). These medications have received FDA approval within the past three years. They promote significant weight loss by acting on the brain to alter food reward salience and enhance satiety. Avoidant restrictive food intake disorder (ARFID) involves reduced interest in or concerns about the adverse effects of eating, leading to weight loss, nutritional deficiencies, or significant functional impairment.

Case Description

Two patients who had previously undergone bariatric surgery were started on tirzepatide (patient A) or setmelanotide (patient B) for weight loss at an obesity medicine clinic. Within two weeks of starting tirzepatide, patient A developed acute food aversions and consumed less than 400 calories daily. She lost 27 lbs within five months. Within three months of starting setmelanotide, patient B developed acute food aversions and obsessions about weight loss requiring fluoxetine. She consumed less than 500 calories daily. She lost 46 lbs within two months. After these medications were discontinued, these patients had improvements in their appetites but did not regain their weights.

Discussion/Conclusion

GLP-1 receptor stimulation in the ventral tegmental area of the brain increases dopamine transporter function, which is associated with reduced synaptic dopamine levels. Low synaptic dopamine levels may reduce the hedonic value or motivational salience of food. Setmelanotide promotes the effects of leptin and POMC, which include reducing food cravings. Overall, tirzepatide and setmelanotide may possibly play a role in ARFID-like syndrome in these patients by reducing hedonic tone and food incentive salience, thereby leaving unchecked obsessive tendencies toward food. Findings from this case series can be used to further characterize the spectrum of adverse effects associated with GLP-1, GIP, and MC4 agonists and treatments that can be used to manage these effects.

Obstetrics and Gynecology

<u>Abstract #11</u> Placenta Percreta in a 53-year-old Preeclamptic Jehovah's Witness Patient

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Introduction

The increasing rates of cesarean deliveries nationwide has contributed to the rising incidence of placenta accreta spectrum (PAS). As reported by the Centers for Disease Control and Prevention (CDC), the prevalence of severe maternal morbidity(SMM) continues to climb despite decreasing levels of maternal mortality. Cesarean hysterectomy in combination with multiple comorbidities can result in major complications significantly affecting the short and long-term consequences of a woman's health.

Case Description

A 53-year-old Jehovah's Witness(JW) parturient was affected by PAS. More specifically, she was diagnosed with placenta percreta, where the placenta invaded through her uterus to involve her bladder. This life-threatening condition can only be carefully treated via cesarean hysterectomy by experienced obstetricians, maternal fetal medicine specialists, and gynecology oncology surgeons. In addition, she was plagued by other comorbid conditions threatening her health: preeclampsia, asthma, anemia, and advanced maternal age. The combination of these factors placed this patient at extremely high risk for SMM and maternal mortality and caring for her required a multidisciplinary approach with consultation of specialists in the field of obstetrics and anesthesiology. She underwent a general anesthetic by an obstetric anesthesiologist to facilitate the delivery of a healthy preterm neonate via cesarean delivery. Afterwards, the obstetrician worked closely with a gynecologic oncologist to safely perform a hysterectomy. Cell saver was utilized and the patient had an estimated blood loss of 1000 mL.

Discussion/Conclusion

The management of women affected by PAS can vary depending on comorbidities and severity of the disease process. In this case report, we discuss the epidemiology of PAS, rates of SMM, and anesthetic management of patients undergoing cesarean hysterectomy. Patients with multiple risk factors for adverse events require careful planning and coordination of care by a team of healthcare providers. We illustrate a successful anesthetic and delivery of this high-risk patient and discuss considerations affecting the care of a JW patient undergoing a precarious surgery.

<u>Abstract #18</u> When Two Sacs Become One... A Case of the Rupture of the Dividing Membrane Between Twins

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Mr. Guimy Castor – OU-TU School of Community Medicine, Department of Obstetrics & Gynecology

Introduction

Twin gestations carry a higher risk of perinatal morbidity and mortality when compared to their singleton counterparts. Monochorionic twin pregnancies are associated with even higher morbidity and mortality compared with dichorionic twin gestations. Among all twins, monochorionic, monoamniotic (MCMA) gestations have the greatest propensity for poor obstetric outcome. Out of the three types of twin gestations, monoamniotic monoamniotic twinning is rare, occurring in less than 1% of pregnancies. A single amniotic sac with a single placenta is the typical finding in monoamniotic twins. There is a possibility that monochorionic diamniotic twins can have rupture of preexisting inter-twin membrane either spontaneously or iatrogenically from an invasive procedure, and they are then considered pseudo-monoamniotic twins. Once this membrane is disrupted, the pregnancy is functionally similar to monoamniotic twins, with an increased perinatal mortality rate.

Case Description

We present a case of pseudo-monoamniotic twins derived from a mono-chornionic diamniotic twin gestation. A 34 year old G4P1112 with monochorionic diamniotic twin gestation was found to have twin anemia polycythemia sequence (TAPS). She was referred for fetal fetoscopy and laser ablation at 22 weeks estimated gestational age. After presenting for the procedure, preceding ultrasound was performed and global amnion chorion separation was seen with evidence of entangled umbilical cords. This confirmed a now monochorionic monoamniotic twin gestation. The patient was no longer a candidate for fetoscopic surgical intervention. The remainder of the pregnancy was managed inpatient as a monoamniotic twin gestation complicated by twin anemia polycythemia sequence and fetal growth restriction of both twins in the less than third percentile. The patient underwent delivery via primary cesarean section at 26 weeks 6 days estimated gestational age after twin 1 (donor twin) was found to have reversed umbilical artery end diastolic flow. Twin 1 demised on day two of life.

Discussion/Conclusion

Spontaneous rupture of the dividing membrane in Mono Diamniotic twin gestations is a rare condition that poses diagnostic and management challenges. As demonstrated in this case, it can hinder treatment for conditions such as TAPS and twin-twin transfusion syndrome. Literature review demonstrates multiple case reports of pseudo-amniotic twin gestations but there is little understanding of how or why it happens. Further research and analysis of existing data is needed to further understand this complication.

<u>Abstract #57</u> Incidental Endometrial Cancer and Lynch Syndrome following Hysterectomy for Prolapse

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Introduction

Pelvic organ prolapse is a benign condition that is managed conservatively or definitively with surgery. Routine preoperative urogynecologic assessment includes a thorough history, physical exam, and up-to-date preventative screening, including Pap smear. Additional work-up, such as endometrial sampling, is not required preoperatively unless indicated by patient's history to rule out malignancy. In postmenopausal women without abnormal uterine bleeding, the risk of unexpected premalignant or malignant endometrial pathology is up to 2.6% after hysterectomy. If premalignancy or malignancy is found, it warrants further evaluation by a gynecologic oncologist.

