SEVERE EOSINOPHILIC CYSTITIS CONTROLLED WITH BENRALIZUMAB
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Introduction: Eosinophilic Cystitis (EoC) is a rare disorder of the bladder resulting in lower abdominal pain, hematuria and dysuria. Standard treatment of antihistamines, leukotriene antagonists, NSAID’s, cyclosporine, hydroxyurea and intravesicular corticosteroids is not always effective. Benralizumab is a monoclonal antibody directed against the IL-5 receptor, reducing eosinophil production and survival. Unlike other antagonists of IL-5, it is very effective in tissue depletion of eosinophils. It has previously been shown to be effective in EoC.

Case Description: AG is a 16 yo female who presented with excruciating lower abdominal pain and passing blood clots in her urine. Evaluation documented eosinophils in the bladder. Patient failed treatment with cetirizine, montelukast, NSAID’s, so was put on oral prednisone then intravesicular corticosteroids, when sent for extra

Discussion: Because benralizumab is effective in depleting tissue eosinophils, it may be more effective than standard therapy for treatment of recalcitrant EoC. Since benralizumab has orphan drug status for HES, and because she failed previous therapy, including high dose mepolizumab, her insurance company agreed to pay to continue her on this therapy.

A CASE OF ELEVATED TRYPotate
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Introduction: We present a case of hereditary alpha tryptasemia (HαT) which can have a broad spectrum of presenting symptoms and should be considered when serum basal tryptase > 8 mcg/L.

Case Description: A 42 year-old female with progressive decline in cognition and memory, fibromyalgia, hypermobility and swelling of joints, chronic abdominal pain, and recurrent infections presented for evaluation of elevated tryptase. She had tryptase levels ranging from 14-16 mcg/L, in addition to two reported episodes of idiopathic anaphylaxis.

Discussion: Physical exam was most remarkable for delayed response to commands with slow speech and impaired recall. No abnormalities found with CBC, CMP, thyroid studies, urine mast cell mediators, or immune evaluation including immunoglobulins and vaccine responses. ESR, CRP, ANA, and extensive autoimmune work-up including antibodies for Lupus, Rheumatoid, Sjogren’s, and myositides were negative. MRI yielded brain atrophy of unclear etiology. Tryptase genetic testing was positive for extra α tryptase copy, being diagnostic for HαT.

High dose antihistamines and mast cell stabilizers were ineffective in treatment so we opted for experimental therapy with dasatinib, a tyrosine kinase inhibitor which has been shown to decrease mast cell number and lower serum tryptase. She recently started on this treatment with reported improvement in multi-system complaints and we will continue to monitor progress in months to come.

Discussion: It is unclear how elevated basal serum tryptase levels translate to the multi-system disorder we may observe in HαT patients. Our case demonstrates a situation of elevated tryptase where HαT testing was performed and helped add to known phenotypes associated with the disease.

RELAPSED LEUKEMIA PRESENTING AS SM-AHN AND HIP PAIN
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Introduction: Systemic mastocytosis (SM) is a rare condition characterized by mast cell infiltration of extracutaneous organs, which can cause allergic symptoms including anaphylaxis. More rarely still, SM can be associated with a hematologic neoplasm (SM-AHN). Allergy was consulted for treatment options in a hospitalized patient with SM presenting with severe hip pain.

Case Description: A 57-year-old male with history of acute myelomonocytic leukemia (AMML) status-post allogeneic stem cell transplantation presented with acute atraumatic left hip pain. Given concern for AMML relapse, he underwent bone marrow biopsy, which showed no leukemic cells but did show over 25% mast cells with spindling and CD25 positivity. Additional finding of persistently elevated serum tryptase diagnosed SM. He did not improve with oral antihistamines, mast cell stabilizer, or midostaurin. After biopsy of a new skin lesion showed leukemia cutis, repeat bone marrow biopsy confirmed AMML recurrence. With initiation of cytoreductive chemotherapy the patient had significant improvement in hip pain and normalization of serum tryptase, suggesting SM-AHN as the most correct explanation for previous findings.

Discussion: SM-AHN is an uncommon form of a rare disorder. It typically presents with a myeloproliferative neoplasm or myelodysplastic syndrome, with mastocytosis potentially clouding diagnosis of malignancy. This case is remarkable for its association with AMML and presentation after SCT, which can be used as a treatment for cases of advanced SM. This case reminds us that diagnoses are not always straightforward, and further evaluation is required when clinical course does not follow expected trajectory based on available data.

A CASE OF ISOLATED ENTEROCOLIC MUCOSAL MAST CELL AGGREGATES
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Introduction: Gastrointestinal infiltration by atypical mast cell aggregates is a key feature of systemic mastocytosis (SM). However, not all patients with this finding meet SM criteria. We describe one such patient’s presentation.

Case Description: A 35 year old female with a history of obesity and hypertension presented with abdominal bloating, diarrhea, 40 lb unintentional weight loss and generalized pruritus for 4 years. Physical exam was unremarkable. Tryptase was normal and peripheral blood PCR was negative for c-kit mutation. Bone marrow biopsy revealed <5% tryptase positive mast cells, scattered CD2 positive small T lymphocytes, and extremely rare CD25 positive cells. Cytospin showed an increase in eosinophils. Colonoscopy identified increased eosinophils and multifocal dense infiltrates of atypical spindle-shaped, CD117/ tryptase positive, CD2/CD25 negative mast cells throughout the colon. EGD was normal. The patient was started on cetirizine, montelukast and oral cromolyn sodium, with significant improvement in abdominal bloating and recovery of weight loss within 4 months.

Discussion: There are reports of patients with atypical enterocolic mast cell aggregates without suspected or established SM. In contrast to this large case series, our gastrointestinal biopsy cells were stained negative for CD25. Immunoreactivity for CD25 in GI mucosal mast cells has been reported to be specific for SM. In our patient, symptoms were substantial, resulting in significant weight loss, and responsive to oral cromolyn sodium, systemic antihistamines and a leukotriene inhibitor. Whether this isolated organ abnormality may progress to SM remains to be seen, but providers should be aware of this entity and treatment options.