

# Angiolymphoid Hyperplasia with Eosinophilia of Orbit in Young Male

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## ABSTRACT

Angiolymphoid hyperplasia with eosinophilia (ALHE) is an uncommon benign clinical entity characterized by the presence of a variable number of papules, plaques or nodules of the dermis and subcutaneous tissues. ALHE shows a predilection for the head and neck area. Orbital involvement is unusual. A 20-year-old male presented with a 4-month history of swelling of the upper lid of right eye. Magnetic resonance imaging orbit revealed altered signal density seen in supero-medial aspect of right eyeball with adjacent inflammatory changes. The patient underwent anterior orbitotomy and excision biopsy of the mass under general anesthesia. Histopathology of the mass showed vascular proliferation and chronic inflammation accompanied by infiltration of eosinophils, which confirmed the diagnosis of ALHE. The patient recovered both functionally and cosmetically and had no signs of recurrence after 6 months. Angiolymphoid hyperplasia with eosinophilia with orbital involvement in males is a rare clinical entity, and further work is required to accurately describe its incidence, etiology and presentation. ALHE can be diagnosed and differentiated from Kimura's disease (KD) on histopathological grounds. The presence of vascular hyperplasia with plump of endothelial cells protruding into the lumen is the most important discriminator in establishing the diagnosis of ALHE. Such distinction is crucial for the patient because ALHE is not associated with any of the systemic manifestations present in KD.

**Keywords:** Angiolymphoid hyperplasia, eosinophilia, orbit

## INTRODUCTION

Angiolymphoid hyperplasia with eosinophilia (ALHE) is an uncommon benign clinical entity characterized by the presence of a variable number of papules, plaques or nodules of the dermis and subcutaneous tissues.<sup>[1]</sup> ALHE shows a predilection for the head and neck area.<sup>[2]</sup> Orbital involvement is unusual. ALHE is a benign neoplasm of the vascular endothelium. Recently, ALHE is being called epithelioid hemangioma, a term that better describes the most distinguishing feature of this entity: The abnormal proliferation of endothelial cells.<sup>[3]</sup> The etiology of

ALHE is obscure; however, it has been reported to occur in pregnancy and also following trauma in some cases.<sup>[2]</sup> ALHE and Kimura's disease (KD) share many clinical and histopathological features.<sup>[4]</sup> Although they were once considered different stages of the same disease, they are now known to represent separate entities.<sup>[5]</sup> Histopathological features of ALHE and KD are similar, with both having dermal involvement with vascular proliferation and chronic inflammation accompanied by infiltration of eosinophils. However, it is possible to distinguish the two conditions by determination of endothelial cell morphology. This report presents a patient with orbital involvement of ALHE.

## CASE REPORT

A 20-year-old male presented with a 4-month history of swelling of the right upper lid [Figure 1]. On examination, a well-defined, soft lesion in the supero-medial part of right upper eyelid was palpated, just below the superior orbital rim, with mild inflammatory signs. It

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measured approximately 2 cm × 2 cm and was round to oval in shape. Extraocular movements were normal. Ptosis was present due to lid edema and presence of mass. Examination of anterior and posterior segment of both eyes revealed no abnormality. Both eyes had visual acuity 6/6 with intraocular pressure 17 mm Hg on applanation tonometry. Systemic examination was normal. There was no preauricular or submandibular lymphadenopathy on examination. Renal function tests and rest of the blood counts were normal. Blood eosinophil count was normal (5%). Magnetic resonance imaging contrast of the orbit revealed 9.2 mm × 6.1 mm × 6 mm sized area of altered signal density seen in supero-medial aspect of right eyeball supero-medial to superior oblique and medial rectus muscles with adjacent inflammatory changes [Figure 2]. There was no bony erosion.

Anterior orbitotomy was planned to remove the mass under general anaesthesia [Figure 3]. A 3 cm incision was taken superomedially near the orbital margin, the

mass was identified, and the fibrovascular attachments were dissected. An irregular, greyish white firm tissue mass was excised. Size of the lesion was more on excision due to the deep location of the mass, which was not palpable on examination. Intraoperatively drain was put. Histopathological examination revealed ALHE and there was no evidence of any malignant cells [Figure 4]. The patient had an uneventful postoperative recovery with no complaints. The patient is undergoing regular follow-up for 6 months, and there has been no evidence of recurrence of the condition.