Case Description

A 69-year-old postmenopausal female G3P3003 presented for initial evaluation of pelvic organ prolapse. Preoperative work-up consisted of a prior up-to-date negative Pap smear. Family history notable for father with lung cancer. No ultrasonography or endometrial sampling were obtained as not indicated per patient's history. Patient desired definitive management with surgery and underwent a total vaginal hysterectomy with concurrent prolapse procedure. Her postoperative course was complicated by postoperative bleeding requiring removal of bilateral ovaries. On review of pathology specimens, a grade 1 endometrial endometrioid adenocarcinoma was identified. Immunohistochemical analysis showed lack of MSH6 gene, which is highly suggestive of Lynch syndrome. Due to findings, patient was referred to Gynecology Oncology. To date, patient is undergoing surveillance of her malignancy with Gynecology Oncology and referred to a genetic counselor for evaluation of possible Lynch syndrome.

Discussion/Conclusion

Endometrial cancer following hysterectomy is rare. Staging for endometrial cancer is completed surgically. Surgical staging involves a total hysterectomy, bilateral salpingo-oophorectomy, and lymph node assessment. In this patient, only lymph node assessment was not completed. The risk of lymph node involvement with her malignancy was discussed, determined to be low risk, and a shared decision was made for surveillance. Also, due to the risk of Lynch syndrome on pathology, the patient was also referred to a genetic counselor. Lynch syndrome increases the risk of malignancies, including endometrial cancer. The estimated risk of endometrial cancer is 16-61% in women with Lynch syndrome through age 70. As this patient has not yet completed genetic counseling and testing, the diagnosis of Lynch Syndrome is undetermined. However, if the patient were diagnosed, she would require colon cancer screening every 1-2 years. Furthermore, the patient's family would need further genetic counseling and testing. This case highlights the rarity of endometrial malignancy with possible concurrent Lynch syndrome following hysterectomy and the importance of an extensive family history preoperatively.

Abstract #62 Ovarian Cancer Post-Hysterectomy with Unilateral Oophorectomy

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Introduction

Hysterectomies without a unilateral/bilateral oophorectomy are one of the most common surgeries performed in the United States (US). One of the main benefits to leaving the ovaries intact after a hysterectomy is prevention against cardiac disease. The risk of ovarian cancer remains with retained ovaries. Informed consent needs to be utilized when discussing the risks and benefits of oophorectomy with patients.

Case Description

Patient is a 74 y/o female who presented to her primary care physician with a several month history of abdominal pelvic pain, fatigue, night sweats, weight loss, hematochezia, stool incontinence, and malodorous vaginal discharge. Past medical history included hysterectomy with unilateral oophorectomy in 1995 for benign reasons and normal colonoscopy in 2021.. Family history was positive for CAD and hypertension. There was no known family history of cancer.

Workup of current symptoms revealed an elevated WBC, iron deficiency anemia, significantly elevated CA-125 level, and unremarkable CEA levels. Physical Exam and imaging studies revealed a large pelvic mass that was consistent with a malignancy vs a chronic abscess communicating with the rectum. Patient was taken to the OR for exploratory laparotomy, tumor debulking, sentinel lymph node dissection, and colostomy formation. Biopsy revealed advanced high-grade adenocarcinoma, primary gynecologic tumor.

Discussion/Conclusion

Fifty five percent of hysterectomies performed in the US are performed in conjunction with bilateral oophorectomy. This premenopausal patient underwent a unilateral oophorectomy and hysterectomy for benign reasons. Leaving one or both ovaries intact has been shown to prevent the estrogen deficiency which increases the risk of cardiac disease, bone demineralization, subsequent fractures, and depression. Cardiac disease is one of the major causes of death in the US, and the risk increases significantly in post menopausal women.

This case study is a unique look at the consequence of developing a gynecologic cancer post-hysterectomy and unilateral oophorectomy. In this case, if both ovaries were removed prophylactically, ovarian cancer would have been prevented, but it would have put her at an increased risk of the consequences of estrogen deficiency. Informed consent is crucial when deciding whether or not to perform a prophylactic bilateral oophorectomy along with a hysterectomy.

Pediatrics

Abstract #2 Orbital Cellulitis with Thrombophlebitis

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Introduction

Orbital cellulitis is an infection of the orbit, involving the muscle and fat, but excluding the globe. Typical symptoms include eye swelling and erythema, ocular pain, proptosis, ophthalmoplegia, and pain with eye movements. Most cases are associated with coexisting rhinosinusitis, while less common causes are ethmoid and pansinusitis. Staphylococcus aureus and streptococci are the most common bacteria identified. Rare complications result from infraorbital or intracranial extension, including vision loss, empyema, brain abscess, meningitis, or cavernous sinus thrombosis. This case demonstrates the rare and serious complication of thrombophlebitis in an adolescent with orbital cellulitis.

Case Description

A 15-year-old adolescent presented to the emergency department with three-day history of progressive right eye swelling, proptosis, ophthalmoplegia, pain with eye movement, fever, headache, cough, nasal congestion, neck stiffness, and pain with rightward rotation. Pertinent labs showed mild leukocytosis (10.8) with elevated inflammatory markers (CRP 16.92, ESR 31). CT orbits with contrast demonstrated right periorbital cellulitis, right sided pansinusitis, and septic thrombophlebitis of the right superior ophthalmic vein. MRI brain and orbits confirmed right thrombosis of superior ophthalmic vein consistent with thrombophlebitis.

Vancomycin was initially started but switched to ampicillin/sulbactam after consultation with Infectious Disease. ENT was consulted and an endoscopic sinus surgery with right maxillary antrostomy, right total ethmoidectomy, and right frontal sinusotomy was performed. Postoperatively, 24 hours of dexamethasone was completed alongside a three-day regimen of nasal saline rinse and oxymetazoline. Maxillary sinus culture grew coagulase negative staphylococcus and Eikenella corrodens. Blood culture grew viridans streptococcus which was considered a contaminant. Pediatric Hematology recommended enoxaparin for the ophthalmic vein thrombophlebitis. Thrombophilia work-up included PT, INR, PTT, and Fibrinogen which were unremarkable. Ophthalmology exam indicated healthy optic nerves prompting recommendations for continued antibiotics and outpatient follow up. After six days, repeat MRI showed improved orbital swelling without abscess and stable ophthalmic vein thrombophlebitis. The adolescent was discharged after seven days with enoxaparin and three weeks of amoxicillin/clavulanate with plans to follow up with Pediatric Hematology.

