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### M035

### HEREDITARY ALPHA TRYPTASEMIA MASKED AS ASTHMA EXACERBATION

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**Introduction:** Clinical phenotypes of hereditary alpha tryptasemia (HAT) are variable and can range from asymptomatic to anaphylaxis. Due to this broad range of presentations, diagnosis can be challenging, particularly if only one organ system is involved.

**Case Description:** An 8-year-old male presented to the emergency department with hypoxemia and stridor. His exam was notable for inspiratory and expiratory stridor with increased work of breathing. He received nebulized racemic epinephrine without improvement, and then had resolution of symptoms with intramuscular epinephrine and oral corticosteroids. Spirometry was suggestive but not diagnostic of asthma. Due to symptoms localized only to the respiratory tract, asthma was initially suspected. However, mast cell disorders were also considered since he required epinephrine before symptom improvement. Serum tryptase was elevated on initial and repeat testing. Genetic testing demonstrated increased copies of *TPSAB1*, consistent with HAT.

**Discussion:** Hereditary alpha tryptasemia is an autosomal dominant disorder resulting from increased copies of *TPSAB1* which encodes alpha tryptase. Disease severity varies from asymptomatic to anaphylaxis. Higher tryptase levels and increased symptom severity parallels increased number of *TPSAB1* copies. Serum tryptase level is the initial screening test. If elevated, genetic testing for *TPSAB1* should be performed. Though presentations with single organ involvement are not normally concerning for systemic disease, severe presentations of atopic diseases, including asthma, should prompt consideration of HAT. Treatment is symptom directed and may include antihistamines, mast cell stabilizers, leukotriene inhibitors, and epinephrine.

### M036

# PARANOID ABOUT RASHES: OLANZAPINE INDUCED DRUG REACTION WITH EOSINOPHILIA AND SYSTEMIC SYMPTOMS

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**Introduction:** Drug Reaction with Eosinophilia and Systemic Symptoms (DRESS) is a severe adverse reaction caused by more than 50 prescription drugs. Although antiepileptics and antibiotics account for most cases, the variety of potential drug classes can make identifying the causative agent challenging.

Case Description: Thirty-year-old male was initially admitted to the medical ICU for treatment of alcoholic hepatitis. Hospital course was complicated by an upper GI bleed, hepatic encephalopathy, respiratory failure, and multiple infections requiring repeated courses of antibiotics. Two months into the hospital stay, he developed a diffuse pruritic maculopapular rash that progressed to cover his trunk, neck and extremities. Workup revealed elevated aminotransferases along with worsening creatinine, leukocytosis, and eosinophilia concerning for DRESS. Medication review revealed the patient received several days of vancomycin in addition to a month-long course of olanzapine for paranoia suffered while inpatient. Patient first received olanzapine 8 weeks prior to symptom onset. HLA-A\*32:01 was obtained and negative. Topical steroids were applied, and potential offending medications held. Rash resolved over the next few weeks followed by clinical improvement. Olanzapine was added to the allergy list to avoid future episodes. Discussion: The diagnosis of DRESS can be challenging as it often occurs in the inpatient setting with patients exposed to varying amounts of multiple drugs, including less common agents such as olanzapine. When multiple culprit drugs have been administered, evaluating factors such as timeline of exposure and genetic predisposition may be helpful in identifying the cause. This can lead to prompt removal of the offending drug minimizing morbidity and mortality.

### M037

### SUCCESSFUL IMIGLUCERASE DESENSITIZATION IN A PATIENT WITH SEVERE ARTHRALGIA AND MYALGIA

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**Introduction:** A patient with Type I Gaucher disease experienced severe musculoskeletal pain within minutes upon first and second administration of imiglucerase, which is a rare adverse event. He underwent successful desensitization, which has only been reported in 3 cases, and continues to receive imiglucerase therapy.

