

Angiolymphoid hyperplasia with eosinophilia- A rare case report.

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Abstract:

Introduction: Angiolymphoid hyperplasia with eosinophilia (ALHE) is a rare benign vascular tumor affecting primarily the head and neck region of young females. Microscopic analysis reveals hyperplastic vessels lined by hypertrophic endothelium. An inflammatory infiltrate rich in eosinophils is also present. Etiology of the lesion is unknown. Various treatment modalities have been described. We present a case successfully treated by excision followed by steroid infiltration. **Case History:** A 24 year old female presented at Out Patient Department of Dermatology-Civil Hospital, Ahmedabad with complaint of papulo-nodular lesion over right ear, which was gradually increasing in size since 2 months. The lesion was 2cm X 1.2 cm in size, non-tender, non-itchy, and associated with bilateral cervical lymphnode enlargement. **Investigation & Diagnosis:** A punch biopsy was taken from the lesion and sent for histopathological evaluation. Hematoxyline& Eosin method was used for staining. Histopathological examination showed aggregates of lymphoid tissue and numerous proliferated blood vessels with prominent endothelial cells and eosinophils in background. Overall findings were in favour of ALHE. **Conclusion:** ALHE is rare condition with a challenging diagnosis and treatment. In spite of benignity of the disease, it causes therapeutic dilemma because of cosmetic defects and frequent resistance to treatment.

Keywords: Angiolymphoid hyperplasia with eosinophilia (ALHE), Epithelioid Haemangioma, Histiocytoid Haemangioma, Kimura's Disease

Introduction:

Angiolymphoid hyperplasia with eosinophilia (ALHE, Epithelioid Hemangioma, Inflammatory angiomatous nodule, Histiocytoid Hemangioma) is an uncommon, benign, reactive vaso-proliferative disease, presenting with painless vascular nodules in the dermal and subcutaneous tissues of the head and neck, particularly around the ear¹. ALHE has also been reported in the scalp, lip, tongue, orbits and the conjunctiva¹⁻⁴. Although frequency is unknown, cases have been reported worldwide. Although it may be more common in Japan than in other countries. ALHE can persist for years, but serious complications (e.g., malignant transformation) do not occur and have never been reported. ALHE is seen most commonly in Asian, followed by Caucasian. It is rare in elderly patients and in the non-Asian paediatric population. ALHE is somewhat more common in females⁵. It presents most commonly in patients aged 20 - 40 years⁵. ALHE is characterized clinically by single to multiple red brown dome shaped papules or subcutaneous nodules¹⁻⁴. About



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1/5 of patients have blood eosinophilia and Lymphadenopathy².

Case Report:

A 24 year old female presented at Civil Hospital, Ahmedabad with complaint of skin lesion over right ear. **[Image 1]** The lesion was 2 cm X 1.2 cm in size, gradually increasing in size since 2 months. There was no history of pain or tenderness. The skin over other parts of her body did not show any abnormality. She did not give a positive history for fever, skin rashes, asthma and respiratory infection, or any history of louse infestation of the scalp since childhood. On clinical examination the right ear showed a papulo nodular skin lesion. Bilateral cervical lymph node enlargement was noted. There was no evidence of hepatosplenomegaly.

Image 1:- Lesion over Right Ear



Routine blood and urine investigations were within normal limits. No blood eosinophilia was noted. Chest screening showed no abnormality.

A 4mm punch biopsy was taken from the lesion and sent for histopathological evaluation. Routine H&E method was used for staining. **[Image 2]**

Image 2 ALHE – H&E Stain [10 X]

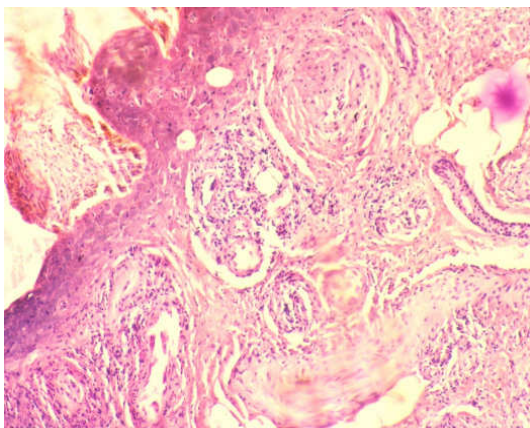
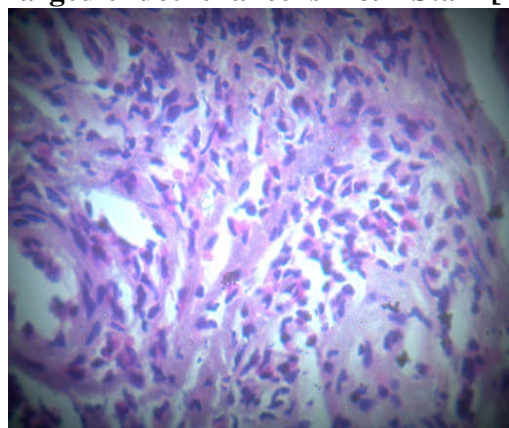