One month after discharge, repeat MRI orbits showed stable chronic thrombosis of the right superior ophthalmic vein with improvement of cellulitis. Hematology continued enoxaparin treatment for an additional 3 months with plans to repeat MRI orbits and test for Protein C deficiency after completing anticoagulation therapy.

Discussion/Conclusion

Thrombophlebitis is a rare complication of orbital cellulitis. Clinical suspicion of orbital cellulitis should prompt early neurologic imaging and a multidisciplinary approach to reduce morbidity and mortality of thrombophlebitis in children.

Abstract #23 Alveolar Rhabdomyosarcoma in Adolescent with Hip Pain

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Introduction

Rhabdomyosarcoma is a relatively rare cause of pediatric cancer but is the most common soft tissue sarcoma in childhood. While thought to originate from precursor cells destined to form skeletal muscle, they can arise in mesenchymal tissue anywhere in the body. This is especially true in adolescents, as they most commonly originate in an extremity. We present an adolescent male with metastatic Rhabdomyosarcoma presenting with unilateral leg swelling.

Case Description

A 16-year-old with recent history of acute pancreatitis presented to the pediatric emergency center with a two-week history of worsening left lower extremity pain and swelling. The patient's primary care physician had recently initiated workup with a left lower extremity venous doppler which demonstrated no evidence of deep vein thrombosis (DVT) but multiple enlarged lymph nodes in the groin. This was further evaluated with a CT chest, abdomen, and pelvis. Imaging showed multiple sites of massive lymphadenopathy in various regions of the pelvis and abdomen, as well as a right lung lesion. The adolescent was found to have significantly elevated LDH, mildly elevated uric acid, and significant left inguinal, mediastinal, and intra-abdominal lymphadenopathy with venous congestion and compression of the left external iliac vein and portal vein without thrombosis. Further history revealed a 27-pound weight loss in the preceding 3 months.

Pediatric hematology and oncology was consulted, and the adolescent was started on IV hydration and allopurinol due to concern for tumor lysis syndrome. Pediatric surgery was consulted for inguinal lymph node biopsy with a preliminary report describing a small round blue cell tumor. Final biopsy results were consistent with Alveolar rhabdomyosarcoma with FOX01 fusion protein. Tumor markers AFP and beta-hCG were obtained and within a normal range. MRI brain was completed with no metastasis found. Subsequent imaging would identify the left foot as the primary tumor site.

Discussion/Conclusion

While the initial presentation of rhabdomyosarcomas is variable, and lymph node involvement is common, a non-tender mass is often present if the primary tumor site is within an extremity. This adolescent's presentation was unusual, as there were no clinical findings at the primary site. This led to the disease going unnoticed until presenting with metastatic disease. This case emphasizes the importance of ongoing evaluation in patients with persistent leg swelling with systemic symptoms.

<u>Abstract #28</u> Failure to Thrive Leading to Delayed Diagnosis of Severe Coarctation of the Aorta

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Introduction

Failure to thrive can be a symptom of a more pervasive medical diagnosis such as nutritional neglect; however, it may also be the result of an underlying condition. As such, when a medical provider is confronted with a child with poor weight gain, a thorough medical history, including feeding history, and exam should be completed to evaluate for all causes of failure to thrive.

Case Description

A 6-month-old female dichorionic diamniotic twin born via c-section at 38 weeks presented to the local children's advocacy center with child protective services for an evaluation of failure to thrive. Previous medical and social history included multiple missed appointments and poor weight gain following 100 grams of weight loss over 2 months, among other social concerns, prompting a CPS referral. Newborn screening, including the critical congenital heart defects screen, was normal. At the time of presentation, the infant's diet consisted of 4-6 ounces of formula every 4 hours, rice cereal, and baby foods. Notably, the other twin was healthy. Upon physical exam, the infant was found to be significantly tachypneic with retractions, head bobbing, and non-palpable femoral pulses. Respiratory distress led to admission to the local children's hospital where oxygen was started. Physical exam revealed a SpO2 of 100%, gallop, weak 1+ brachial and femoral pulses bilaterally with brachio-femoral delay. Tachypnea continued with difficulty feeding. An RPP was positive for Rhino/Enterovirus and Adenovirus. Initial chest x-ray showed cardiomegaly and bilateral perihilar pulmonary infiltrates prompting a consult to pediatric cardiology. An echocardiogram demonstrated severe dilation of the left atrium, severe dilatation and moderate hypertrophy of the left ventricle with severely decreased systolic function, bicuspid aortic valve, and severe coarctation of the aorta. The infant was transferred to the PICU due to cardiogenic shock where intubation, alprostadil and milrinone were initiated after stabilization. The infant was then transported to another facility where extended end-to-end anastomosis and PDA ligation was completed. Regular follow-up with pediatric cardiology was initiated.

Discussion/Conclusion

Failure to thrive can be multifactorial in etiology. While most cases of failure to thrive are driven by an inorganic cause including improper mixing of infant formulas, feeding refusal, or parental neglect, this case reinforces the importance of keeping a broad differential. Additionally, the literature indicates the most common lesion to be missed during CCHD screen is coarctation of the aorta. This case highlights the crucial need for thorough physical exams, broad differential, and detailed documentation.

<u>Abstract #32</u> Primary Spontaneous Pneumothorax in An Otherwise Healthy Term Neonate

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Introduction

Primary spontaneous pneumothorax (PSP) is air accumulating between the visceral and parietal pleurae following visceral pleura rupture in the absence of explainable underlying lung disease. PSPs are typically caused by significant changes in transpulmonary pressure causing alveolar distension and eventual rupture. Common causes of respiratory distress leading to neonatal PSP include respiratory distress syndrome, transient tachypnea of the newborn, and persistent pulmonary hypertension. While small pneumothoraces are often asymptomatic and incidentally discovered, large pneumothoraces in newborns cause significant distress. Symptomatic pneumothoraces occur in approximately 0.02% percent of live births with increasing rates in infants who are premature, large for gestational age, or have underlying causes for lung disease. This case highlights an otherwise healthy term neonate without risk factors who developed spontaneous pneumothorax at 24 hours of life.