**Case Description:** A 37-year-old male with Type I Gaucher disease developed severe arthralgias and myalgias without skin changes nor symptoms of anaphylaxis within 5 minutes of the first infusion of imiglucerase enzyme replacement therapy. The infusion was stopped, and he was treated with intravenous (IV) fluids, IV hydrocortisone, and acetaminophen. He experienced severe back pain at a second infusion a week later despite pretreatment with IV diphenhydramine and prednisolone. A desensitization procedure was performed using an intermediate risk (3 bag) protocol (44U, followed by 440U and 4400U). Premedication included intravenous (IV) dexamethasone twelve hours in advance, and IV dexamethasone, aspirin 325mg, IV diphenhydramine, and IV famotidine 30 minutes prior to the first bag. He tolerated the desensitization protocol without a reaction and has progressed to monthly infusions with pretreatment without adverse reactions.

**Discussion:** Adverse reactions to imiglucerase are an infrequent setback in Gaucher disease treatment, with mild cutaneous or respiratory symptoms primarily reported resolving with slowed infusion rates. Although hypersensitivity reactions suspected to be IgG-mediated have been described within the first six months of therapy, this case represents an even more rare immediate muscular hypersensitivity reaction upon first imiglucerase exposure with a successful model for desensitization and continued current tolerance of active therapy.

### M038

## MRNA COVID-19 VACCINE-ASSOCIATED BULLOUS FIXED DRUG ERUPTION

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Introduction: Fixed drug eruptions are unique cutaneous drug reactions that recur in the same location upon re-exposure to the offending agent. We report a dilemma of how to proceed with the vaccination after a rare complication of the mRNA covid-19 vaccine. Case Description: A 47-year-old female referred to us after developing a rash to the mRNA covid-19 vaccine for evaluation of safety and plan on how to proceed with the booster vaccine. After the 1st dose on her left deltoid, she developed persistent right arm pruritus from mid-forearm to wrist that gradually became well-demarcated and erythematous by day four. Rash improved within seven days of using hydrocortisone but left hyperpigmentation of the skin. Two weeks after receiving her 2<sup>nd</sup> dose on her left deltoid, the same location on her right forearm flared up with more pronounced pruritus, erythema and a few bullous lesions. Rash improved after a week of using hydrocortisone leaving behind hyperpigmentation. The patient denied using any prior medications and had no other skin or mucosal involvement. We decided to proceed with another brand of mRNA covid-19 vaccine instead and prophylactically apply clobetasol twice daily to the affected area for 7-days post-vaccine. She tolerated the vaccine without any flare-up of the rash.

**Discussion:** Fixed drug eruptions more frequently occur with drugs including antimicrobials, NSAIDs, acetaminophen and anti-convulsants. Very rarely has it been reported in association with the administration of the Covid-19 vaccines. This case illustrates a unique predicament after an unusual complication of bullous fixed drug eruption after mRNA covid-19 vaccine administration.

Figure. Bullous fixed drug eruption.



Erythema extending frommid-forearm to wrist with few bullous lesions.

### M039

### SECONDARY URTICARIAL VASCULITIS AND CHRONIC SPONTANEOUS URTICARIA ASSOCIATED WITH COVID-19 VACCINATION

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**Introduction:** COVID-19 vaccines are generally safe, however there are associated adverse reactions including delayed cutaneous manifestations. This is a case of secondary urticarial vasculitis and chronic spontaneous urticaria associated with COVID-19 vaccination.

Case Description: A 29-year-old male with allergic rhinitis and dermatographism presented with recurrent urticaria. He received his first dose of COVID-19 vaccine and started to have pruritic urticaria on his abdomen after 5 days. Punch biopsy of the lesion showed dermal hypersensitivity response and findings consistent with secondary urticarial vasculitis. His rash improved with topical steroids and oral antihistamines. After the second vaccine, he immediately developed similar symptoms on his face and body. Rheumatologic and infectious work up were unremarkable, and the concern for systemic vasculitis was low. Secondary urticarial vasculitis and hypersensitivity response related to the vaccine was high on the differential given the temporal association. Due to his recurrent pruritic urticaria refractory to high dose of oral antihistamines, omalizumab was started with significant improvement. He is now doing well off omalizumab and on as needed antihistamines only.

