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AN INFANT WITH HEREDITARY ALPHA TRYPTASEMIA AND ELEVATED INTESTINAL MAST CELLS IMPROVES WITH CYPROHEPTADINE TREATMENT

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Introduction: Hereditary alpha tryptasemia (HaT) causes symptoms in multiple organ systems, including gastrointestinally. We present a patient with HaT who had chronic reflux and diarrhea that resolved with cyproheptadine treatment. Histology demonstrated increased numbers of mast cells within the duodenum and colon, further supporting the role of HaT in the pathophysiology of his symptoms.

Case Description: A 10-month-old boy with a history of premature birth at 33 weeks presented to allergy clinic for two episodes of intractable vomiting and lethargy after consuming oats. His parents also reported a history of severe reflux refractory to famotidine and intermittent loose stools. The patient’s mother and two siblings have HaT confirmed via genetic testing. Laboratory evaluation revealed undetectable IgE to oat and elevated tryptase of 15.3 ng/mL, lending diagnoses of oat FPIES and likely HaT. Trials of cetirizine and omeprazole did not alleviate his symptoms. Workup for infectious or malabsorptive causes was unrevealing. He underwent esophagogastroduodenoscopy with biopsy, which showed normal architecture of his stomach and intestinal mucosa. Immunostaining for CD117 (c-kit) and tryptase highlighted greater than 63 and 77 mast cells per high power field (HPF) within the duodenum and colon respectively. He was initiated on cyproheptadine 1 mg daily, as this had helped his sibling’s similar symptoms, and had complete resolution of his reflux and loose stools.

Discussion: Increased numbers of mast cells in the intestinal mucosa may contribute to gastrointestinal symptoms in HaT, a finding not previously reported in pediatric literature. Cyproheptadine may be a useful treatment for patients with HaT who experience significant gastrointestinal symptoms.

Immunohistochemical staining for CD117 (c-kit) highlights an increased number of mast cells in the duodenum (40x magnification).

Increased number of intestinal mucosal mast cells have been reported in adult patients with HaT and patients with inflammatory bowel disease. Our study is the first to our knowledge to identify increased intestinal mast cells in a pediatric patient with HaT.

HYPERSENSITIVITY TO HUMAN SEMINAL PLASMA IN A TRANSGENDER MALE

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Introduction: Human seminal plasma (HSP) hypersensitivity is a rare allergy to human semen proteins that can present in female patients as local or systemic reactions. Here, we report a case of HSP hypersensitivity in a transgender male.

Case Description: A 21-year-old transgender male (female-to-male) with allergic rhinitis presented with a 5-year history of recurrent vaginal pain after coitus. Vaginal intercourse causes a severe burning sensation for 3-4 hours and is sometimes associated with itching that lasts for days. It is prevented by the use of condoms. Skin contact with semen immediately causes pruritic lesions for 1-2 hours. Receptive oral intercourse causes lip tingling. He has developed abdominal pain and loose bowel movements after swallowing his partner’s semen. No associated respiratory or cardiovascular symptoms or angioedema reported. Patient denied using lubricants or spermicides that may cause other allergic reactions. Vulvovaginitis was ruled out with normal pelvic exam per gynecology. For management, prevention of seminal fluid exposure by using condoms was recommended. Also recommended were antihistamine and prophylactic use of cromolyn.

Discussion: The diagnosis of HSP hypersensitivity is made based on a history of symptom development upon HSP exposure and prevention of symptoms with the condom use. No skin testing was performed because localized reaction to HSP is considered non-IgE mediated and no standardized testing is available. Intra-vaginal/subcutaneous desensitization is not very effective for local reactions.

SARS-COV-2 INFECTION ASSOCIATED WITH NEW SEVERE HYPEREOSINOPHILIA AND MULTIPLE ORGAN DAMAGE

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Introduction: SARS-CoV-2 infection leads to a wide spectrum of clinical disease characterized by hyperinflammation. The role of eosinophils in SARS-CoV-2 infection is unclear, and rare eosinophilic complications have been reported. We present a case of severe hypereosinophilia developing concomitantly with SARS-CoV-2 infection, leading to multiple end-organ damage to the extent not previously described.

Case Description: A 24-year-old female with bipolar disorder presented with neck/throat and muscle pain in January of 2021, during an Omicron variant wave of SARS-CoV-2. Initial testing detected positive SARS-CoV-2 PCR, leukocytosis $37.7 \times 10^{9}/\mu L$ and eosinophilia $24.39 \times 10^{9}/\mu L$. Over the next month, patient’s leukocytes and eosinophils persistently increased to maximum 96.5 and 71.5 $\times 10^{9}/\mu L$ respectively. She developed muscle weakness and vision changes. Physical exam showed forearm edema, tremors, and splinter hemorrhages. Imaging confirmed myocarditis, multiple strokes, with regional myositis, cellulitis, and tenosynovitis. Lab workup was negative for other infections, ANCA vasculitides, KIT-mutation, tryptase, vitamin B12, FISH panel for myeloproliferative disorders, and next generation sequencing (PDGFRA, PDGFRB, FGFR1, FGFR2, JAK2). Bone marrow biopsy demonstrated leukocytosis, absolute eosinophilia with reactive features. She was treated with hydroxyurea and systemic steroids, after which the eosinophil count declined and she started to note clinical improvement. At this time, she has successfully transitioned to mepolizumab monotherapy.