Case Description

A multigravida woman with an unremarkable prenatal course delivered a 2840-gram neonate at 37 weeks gestation via routine spontaneous vaginal delivery. The neonate had APGAR scores of 8 and 9 and received routine care immediately after birth. At 16 hours of life, the neonate was stable and breastfeeding well. At 24 hours of life, routine critical cardiac congenital heart disease (CCHD) screening revealed a pre-ductal value of 88% and a post-ductal value of 92% with desirable values > 95%. Upon physical exam, the neonate had associated tachypnea, grunting, subcostal retractions, and tracheal tugging. Following an immediate transfer to the NICU, chest X-ray revealed a large, right-sided pneumothorax with leftward mediastinal shift. Needle thoracentesis decompression was successful, and the neonate was supported with high-flow oxygen. Subsequent evaluation did not discover an underlying cause. Repeat chest X-ray at 48 hours displayed resolution of the pneumothorax without recurrence. High-flow oxygen support was utilized for a total of 4 days and the neonate was discharged in good health on day 18 of life after tolerating oral feedings.

Discussion/Conclusion

While the most common causes of respiratory distress in term neonates are typically seen within the first several hours following birth, spontaneous pneumothoraces can sometimes occur without provocation and can have a high associated morbidity and mortality if not treated swiftly. Managed conservatively, fifty to sixty percent of neonates with PSP will experience recurrence while pleurodesis and thoracentesis greatly reduce the probability of recurrence. This case highlights the importance of monitoring healthy neonates for a full 24 hours after birth and completing all necessary screens prior to discharge to avoid unforeseen complications.

Abstract #35 Burkitt Lymphoma

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Introduction

Burkitt lymphoma (BL) is an aggressive type of non-Hodgkins's lymphoma (NHL) that affects B-cells most commonly in children, and typically presents as enlarging lymph node masses found in the chest and/or the abdomen. Primary tumors commonly spread to the brain and/or spinal cord, but can also be found in the bone marrow, spleen, and liver. Endemic BL is highly associated with EBV in Africa and other locations where malaria is found, but sporadic BL is rarely associated with this viral illness.

Case Description

A 7-year-old male presented to the hospital for an evaluation of a right sided neck mass. Past medical history included complaints of right sided neck pain for the last two months in addition to fatigue and intermittent back pain. The physical exam was significant for poor dentition and a firm, fixed, 6 x 4 cm mass below the right side of the mandible. Labs were grossly unremarkable with normal CBC and no sign of renal dysfunction or liver infiltration were evident. Additionally, LDH and uric acid levels were WNL at 242 and 4.5 respectively. There was leukocytosis of 15.3 with elevated platelets to 484 and an ESR at the upper limits of normal at 9. Serology testing was only positive for past exposure to CMV and EBV. A three-day course of clindamycin was prescribed. CT was obtained which showed an infiltrating mass on the sternocleidomastoid causing a mass effect on the right sided jugular vein, carotid sheath, and submandibular gland. Based on these findings, the differential included sarcoma, branchial cleft cyst, dermoid, lymphoma and teratoma. Pediatric Hematology/Oncology was consulted and expressed concerns for a possible catecholamine producing tumor. Plasma metanephrine levels were obtained to rule out a possible paraganglioma before a fine needle aspiration and core needle biopsy could be obtained. Upon pathology evaluation of the biopsy, the child was diagnosed with Burkitt Lymphoma.

Discussion/Conclusion

Although our child in this case had fatigue and right sided neck pain, he did not exhibit the other clinical signs often seen in BL such as painless lymphadenopathy, fatigue, weakness, loss of appetite, weight loss, rapid growth rate, hepatosplenomegaly, and cytopenia or other lab abnormalities. His presentation, which resembled lymphadenitis, likely delayed appropriate diagnosis. As such, this case highlights the importance of an in-depth history and physical exam followed by proper diagnostic procedures to correctly diagnose unexpected disease.

Abstract #44 Accidental Gunshot Wound in Pediatric Patient

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Introduction

Blunt or penetrating thoracoabdominal firearm trauma necessitates a multi-disciplinary approach in the acute and long-term settings. We report an accidental self-inflicted gunshot wound to the upper abdomen resulting in a perforated inferior vena cava (IVC), shattered thoracic vertebrae, perforated stomach, duodenum, and jejunum, spinal cord injury with lower extremity paraplegia, nutritional intolerance, and severe emotional distress.

Case Description

A 2-year-old presented to the emergency department in hemorrhagic shock after an accidental self-inflicted gunshot wound through the abdomen, necessitating an emergent exploratory laparotomy and massive transfusion protocol. The child was admitted to the PICU, initially requiring central and arterial line placements, intubation, vasopressors, and post operative antibiotics. During this stay, the child regressed to a mostly non-verbal state and was clearly distressed by medical attention without a mature coping mechanism to alleviate fear. The child developed traumatic rhabdomyolysis, pancreatitis, and hemiplegia of the left leg due to L3/L4 vertebral fractures with bone fragments in the left side of the spinal canal. The child's IVC injury resulted in significant lymphedema with pressure ulcers in the lower extremities, and an intolerance of oral feeds resulted in ultimate dependence on Jejunostomy tube feeds for nutrition. After a 20-day hospital stay, the child was discharged to a pediatric rehabilitation center with significant improvement in stability and movement.

The child's extensive inpatient care team prioritized presenting to the parents as a unified front to minimize confusion and ongoing trauma associated with unclear expectations and advancements of care. Nurses worked to coordinate care with physicians to minimize interruptions and avoid unnecessary emotional distress. With DHS involvement, the child's parents were allowed to remain at bedside and make all medical decisions throughout hospital stay, helping the child adjust to unfamiliar environments, anxiety, and stress.

Discussion/Conclusion

In this case, interdisciplinary communication was imperative, yet the varying schedules and plans of different specialties presented a logistic challenge to organize care cohesively. At times, it was necessary for the general pediatric hospitalist team to address conflicting advice between specialties and work with various providers to come to the best solution for the child in regards to all affected organ systems. Optimizing recovery included minimizing the impact on child development. Identifying medical and social challenges, utilizing resources to promptly address evolving needs, and effective communication with all care providers is challenging, but essential in the hospital setting.

<u>Abstract #45</u> Atypical Presentation of DRESS Syndrome in an African American Male

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Introduction

Drug reaction with eosinophilia and systemic symptoms (DRESS), a rare yet potentially fatal drug reaction, typically presents with an extensive maculopapular erythematous rash 2-4 weeks after exposure to the causative agent. Systemic symptoms involve fever and hematologic irregularities, with eosinophilia, transaminitis, and leukocytosis being most common. Its mortality rate is about 10%, highlighting the critical need for early recognition and adequate treatment. We present a severe case of DRESS requiring high dose IV steroids with an atypical mild eczematous rash presentation.