Image 3 ALHE-Aggregate of lymphoid tissue & Enlarged endothelial cells H&E Stain [40X]



Gross examination:

One skin covered greyish brown soft tissue structure measuring 0.4cm X 0.4cm was received.

Microscopic examination:

Section showed mild hypertrophy of epidermis with acanthosis & downward proliferation of rete ridges. In the superficial dermis there was a lobular proliferation of small to medium sized blood vessels lined by *enlarged & prominent endothelial cells*. [Image 3] There was marked perivascular infiltration of chronic inflammatory cells mainly lymphocytes, plasma cells & eosinophils. In the deep dermis there was increased fibro collagenous tissue with *chronic inflammation & eosinophilia*. Overall histology with clinical findings were suggestive of epithelioid haemangioma/angiolympoid hyperplasia with eosinophilia.

The patient was then treated by excision of the lesion followed by local infiltration of steroids after one month. After 3 months of follow up, there is no recurrence.

Discussion:

Angiolympoid hyperplasia with eosinophilia (ALHE) is a rare condition affecting muscular arteries, typically of the head and neck¹. It was first described in 1969 by Wells and Whimster⁴. Initially it was thought to be related to Kimura's disease but recent histological studies indicate that Kimura disease differs from ALHE in several clinical and histopathologic characteristics^{2,6-8}. [Table 1]

Table 1: Kimura's Disease V/S ALHE

Clinicopathological features	Kimura's Disease	ALHE
Age of onset	Younger age	Elder age
Duration of disease	Longer	Shorter
Clinical appearance	Deep seated large soft tissue mass	Single to multiple papulo-nodular eruptions
Overlying skin	Normal	Normal/Ulcerated
Blood Eosinophilia	++	+/-
Lymphadenopathy	+/-	+/-
Elevated serum Ig E	+/-	-
Blood vessels	Thin walled	Thick walled and concentric
Plump endothelial cells	-	++
Lymphoid follicles with active germinal centres	+	+/-
Fibrosis	+	-

Histologically the lesions are characterized by a reactive proliferation of small blood vessels, often surrounding a muscular artery or vein, with peripheral inflammatory infiltrates consisting of mononuclear cells and eosinophils. The endothelial lining of blood vessel undergoes epithelioid changes, leading to the terms "histiocytoid" or, more recently "epithelioid" haemangioma⁶.

The pathogenesis of ALHE remains unclear. Some authors consider ALHE as a neoplasm developing from endothelial cells; others suggest that it is secondary to an inflammatory vascular reaction secondary to complex immunologic mechanisms. Many other hypotheses have been reported implicating environmental factors such as insect bite, trauma,

and infections. Some authors consider that arterio-venous shunt is the main etiopathogenetic mechanism observed in 42 % of the cases⁷.

Serum hypereosinophilia is inconstant (21%) and is not required to make the diagnosis. Positive diagnosis is based upon histological findings. Other differential diagnosis based on clinical and/or histopathologic findings includes.

1. Insect bites- can be ruled out from the history.
2. Capillary hemangioma- histopathological findings suggesting proliferation of numerous thin walled capillaries with scanty connective tissue.
3. Granuloma pyogenicum with satellite lesions- rapidly growing, polypoidal mass, histopathologically a central branching vessel usually devoid of red blood cells, surrounded by a hypercellular proliferation of neofomed endothelial cells and perithelial cells.
4. Angiosarcoma of the face and scalp, Kaposi's sarcoma- histopathological findings consistent with malignancy.

In the absence of treatment, lesions may either increase progressively or decrease spontaneously. Surgical treatment remains the treatment of choice. Recurrences, essentially after incomplete excision, are observed in 30% of the cases⁷. No metastatic cases have been reported⁷. Considering the possible spontaneous involution of the lesions, a simple follow up is recommended 3 to 6 months before surgical excision or other extensive therapeutic modalities are attempted.

Conclusion:

ALHE is rare condition with a challenging diagnosis and treatment. In spite of benignity of the disease, it causes therapeutic dilemma because of cosmetic defects and frequent resistance to treatment.

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