## DISCUSSION

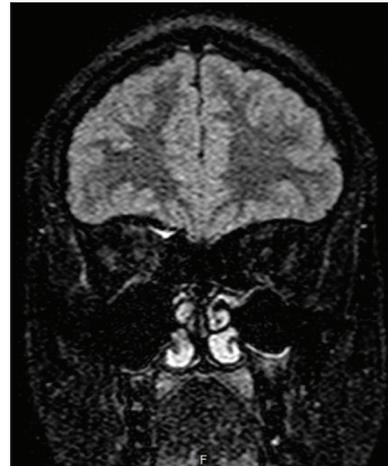
Wells and Whimster were the first to describe ALHE in 1969.<sup>[6]</sup> It has been reported from many parts of the world and can occur in all races. This condition is characterized by abnormal proliferation of endothelial cells with infiltrates of lymphocytes



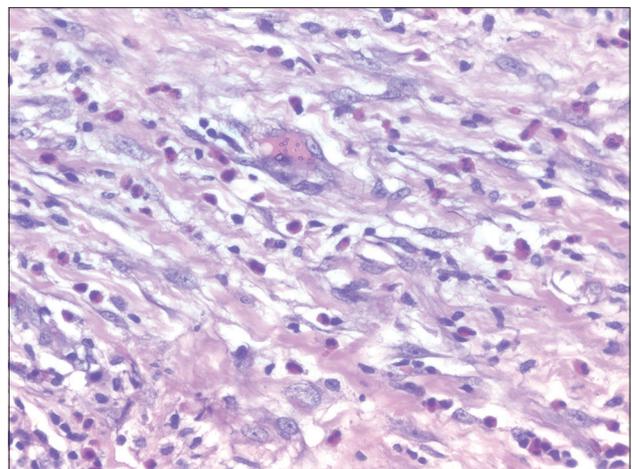
**Figure 1:** Preoperative clinical photograph showing lid edema with ptosis



**Figure 3:** Anterior orbitotomy approach during surgery



**Figure 2:** Magnetic resonance imaging contrast orbit showing alter signal density in T1 section



**Figure 4:** The capillaries are lined by plump endothelial cells with a marked histiocytoid appearance. There is a pronounced cellular infiltrate (arrow) containing many eosinophils (H and E, x400)

and eosinophils, as demonstrated by this case. ALHE is also known as eosinophilic lymphofolliculitis or histiocytoid hemangioma.<sup>[7,3]</sup> ALHE should be distinguished from a variety of benign and malignant vascular proliferations including pyogenic granuloma, epithelioid hemangioendothelioma and Kaposi's sarcoma – all of which lack a noticeable eosinophil infiltrate. The disease usually occurs in middle-aged women, presenting as a subcutaneous nodule, papule, or red plaque. These lesions have been reported to occur over the site of arteriovenous malformation. In the orbital cavity, this condition may present as conjunctival or lacrimal gland tumor.<sup>[8]</sup> Regional adenopathy is rare in this disease, despite its similarities to KD.<sup>[8]</sup> Bleeding may occur but are usually uncomplicated.<sup>[9]</sup> The clinical course ranges from self-limiting to chronic disease, although it usually regresses over a period of 1–4 years and malignant transformation does not occur. ALHE usually presents as benign nodules with erythematous papules mostly in the head and neck region. However, the orbital involvement seen, in this case, is very unusual. Features of orbital involvement may include proptosis, watering, pruritus around the eyes and blurring of peripheral vision.<sup>[9]</sup> Our patient was successfully treated surgically before he developed any such symptoms.

Surgical excision may be beneficial in the case of a solitary small tumor but there is a risk of recurrence at the surgical site.<sup>[4]</sup> Systemic and intralesional steroid administration, interferon therapy, cryotherapy, laser therapy and topical application of tacrolimus have been used with success.<sup>[4]</sup> In conclusion, ALHE with orbital involvement in males is a rare clinical entity, and further work is required to accurately describe its incidence, etiology and presentation. ALHE can be diagnosed and differentiated from KD on histopathological grounds. The presence of vascular hyperplasia with plump of endothelial cells protruding into the lumen is the most important discriminator in establishing the diagnosis of ALHE. Such distinction is crucial for the patient because ALHE is not associated with any of the systemic manifestations present in KD. The former is a localized hyperplasia of atypical endothelial cells with no systemic involvement. On the other hand, the latter can course with lymphadenopathy, blood eosinophilia, and nephrotic syndrome due to IgE deposition in the renal glomeruli.

## CONCLUSION

Angiolymphoid hyperplasia with eosinophilia with orbital involvement in males is a rare clinical entity, and further work is required to accurately describe its incidence, etiology and presentation. ALHE can be diagnosed and differentiated from KD on histopathological grounds. The presence of vascular hyperplasia with plump of endothelial cells protruding into the lumen is the most important discriminator in establishing the diagnosis of ALHE. Such distinction is crucial for the patient because ALHE is not associated with any of the systemic manifestations present in KD. The former is a localized hyperplasia of atypical endothelial cells with no systemic involvement. On the other hand, the latter can course with lymphadenopathy, blood eosinophilia, and nephrotic syndrome due to IgE deposition in the renal glomeruli.

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