Case Description

A 17-year-old African American male presented to the ED with one-month history of diffuse and pruritic rash, myalgias, and weight loss, along with 2-week history of intermittent fevers, increased somnolence, and facial swelling. Symptoms had not improved despite prior treatments with diphenhydramine and prednisone, followed by amoxicillin and cephalexin for presumed infections at urgent care. Of note, the rash markedly worsened after cephalexin use. On presentation, he was febrile to 102.2 F and tachycardic at 141. Physical exam revealed dry, eczematous skin on the trunk, back, bilateral arms, and bilateral legs. Ulcerated lesions measuring 1mm in diameters with a pink base were also noted on upper and lower back with accompanying abdominal striae. No marked skin sloughing or oral ulcers were observed.

Lab work revealed leukocytosis, eosinophilia, normocytic anemia, and transaminitis in addition to an elevated CRP (15.8) and ferritin (> 13,000). Clindamycin was initiated for concern of super-imposed bacterial infection, and Allergy/Immunology was consulted. Peripheral smear and parasitic testing were negative. Repeat labs the following day showed marked increase in LFTs and eosinophilia, most consistent with DRESS Syndrome. High dose IV methylprednisolone 1mg/kg twice daily was initiated along with hydroxyzine and cetirizine for pruritis. Repeat labs prior to discharge showed marked reduction in LFTs, eosinophils, and ferritin and resolution of symptoms. The patient was then discharged with a 4-week corticosteroid taper with outpatient follow up in allergy clinic.

Discussion/Conclusion

Rapid recognition of DRESS and discontinuation of the causative drug, coupled with timely initiation of treatment, is imperative for clinicians to reduce patient mortality and morbidity. In our case, the initial rash likely had a different etiology, but the exposure to cephalexin or amoxicillin is what worsened his clinical status and caused DRESS. Fortunately, he had already been off of antibiotics for >1 week at presentation. Clinicians should be vigilant regarding high-risk medications and recognize atypical rash presentations to ensure early and accurate diagnosis of DRESS.

Abstract #47 Septic Shock and Pancytopenia Associated with Parvovirus B19

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Introduction

Human Parvovirus B19 (PB19) is a single-stranded DNA virus. Certain risk factors including pregnancy, blood disorders, and immunosuppression can increase risk of significant complications from PB19. Septic shock and pancytopenia are two potential life-threatening complications, but are extremely uncommon, especially in otherwise healthy people. We report a pediatric case of septic shock and pancytopenia associated with a PB19 infection in a previously healthy patient.

Case Description

A 6-year-old female with no significant past medical history was admitted to the PICU after initially presenting to an outlying ED with rash, fever, fatigue, and swollen hands/knees for six days. She had hypotension in the 80s/30s despite multiple fluid boluses, norepinephrine drip, and antibiotics prompting transfer to our PICU with worsening septic shock and possible bacterial meningitis.

Upon arrival, she had generalized pain, petechial rash, ecchymosis, edema with refractory septic shock, generalized anasarca, DIC, bandemia, pancytopenia, hypomagnesemia, hypoalbuminemia, hypophosphatemia, elevated inflammatory markers, and acute hypoxic respiratory failure requiring intubation.

LP showed no evidence of meningitis. Antibiotics, IVF, inotropic medications, and FFP/platelet transfusions were administered. Imaging showed mild hepatosplenomegaly/ascites. Differential included autoimmune disorders, infectious organism, and rare conditions like MISC and HLH. ID, Hematology/Oncology and Allergy/Immunology were consulted prompting further work-up for HLH and other infectious etiologies.

Comprehensive immunodeficiency work-up including ANA, rheumatoid factor, immunoglobulin levels, and serum CXCL9 were unremarkable. Blood, urine, and CSF cultures showed no growth. Subsequent labs were negative for tickborne disease and immunodeficiency, and positive for SARS-COVID 2 antibodies and PB19 PCR. CXR revealed pleural effusion, treated with furosemide. IV hydrocortisone was given for low cortisol. MRI brain performed for acute mental status change resulted normal. IVIG 2g/kg was given on day 3 with improvement in symptoms, and resolution of pancytopenia. Patient was extubated on day 5 and transferred to the general floor. PB19 IgM and IgG antibodies were elevated indicating acute infection, while ASO titers and antibodies for CMV, EBV, and HHV-6 were negative. The positive PB19 PCR with elevated IgG/IgM along with lab results and presentation made PB19 viral sepsis the most likely diagnosis.

Discussion/Conclusion

This case showcases a unique presentation of septic shock and pancytopenia in PB19 infection, deviating from typical presentations. It also highlights the parallels between PB19 viral sepsis and HLH, including fever, cytopenia, elevated ferritin, and neurologic findings. Clinicians should be aware of the existence of parvovirus-induced septic shock and pancytopenia, keeping it in their differential when encountering patients with these atypical manifestations.

Abstract #55 Refeeding Syndrome in the Setting of Child Neglect

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Introduction

Refeeding syndrome is a serious and potentially life-threatening condition that may arise following rapid initiation of refeeding following a sustained period of undernutrition. Close monitoring of lab work is key in identifying early signs of refeeding syndrome as there are specific associated electrolyte imbalances such as hypophosphatemia, hypokalemia, and hypomagnesemia. Additional important potential lab findings include thiamine deficiency, elevated creatine phosphokinase and worsening anemia. Electrolyte imbalances can have both short and long-term effects on the cardiovascular, gastrointestinal, and neurologic systems. Arrhythmias, blood pressure instability, heart failure, ataxia, vertigo, paralysis and respiratory distress are all symptoms that may occur in the setting of refeeding syndrome.

Case Description

A 7-year-old presented to an outside emergency room with altered mental status and parental concern for melatonin overdose. The child was emaciated with a BMI-for-age z-score of -5.94 and bruising to the forehead and eyes. A DHS referral was immediately placed, and the child was transferred to a facility with a pediatric intensive care unit. There the child was found to have severe hypoglycemia, hyponatremia, hypothermia, bradycardia, hypotension, thrombocytopenia and transaminitis. Given the severity of malnutrition, the child was at high risk for refeeding syndrome. They underwent close laboratory and weight monitoring while physicians worked with gastroenterology and dietitians to gradually increase caloric intake. The patient developed clinical signs of refeeding syndrome, though these were minimized via close monitoring of electrolytes and appropriate electrolyte supplementation.

A medical evaluation and forensic interview by the child abuse protection team was completed. Given the severity of the initial presentation and the reported history, the child was diagnosed with child neglect, child torture, and child physical abuse. At time of discharge from their 15-day hospitalization, they were tolerating a normal pediatric diet without restriction along with vitamin and phosphate supplementation. Discharge planning included follow-up at the Child Advocacy Center and with a primary care pediatrician, psychiatry, cardiology and gastroenterology.