**Discussion:** There are reports of delayed cutaneous reactions associated with COVID-19 vaccines. Although rare, it is important to recognize these manifestations of urticarial vasculitis and chronic spontaneous urticaria associated with COVID-19 vaccination.

### M040

### SYMMETRIC DRUG-RELATED INTERTRIGINOUS AND FLEXURAL EXANTHEMA (SDRIFE) INDUCED BY CIPROFLOXACIN

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**Introduction:** Symmetric Drug-Related Intertriginous and Flexural Exanthema (SDRIFE) describes a symmetrical erythematous rash

seen on gluteal and intertriginous areas after first or repeated exposure to systemic drugs. Although beta-lactam antibiotics are the most common causative agents, fluoroquinolones and topical medications have only rarely been implicated in SDRIFE. Here we describe a case of SDRIFE to ciprofloxacin.

**Case Description:** A 19-year-old male with microtia was treated for otitis externa. He had no prior history of reactions to antibiotics. He received one week of ciprofloxacin otic drops followed by the addition of oral amoxicillin and ciprofloxacin. After one dose of oral ciprofloxacin, he developed pruritus on his neck, chest, and ear and was subsequently admitted for intravenous (IV) sulfamethoxazole-trimethoprim and ciprofloxacin along with otic ciprofloxacin due to worsening otitis. After 2 doses of IV antibiotics, he developed a maculopapular pruritic rash over the decolletage and back with symmetrical eczematous plaques to bilateral axillae, posterior auricular folds, and neck. Despite discontinuing IV antibiotics, the rash initially worsened but rapidly improved after stopping otic ciprofloxacin.

**Discussion:** We report one of the few cases of flouroquinolone induced SDRIFE previously described only in patients receiving systemic therapy. In this case, exposure to topical fluoroquinolones continued to drive the hypersensitivity reaction despite discontinuing all systemic fluoroquinolones. Although fluoroquinolones and topically administered drugs remain less recognized culprits of SDRIFE, this case highlights the importance of avoidance of all forms of a culprit drug in patients with drug hypersensitivity.

**Figure.** Symmetric Drug-Related Intertriginous and Flexural Exanthema (SDRIFE) Reaction.



Erythematous Plagues on Bilateral Axillae.

### M041

### A CASE OF TOXIC EPIDERMAL NECROLYSIS DURING IMMUNE CHECKPOINT INHIBITOR AND PLATIN TREATMENT FOR CANCER

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**Introduction:** Immune checkpoint inhibitors have revolutionized cancer therapy but have unforeseen side effects. We present a case of metastatic gastroesophageal cancer treated with nivolumab and FOLFOX causing toxic epidermal necrolysis (TEN). This case highlights the importance of balancing cancer treatment with screening and management of adverse events.

**Case Description:** A 57-year-old male with stage IV gastroesophageal adenocarcinoma was started on nivolumab and FOLFOX every two weeks. He reported a mild rash after the first 3 cycles that resolved spontaneously. After the fourth cycle, he presented to the ED with a painful, erythematous, pruritic, full-body rash from the neck down with blistering and desquamation around the fingers and toes. He received intravenous solumedrol, famotidine, and diphenhydramine and was discharged on a prednisone taper and topical triamcinolone. Labs were notable for a mildly elevated CRP of 6.57 and lactate of 2.6. Skin biopsy showed epidermal necrosis consistent with TEN, erythema multiforme (EM), or EM-like drug eruption. When prednisone was tapered down, the rash worsened requiring hospitalization and high-dose steroid treatment. Nivolumab was halted, but another cycle of FOLFOX was administered. The rash worsened again two weeks later requiring additional steroids. FOLFOX was subsequently discontinued and capecitabine was