Eosinophilia in a pediatric patient with orbital and facial swelling

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Rationale

Hypereosinophilic Syndrome (HES) is rarely encountered in the pediatric population. We describe a previously healthy 12-year-old boy with swelling of the face and orbit, and peripheral eosinophilia.

Introduction

A previously healthy 12 year old boy presented to the emergency room with worsening left sided facial edema and jaw pain without fevers, vision changes, congestion or pain with eye movements. He had recently been discharged from an inpatient pediatric unit after receiving treatment with IV vancomycin and ampicillin-sulbactam for concern for infectious etiology of orbital edema and was transitioned to amoxicillin-clavulanic acid without improvement in symptoms.

Methods

This patient underwent thorough evaluation, including MRI of the orbit, face and neck, orbital tissue biopsy and laboratory analyses to investigate etiologies and potential sequelae of hypereosinophilia.

Differential Dx

Differential included an exaggerated response to insect bite, malignancy, fungal, atypical bacterial or parasitic infection, drug hypersensitivity, orbital pseudotumor, eosinophilic granuloma, sarcoidosis, granulomatosis with polyangiitis (EGPA), Kimura Disease or other collagen vascular diseases.

Figure 1* : Left sided perioribital swelling

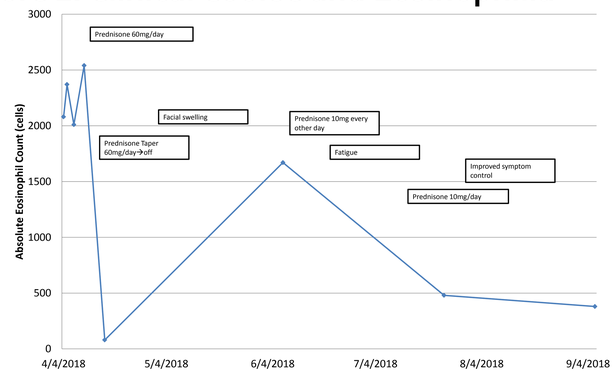
*Photo presented with permission by patient and parent

Results

Table 1: Diagnostic or laboratory testing

	Result
MRI of the orbit, face and neck	Infiltrative left sided orbital/retro maxillary process involving medial/lateral/inferior recti, temporalis muscle and focal erosion of base of left sphenoid bone
Echocardiogram	Normal
Pulmonary Function Tests: Spirometry with FeNO and DLCO	Normal
Troponin (ng/mL)	<0.02
Abdominal ultrasound	Two isoechoic lesions in the right hepatic lobe, largest measuring 9.4cm
MRI of the abdomen	Well-circumscribed, single lobulated lesion in the right hepatic lobe measuring 10.3 x 9.2 x 10 cm with imaging features consistent with a focal nodular hyperplasia
F1P1L/PDGFRA	Negative
TCR clonality	Polyclonal
Serum IGAM (mg/dL) IgA IgG IgM IgE	Normal 58 (46-218) 850 (685-1620) 134 (27-151) 107 (8-631)
B12 level (pg/mL)	306 (310-1379)
Tryptase (ng/mL)	2.0 (1-11.4)
Strongyloides serology (IgG)	Negative
Stool ova and parasite	Negative on two evaluations
ANA	Titer 1:80 (low positive)
Aldolase (unit/mL)	5.8 (3.3-9.7)
Anti-PR3 antibody (AI)	<0.1
Myeloperoxidase antibody (AI)	<0.1
Muramidase (mcg/mL)	6.8 (5-11)
Angiotensin converting enzyme inhibitor (unit/L)	68 (13-100)
QuantiFERON®-Tb Gold	Negative
Biopsy of L orbital/facial soft tissue	Lymphocytic and eosinophilic perivascular and panniculitic infiltration also involving adjacent skeletal muscle but no evidence of clear vasculitis, granuloma, foreign body reaction, myopathic process, tumor and microorganisms on special stains. Stains demonstrated a mixed population of T and B cells, rare IgG4 and CD1a positive cells.

Figure 1: Clinical Course and Eosinophilia



Management

He had no evidence of parasitic infections, hypersensitivity disorders or neoplasm and similarly, laboratory testing and biopsy findings made vasculitis less likely, though perivascular involvement could have represented early disease. This patient was started on prednisone 60mg/day and weaned to 10mg/day with clinical improvement noted. He has been unable to wean off steroids after many months due to recurrence of orbital edema, and so other therapeutic options are being investigated. After consultation with Rheumatology methotrexate was recommended as a steroid sparing agent but not pursued given family reluctance. He is currently being considered for anti-IL5 therapy.

Conclusions

We describe a rare case of a pediatric patient with idiopathic HES versus early eosinophilic vasculitis presenting with facial and orbital swelling. Non-specific clinical and laboratory findings make diagnosis, treatment and prognosis challenging.

References

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