Discussion/Conclusion

Refeeding syndrome is a rare but serious complication following severe prolonged malnutrition. Interdisciplinary teamwork is a necessity both during a hospitalization and follow-up for refeeding syndrome. Close monitoring of lab work with regular replacement of electrolytes and vitamins is key to reducing the severity of refeeding syndrome. When severe malnutrition is present in a pediatric patient, an evaluation for child maltreatment is warranted. Diagnoses of child neglect and child torture are likely to result in lifelong negative consequences and necessitate close follow-up with a multi-disciplinary team.

Abstract #65 Undiagnosed Hyperimmunoglobulin Syndrome

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Introduction

Hyperimmunoglobin E Syndrome (HIES) is a recurrent infection syndrome associated with an increase in serum levels of IgE. It is characterized by recurrent pulmonary infections, eczematous dermatitis, and an excoriated and severely pruritic rash due to the presence of Candida and staphylococci in the skin. Facial features such as a broad nasal bridge, protrusion of the forehead, and notable thickening of the soft tissue of the face, give the skin the described appearance of "coarse facies." Patients also tend to have skeletal abnormalities such as scoliosis and osteoporosis associated with minor trauma bone fractures.

Case Description

A 14-year-old, with history of atopic dermatitis and asthma, presented with worsening shortness of breath, cough, acute hypoxic respiratory failure, and wheezing. They were found to have hyperglycemia, leukocytosis, thrombocytosis, eosinophilia, and acute kidney injury. Upon initial presentation, patient was on 3L nasal cannula of supplemental oxygen. Patient's initial physical examination revealed distinguishable features of HIES such as the excoriated bleeding skin manifestations, facial features of frontal bossing and deep-set eyes, and complaints of skeletal abnormalities with back and joint pain. The adolescent was quickly weaned to room air and his asthma was controlled with mometasone/formoterol. Due to a positive MRSA blood culture at 22 hours, Pediatric Infectious Diseases was consulted and recommended IV vancomycin for seven days as well as an echocardiogram, which was unremarkable. Repeat blood cultures were obtained during hospitalization which showed no growth and clinically, the patient's skin drastically improved with daily use of topical ointments.

Upon further investigation of their medical history, the adolescent was seen by Pediatric Rheumatology, four months prior to admission. Blood work revealed IgE levels were elevated at 35,844, with accompanying elevation in eosinophils. The family was unaware of these lab abnormalities. At that time, a referral to Allergy and Immunology was made. However, due to lack of specialist availability, the adolescent was unable to be seen prior to this admission. The adolescent had inconsistent follow up with Dermatology and Pulmonology for the previous diagnoses, which contributed to worsening symptoms requiring hospital admission.

Discussion/Conclusion

This case shows that those with suspected HIES require specialized, close care. Failure to follow-up and lack of accessibility to specialties can contribute to issues that require admission, as management requires a multidisciplinary team. With management focused on skin care, prevention of infection, and control with pulmonary and skeletal complications, patients have a successful prognosis.

Surgery

<u>Abstract #1</u> Persistent Intra-operative Hypotension in the Prone Position During Posterior Fossa Craniotomy

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Introduction

Intraoperative hypertension frequently presents itself as a significant complication in patients undergoing surgery under general anesthesia, particularly when placed in the prone position. Primary risk factors and dynamic hemodynamic shifts may contribute to this change during surgical procedures. Various management strategies, including the use of vasopressors and positioning tools, are employed to modulate and mitigate this shift.

Case Description

A 52-year-old male patient with a past medical history of class II obesity (BMI 38), hypertension, and hyperlipidemia underwent a posterior fossa craniotomy for removal of a cystic mass observed on MRI in the cerebellar vermis with features concerning for possible primary brain tumor or hemangioblastoma. Preoperative anesthesia assessment one week before surgery disclosed a recent onset of right-sided hearing loss. The patient was determined to be ASA category 3b, Mallampati IId, with normal vital signs. Additionally, patient medications included metoprolol and losartan for treatment of chronic hypertension. The neurosurgery team elected to proceed with posterior fossa craniotomy to remove the brain lesion.

Patient vitals were noted to be within normal limits following induction and intubation. However, following the placement of the patient in the prone position, the patient developed severe hypotension with MAP decreasing from 108 to 30 over the next 20-30 minutes. The patient received escalating doses of phenylephrine, norepinephrine, epinephrine, and vasopressin, all without an adequate response. Appropriate adjustments were made to sevoflurane and propofol administration rates, and 2500 ml of normal saline was provided. The lack of response to numerous interventions prompted anesthesiologists to confirm that the arterial line was functioning correctly. Eventually, blood pressure normalized, the remainder of the case was uneventful, and the patient was transported to PACU. In total, the patient was markedly hypotensive for over two hours, with a MAP ranging from 30 to 49.

Discussion/Conclusion

Intraoperative hypotension can be a challenging complication to manage during cases requiring prone positioning, such as a posterior fossa craniotomy. This case presentation describes a patient scenario involving severe persistent intraoperative hypotension in the prone position and the primary causes and management strategies for this condition.

Decreased stroke volume and cardiac index are the main hemodynamic changes attributed to hypotension following prone positioning. Management of the associated hypotension can involve adequate chest support modalities and routine pharmacologic interventions such as phenylephrine, norepinephrine, and vasopressin. Overall, anesthesiologists must consider the effect of patient positioning, especially the prone position, on blood pressure during a case.

<u>Abstract #24</u> Intracecal Aorto-femoral Bypass Graft Requiring Multiple Operative Interventions

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Dr. Lucas Phi - OU-TU School of Community Medicine, Department of Surgery

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Dr. Stuart Hoff - OU-TU School of Community Medicine, Department of Surgery

Dr. Peter Nelson - OU-TU School of Community Medicine, Department of Surgery

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Introduction

Arterial bypass graft infections remain a challenging complication, often requiring multidisciplinary teamwork to manage, with most requiring surgical removal of the graft to gain control. Adding to this complexity, these patients not only require graft removal but also concurrent revascularization. Here we discuss a presentation of graft infection that required a creative, multidisciplinary approach to manage.