Discussion: Hypereosinophilia and eosinophilic inflammation have been described following SARS-CoV-2 infection and vaccination. Hypereosinophilia can inflict end-organ damage, but to our
knowledge this is the first reported case of severe hypereosinophilia coinciding with COVID-19, causing this extensive degree of systemic thrombotic and myopathic disease. Whether SARS-CoV-2 infection was a coincidental or causative, remains uncertain.

Multiple sites of end organ damage in the setting of severe hypereosinophilia

M366
STERIOD SPARING STRATEGY WITH DUAL BIOLOGICS IN TREATING HYPEREOSINOPHILIC SYNDROME
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Introduction: Hypereosinophilic Syndrome (HES) is a multisystem disorder characterized by peripheral blood eosinophilia and organ impairment due to eosinophil infiltration. Treatment primarily consists of reducing eosinophil levels via a variety of modalities, including corticosteroids, which carry significant side effects. Different strategies have been developed to reduce the dosage of chronic steroids that a patient receives.

Case Description: A 62 year-old female patient has been followed in our Allergy and Immunology clinic for 17 years for her Lymphocytic hypereosinophilic syndrome. Her treatment has been changed throughout the years to control symptoms and eosinophil level. For the past few years, she has been stabilized on a regimen of mepolizumab 300 mg subcutaneous q6 weeks, daily cyclosporine, as well as prednisone 7.5 mg daily. Prednisone tapering has been attempted multiple times in the past without success. In March of 2022, dupilumab was added after insurance approval. Dupilumab, which is FDA approved for eosinophilic esophagitis as well as eosinophilic asthma, was chosen as additional therapy blocking the IL-4/IL-13 pathway, unique to mepolizumab as well as eosinophilic asthma, was chosen as additional therapy blocking the IL-4/IL-13 pathway, unique to mepolizumab. She was started on a 600mg loading dose and 300mg q2weeks. Within 4 weeks, her prednisone was successfully tapered to 5mg, the lowest dose in the last 17 years, and further taper is in process.

Discussion: Dual biologic therapy with both dupilumab and mepolizumab is a potential steroid sparing strategy for patients with refractory hypereosinophilic syndrome. Further studies to validate the efficacy and identify ideal dosing combination of such dual biologic therapy in sparing steroids is indicated.

M367
PERSISTENTLY ELEVATED BASELINE TRYPTASE - A CASE OF HEREDITARY ALPHA TRYPTASEMIA
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Introduction: Hereditary Alpha Tryptasemia (HaT) is an autosomal dominant genetic condition resulting from inheriting an extra TPSAB1 gene copy, which encodes alpha and beta tryptase. Most cases involve increased copy numbers of alpha tryptase gene, but documented cases of beta tryptase gene duplications exist. HaT typically presents with tryptase levels greater than 8 ng/mL. Phenotypes vary in severity and history may include idiopathic anaphylaxis, vibratory urticaria, musculoskeletal problems, and dysautonomia. HaT can coexist with clonal mast cell diseases, particularly systemic mastocytosis (SM).

Case Description: A 15-year-old female presented for elevated blood tryptase levels. She reported frequent emesis, diarrhea, anorexia and unintentional weight loss. She reported generalized fatigue, anxiety, and musculoskeletal pain with 2-years of frequent flushing and postural orthostatic hypotension. She denied dysphagia, urticaria, and anaphylaxis. Lab evaluation revealed negative c-KIT mutation. Tryptase range was 12.2-14.2 ng/mL. TPSAB1 gene analysis was obtained demonstrating two copies of alpha-tryptase and three copies of beta-tryptase. She was diagnosed with HaT based on a duplication at TPSAB1 allele. Initiation of cromolyn and high dose cetirizine resulted in suboptimal symptomatic improvement. Management is currently ongoing.

Discussion: When assessing elevated tryptase levels, considering HaT in the differential diagnosis is important, in addition to possible confounding conditions. A diagnosis of HaT also does not preclude concomitant conditions such as SM. This case demonstrates the need for more inclusive nomenclature to account for beta-tryptase duplications sometimes presenting with similar clinical manifestations. We propose HaT be renamed Hereditary Tryptasemia, accounting for cases of additional beta-tryptase copies as demonstrated in this patient.

M368
SYSTEMIC MASTOCYTOSIS PRESENTING WITH KOUNIS SYNDROME AND REFRACTORY SHOCK
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Introduction: Systemic mastocytosis (SM) can rarely present in the setting of refractory shock and/or coronary artery vasospasm triggered by mast cell activation, also known as Kounis Syndrome.

Case Description: A 30-year-old male with a history of urticaria pigmentosa (lost to follow up as an adolescent) and unexplained syncopal episodes was found unconscious at home. Upon EMS arrival, he