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Orbital angiolymphoid hyperplasia with eosinophilia in a pediatric patient: a case report

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ABSTRACT

A 7-year-old female presented to an outpatient eye clinic with a six-week history of left upper eyelid edema and erythema in the absence of trauma without compromise of visual acuity or extraocular muscle movements and was initially treated as a case of pre-septal cellulitis with antibiotic therapy. Failure of resolution with new onset nasal bleeding after two weeks prompted computed tomography (CT) and magnetic resonance imaging (MRI), showing an orbital mass with radiologic concern for lymphoma or rhabdomyosarcoma. Upon biopsy, the histopathological analysis diagnosed angiolymphoid hyperplasia with eosinophilia (ALHE), a rare condition that infrequently presents with ocular manifestations and more rarely presents in the pediatric population. Most therapeutic management for cutaneous and non-cutaneous ALHE have high rates of recurrence. This case was refractory to oral prednisone treatment but responded to a combination of methotrexate and adalimumab therapy with an 18.5% reduction in volume after three months.

KEY WORDS: ALHE; epithelial hemangioma; images

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INTRODUCTION

Angiolymphoid hyperplasia with eosinophilia (ALHE), also called epithelial hemangioma (EH), is a rare, benign vasoproliferative disorder with uncertain pathophysiology that can occur in orbit but most commonly occurs at cutaneous sites of the face and neck. ALHE was first described by Wells and Whimster in 1969, with a later description by Enzinger and Weiss in 1983, coining EH due to focusing on the pathological atypia of endothelial cells [1, 2]. A similar presentation of ALHE that was once considered synonymous but is now increasingly recognized as a separate condition is Kimura Disease (KD), the distinction made by the absence of the characteristic endothelial atypia and the presence of interstitial fibrosis in Kimura disease [3]. The clinical presentation of ALHE is variable in distribution and the number of lesions.

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Still, it is most commonly reported at cutaneous regions of the ear or scalp, with fewer than one percent occurring within the orbit [4]. Symptoms of the single- or multiple-presenting lesions may include pruritis, pain, or spontaneous bleeding, with only 15% of cutaneous manifestations being asymptomatic [4, 5]. Past literature has reported a female predominance, but a 2016 systematic review of 908 patients indicated no difference in manifestation by sex.4 With a mean age at presentation being reported from 37.6 to 40 years old, pediatric cases are rarely reported in the literature [4, 6]. Diagnosis is commonly made following excisional or FNA biopsy in traditional cutaneous ALHE, but in ocular manifestations, biopsy commonly follows imaging techniques such as CT or MRI [6]. Excision is the most commonly chosen treatment modality (44.2%), while intralesional corticosteroids (11.3%), topical corticosteroids (9.1%), and systemic corticosteroids (6.9%) are also utilized.4 Recurrence rates for all modalities are high at 40.8%, 79.1%, 98.2%, and 87.8%, respectively.4 Given the suboptimal outcomes of standard therapy, there is still no consensus on a therapeutic approach.

CASE PRESENTATION

A 7-year-old female presented to an outpatient ophthalmology clinic in Tallahassee, FL, with a six-week history of left upper eyelid edema, erythema, and mild pruritis (Fig. 1). She had no history of ocular trauma or prior instrumentation of the area or significant medical history. Visual acuity measured 20/20 in each eye with full extraocular movements. Physical exam was significant for 1+ edema and 1+ erythema in the left upper eyelid (LUL). The dilated fundus exam and cycloplegic refraction were normal. She was started on oral amoxicillin-clavulanic acid for 2 weeks for concern of pre-septal cellulitis. Following antibiotic course completion, she returned to the clinic with failure to resolve LUL symptoms and a new onset symptom of intermittent, low volume bright red blood drainage from the left nostril.

Failure of treatment and the ipsilateral nasal epistaxis prompted concern for post-septal infection necessitating a CT orbit and sinus which identified a left orbital lesion enveloping the superior rectus muscle inseparable from the lacrimal gland with extension into the preseptal area. The CT findings prompted a contrast-enhanced MRI of the head and orbits. The lesion was found to be 2.8 cm in width, 0.8 cm in vertical height, and 2.9 cm in AP (Fig. 2). It was concerning for possible lymphoma or rhabdomyosarcoma, which prompted an excisional biopsy. After histopathologic evaluation, a diagnosis was made of angiolymphoid hyperplasia with eosinophilia due to the identification of vascular proliferation lined by endothelial cells with abundant eosinophilic cytoplasm and cytoplasmic vacuoles (Fig. 3). The presence of cytoplasmic changes and the absence of interstitial fibrosis of fibro-collagenous components ruled out the previously synonymous Kimura disease.3,7

Immediate post-diagnosis medical management consisted of 25 mg/day of oral prednisone with no change in lesion size on a one-month repeat MRI. After two total months of prednisone therapy without improvement, the patient was seen by a pediatric rheumatologist and started on 20mg of subcutaneous methotrexate once weekly as well as 40 mg of subcutaneous adalimumab every two weeks. A repeat MRI at three months following the additional medical therapy showed a decrease in size to 2.8 cm in width, 0.7 cm in vertical height, and 2.7 cm in AP, an 18.5% decrease in volume. The patient remains clinically stable without changes in visual acuity or extraocular movements.