Case Description

We present a 57-year-old man with hypertension, hyperlipidemia, and aortoiliac occlusive disease who underwent an open aorto-bifemoral bypass graft six months ago. This was immediately complicated by a small bowel obstruction requiring exploratory laparotomy, and not long after this the patient was found to have Klebsiella pneumoniae bacteremia. In this most recent admission he presented with fatigue, fever, and chills. Further work up was notable for methicillin-sensitive Staphylococcal aureus bacteremia as well as imaging findings concerning for a possible aorto-colonic fistula versus intracecal segment of the bypass graft. The patient's antibiotic regimen was broadened and he was taken for an exploratory laparotomy with an expansive team. This included urology for ureteral stent placement, as well as both the vascular and general surgery services. After exploring the abdomen, the right limb of the graft was clamped and divided, and a partial cecectomy and appendectomy was performed to remove the segment en bloc. The specimen was then opened to confirm that the graft was intraluminal. The abdomen was then irrigated and closed, and a femoral-femoral bypass graft was performed to revascularize the right leg. The patient's course involved a prolonged ileus, complex pain management, and physical therapy. He was discharged two weeks after surgery on an 8-week course of intravenous antibiotics based on infectious disease recommendations with plans to transition to lifelong oral suppressive antibiotics.

Discussion/Conclusion

This represents a delayed recognition of a technical complication presenting with polymicrobial bacteremia, recent bowel obstruction, and eventual confirmation of an arterial graft infection. While these presentations may be viewed as isolated events, this case demonstrates the importance of considering the entire patient history for the best assessment. Any obstructive symptoms in the setting of recent intra-abdominal surgery may indicate an iatrogenic bowel injury. Furthermore, this case highlights the importance of proper technique when tunneling any medical device. Likely, this graft was aberrantly placed through the colon in the initial operation which went unrecognized initially despite the obstructive symptoms and ongoing bacteremia. Though definitive surgical management was complex, a thought-out and deliberate multispecialty surgical approach was successful.

Abstract #52 Heterotopic Pancreas: An Unlikely Finding During Bariatric Surgery

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Introduction

Heterotopic Pancreas (HP) is a rare, benign pathology that can be discovered incidentally during surgery or suspected with radiographic imaging. This case provides an overview of the key features and clinical implications of heterotopic pancreas, and management when identified at the time of concurrent planned gastrointestinal surgery.

Case Description

47-year-old female presented for elective laparoscopic Roux-en-Y gastric bypass after successfully completing a multidisciplinary weight loss program. Past medical history included stage IIA melanoma, hypertension, and symptomatic GERD. During surgery, a 2.5 cm exophytic mass was identified on the antimesenteric border of jejunum 15 cm from the Ligament of Treitz. Given the nodular and irregular appearance, the mass was resected with a margin of normal small bowel and frozen section demonstrated benign heterotopic pancreatic tissue. Fluorescent angiography demonstrated that the bowel in the area of resection had adequate perfusion. The gastric bypass was completed without complication and the patient had a normal postoperative course and was discharged on postoperative day 1.

Discussion/Conclusion

Heterotopic pancreas is the presence of ectopic pancreatic tissue, most commonly found in the stomach, duodenum, jejunum, and Meckel's diverticulum. The jejunum is the 3rd most common location behind duodenum and stomach. Most cases are asymptomatic and discovered accidentally during surgical procedures or autopsy with reported incidences of 0.5% and 0.6-14% respectively. Surgical resection is the mainstay of treatment to prevent future complications and obtain a permanent section. While most cases are clinically silent, the most common presenting symptoms can include nausea and vomiting, epigastric pain, ulceration, and weight loss. In some cases, HP can be identified on imaging and will appear as a small oval intramural mass with microlobulated margins and an endoluminal growth pattern.

Heterotopic pancreas, also known as pancreatic rest or ectopic pancreas, is a rare congenital anomaly characterized by the presence of pancreatic tissue in locations outside the normal pancreas. This ectopic tissue retains the histological characteristics of the pancreas, including acini, ducts, and islets of Langerhans. Heterotopic pancreatic tissue can be found in various gastrointestinal sites, most commonly in the stomach, duodenum, and jejunum. Definitive diagnosis is made with pathologic evaluation, and resection of suspected heterotopic tissue can be safely done minimally invasively when identified during abdominal surgery. Frozen section can facilitate real time diagnosis, and also allow for additional surgical management if indicated based on pathologic findings.

<u>Abstract #59</u> Physician Modified Endograft in a Pararenal Abdominal Aortic Aneurysm

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Introduction

Abdominal aortic aneurysm (AAA) is a life-threatening condition necessitating the development of surgical techniques adaptable to the patient's anatomy to optimize outcomes. Endovascular aneurysm repair (EVAR) is a minimally invasive surgical approach that recreates the aortic lumen at the site of aneurysm to prevent further dilation and fatal aortic rupture. Physician modified endografts (PMEG) improve EVAR by enabling surgeons to customize a unique graft based on a patient's anatomy. EVAR with PMEG facilitates AAA repair and preservation of blood flow in major branching arteries of the abdominal aorta. We present a case of a patient in need of one such graft.

Case Description

A 74-year-old man presented to an outside ED with worsening shortness of breath. CTA of the chest confirmed right pulmonary embolism as well as a 6.8 cm x 8.6 cm pararenal AAA. This was an incidental finding as he was asymptomatic for AAA denying abdominal pain, melena, hematochezia, or leg pain with ambulation. A pulsatile mass could be palpated on abdominal exam correlating with the AAA location identified on CTA. An EVAR with four vessel PMEG was planned using a Terumo Treo endograft.

The position of fenestrations on the graft were planned using 3D reconstruction of the patient's anatomy. The graft was then removed from its sheath and the fenestrations were created using cautery. A packing coil was then sutured to the edge of each fenestration to act as a radiopaque marker for later cannulation. The graft was then constrained and then resheathed. Access wait obtained in both femoral arteries and the graft was partially deployed. Fusion imaging was used to assist with locating the orifice of each visceral vessel. Each visceral vessel was then cannulated with wires and the graft was then fully deployed. Stents were placed in each visceral vessel and the rest of the main body and contralateral limb was then deployed. A live CT was then done upon completion. The patient tolerated the procedure well and was discharged in one week.

Discussion/Conclusion

While EVAR with PMEG is not utilized by most vascular surgeons this case illustrates its efficacy for patients with pararenal AAA. Other endovascular options for treating this disease process include using off the shelf branched or fenestrated devices such as the Cook Zenith Fenestrated device but these devices take time to be constructed. The use of PMEG expands the spectrum of disease able to be treated with minimally invasive measures.

Abstract #63 Endovascular Minimally Invasive Management of Multi-Organ

Trauma

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Introduction

Blunt traumatic aortic injury (BTAI) is the second most common cause of death in trauma patients. Of the 20% of patients who survive to hospital arrival, 50% will die within 24 hours. Additionally, splenic injury represents approximately 31-35% of all blunt abdominal traumatic injuries making it the most frequently injured solid organ with total mortality of 6-7% percent. Here we present a case involving a restrained driver status post motor vehicle collision with blunt thoracic and upper abdominal trauma resulting in class III blunt thoracic aortic injury, a grade IV splenic laceration, a maxillofacial fracture and an open book pelvic fracture. Vascular consultation by the trauma team permitted a minimally invasive procedure that addressed multiple lesions in one operation in a coordinated and efficient approach.

Case Description

A 30-year-old male with a past medical history of obesity, who was the restrained driver involved in motor vehicle collision, presented to the emergency department as a tier I trauma secondary to mechanism of injury and loss of consciousness. On arrival he was tachycardic with an obvious open book pelvic fracture. After trauma surveys, CT imaging was obtained that revealed dissection of the proximal descending thoracic injury with hemorrhagic products in the mediastinum (grade III BTAI with high-risk features) and a grade IV splenic laceration with intraparenchymal and subcapsular hematomas. Vascular surgery was consulted, and he was taken to the operating room emergently.

Percutaneous arterial access was obtained under ultrasound guidance through the right common femoral artery. An intravascular ultrasound was then used to evaluate the aortic arch and location of the intimal tear in the aorta 2-3 cm distal to the left subclavian artery, and a conformable thoracic stent graft was placed that covered the lesion. An abdominal aortogram was obtained and extravasation of contrast from the splenic artery, consistent with splenic injury, was noted. The splenic artery was coil-embolized to control the hemorrhage. Lastly, a pelvic angiography evaluated for and ruled out any further source of hemorrhage. The entire procedure was performed through a subcentimeter incision and lasted under 2 hours. The patient was admitted to the intensive care unit, and the orthopedic and maxillofacial injuries were addressed in the subsequent days.

Discussion/Conclusion

This case exemplifies an efficient manner to address thoracoabdominal lesions in a minimally invasive manner that results in reduced morbidity, mortality, hospital length of stay, and physical stress on a patient with significant traumatic injuries.

<u>Abstract #64</u> Juvenile Nasopharyngeal Angiofibroma: Endoscopic ENT and Neurosurgical Resection with Pre-Op Embolization

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Introduction

Juvenile Nasopharyngeal Angiofibromas are one of the most common benign sinonasal tract tumors in adolescent males aged 13-22. They are thought to arise from the pterygopalatine fossa, with the sphenopalatine artery serving as a primary source of perfusion for these tumors. That being said, they are a relatively rare occurrence in clinical practice, and a high degree of suspicion must be raised when the right patient comes through the door. These tumors most commonly occur in adolescent males, with the typical clinical presentation including nasal congestion, recurrent epistaxis, headaches, and a nasopharyngeal mass.

Case Description

The present case is of an 11-year-old male who presented to the ER with refractory epistaxis. He had a 3-month history of gradually worsening nasal congestion that had developed into recurrent epistaxis within the past week. MRI imaging showed an enhancing mass within the paranasal sinuses, nasal cavity, and nasopharynx. Interventional radiology went in 24 hours pre-op to embolize the vasculature feeding the mass. ENT and neurosurgery then proceeded with endoscopic resection. The patient recovered well, and post-op MRI demonstrated complete resection of the mass. The patient underwent typical precautions of any major surgery, including weight and activity restrictions, for several weeks post-op. Within a month, he is expected to be back at school and, within 12 weeks, back to full functioning.

Discussion/Conclusion

Care must be taken when working up patients in the office, relying upon imaging secondary to a high index of suspicion, as exam room biopsies may lead to critical bleeding that is unable to be controlled outside of the operating room. Overall, these tumors have fantastic outcomes with the right team to perform the operation. As of late, preoperative embolization paired with endoscopic resection leads to ideal outcomes with minimal blood loss and low recurrence rates. This case represents a rare clinical condition that can be encountered in both the primary care pediatric and emergency room environments. With a high degree of suspicion and proper coordination with specialized teams, including ENT and neurosurgery, within a provider's community, excellent outcomes can be achieved with low morbidity.

<u>Abstract #70</u> Abdominal Cocoon Syndrome Variant: A Rare Cause of Small Bowel Obstruction

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Introduction

Abdominal cocoon syndrome (ACS) is a rare but intriguing etiology of partial or complete small bowel obstructions. A dense fibrous membrane encapsulates the small bowel, creating its characteristic cocoon appearance. Surgical removal of this membrane may reveal significant thickening of the small bowel. ACS, also called peritoneal encapsulation, leads to constriction and distortion of the small intestine, causing bowel obstructions. Diagnosing ACS is challenging due to its rarity and nonspecific symptoms.

Case Description

A 73-year-old male undergoing ambulatory peritoneal dialysis (PD) presented with abdominal pain, nausea and vomiting. He was found to have fungal peritonitis, and his PD catheter was removed. He developed recurrent episodes of partial small bowel obstructions. He was unable to tolerate a diet per os and developed abdominal pain, nausea, and vomiting. On exam, his abdomen was distended and tympanic. He underwent multiple CT scans showing a serpentine configuration of dilated loops of small bowel. His symptoms failed to resolve with bowel rest and nasogastric tube decompression. Gastrograffin small bowel studies demonstrated delayed transit through the small bowel to the colon. The decision was made to proceed with surgery. Diagnostic laparoscopy revealed a leathery brown appearance of most of the small intestine. The entire small bowel was examined, and adhesions to his prior peritoneal dialysis entry site and retroperitoneum were released from this thickened bowel.

Discussion/Conclusion

The pathogenesis of abdominal cocoon syndrome is linked to peritoneal irritation. The resulting inflammation releases fibrin-like material that encases the small intestine. ACS manifests in both primary and secondary forms. Primary ACS is idiopathic, and secondary ACS is associated with discontinuing peritoneal dialysis, liver cirrhosis, and abdominal tuberculosis. The typical features of ACS include the thickening of the small bowel and the formation of a fibrous capsule around it. However, the presented case deviates from this norm, displaying a thickened and leathery small bowel without the characteristic encasing fibrous membrane. This observation in variability suggests a spectrum within ACS, highlighting the complexity of this rare condition.

The scarcity of abdominal cocoon syndrome presents a unique clinical scenario requiring a high index of suspicion for timely recognition and management. Laparoscopy can reveal a thick membrane potentially requiring removal, though diffuse thickening can occur in isolation. Management requires a multidisciplinary approach, including surgical intervention to release encapsulated bowel and address underlying adhesions. Efforts to optimize peritoneal dialysis techniques and minimize peritoneal inflammation are essential in preventing ACS and ACS variants.