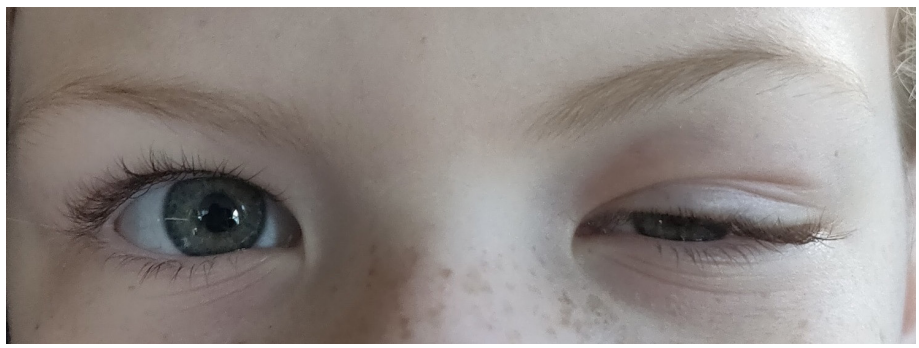

Figure 1. Photograph of patient referenced above on initial presentation demonstrating +1 left upper eyelid edema and erythema

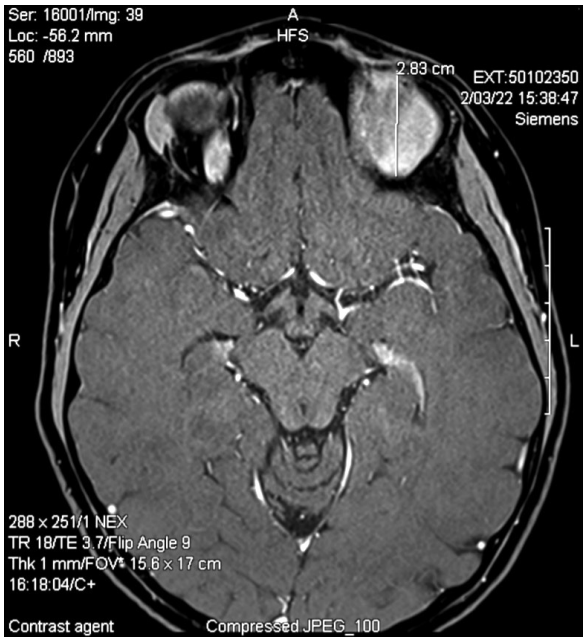

Figure 2. Contrast-enhanced magnetic resonance imaging (MRI) of the head and orbits showing a left orbital lesion, 2.8 cm in width, 0.8 cm in vertical height, and 2.9 cm in anterior-posterior (AP), enveloping the superior rectus muscle inseparable from the lacrimal gland with extension into the preseptal area

Orbital angiolymphoid hyperplasia with eosinophilia is an uncommon condition and only rarely reported in children. As demonstrated in our case, the clinical presentation of orbital ALHE can mimic pre- and post-septal cellulitis. Additionally, on imaging, ALHE can mimic malignancies with a significant mortality rate. Therefore, histopathologic evaluation is essential to the final diagnosis. Additionally, this case report sheds light on the non-surgical or pre-surgical management of ALHE, given the previously mentioned high recurrence rates for many standard therapies. The initial lack of lesion response to systemic prednisone is consistent with most previous literature.2,4 The reported first success of methotrexate for ALHE was by Baker et al. in 2012.8 There are no reported cases of successful adalimumab therapy either as monotherapy or in combination with another medication for orbital ALHE. In our case, the use of concurrent methotrexate and adalimumab resulted in an identified decrease in lesion size of 18.5% after three months of therapy. This may provide additional support for using these agents for ALHE in children.

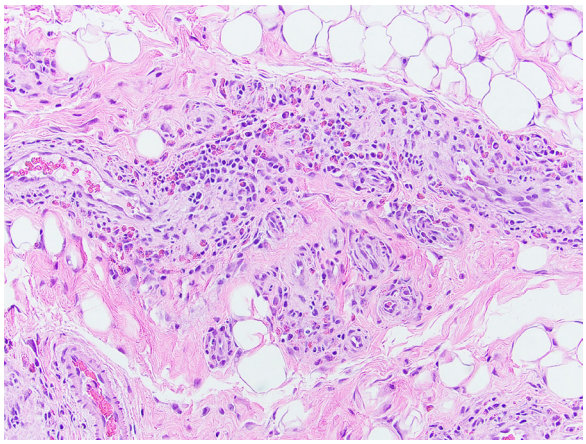

Figure 3. Histopathologic image of the patient's biopsy specimen demonstrating vascular proliferation lined by endothelial cells with abundant eosinophilic cytoplasm and cytoplasmic vacuoles characteristic of angiolymphoid hyperplasia with eosinophilia (ALHE)

Conflict of interest:

The authors have no conflicts of interest to disclose related to this submission.

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