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Angiolymphoid Hyperplasia with Eosinophilia- Think Twice with Scalp Papules!-A Case Report

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Abstract

Angiolymphoid hyperplasia is an uncommon condition characterised by multiple erythematous papules and nodules which are vascular present over head and neck area, predominantly around ears. It is a benign condition but treatment is often sought for the appearance of lesions. We report a case which had recurrence of erythematous lesions after incomplete electrocoagulation and the diagnosis of angiolymphoid hyperplasia with eosinophilia was confirmed on histopathological evaluation after complete excision of the lesion. In conclusion, as angiolymphoid hyperplasia can be clinically misdiagnosed, histopathological evaluation is warranted in erythematous nodules presenting over the sites of predilection.

Key words: Angiolymphoid Hyperplasia, Eosinophilia, Nodules, Papules, Scalp

Introduction

Angiolymphoid hyperplasia with eosinophilia (ALHE), is an uncommon, benign vascular tumor with uncertain etiology. The term was coined in 1969 by Wells and Whimster for describing a distinct type of neoplasm with localised proliferation of blood vessels in a background of dense inflammatory infiltrate consisting of lymphocytes, eosinophils and mast cells.[1] Since then, it has been known by many other names also like epitheloid hemangiomas (EH), pseudopyogenic granuloma, inflammatory angiomatous nodule, histiocytoid hemangioma and atypical granuloma, but ALHE and EH remain the most commonly used nomenclature and their persistence highlights the debated etiology. It usually affects young to middle aged females with head and neck being the sites of predilection. We present a case of ALHE over scalp which was misdiagnosed as pyogenic granuloma elsewhere and had undergone electrocautery with recurrence of lesions after few months.

Case report

A 45 year old female presented to dermatology department with multiple raised red lesions over occipital scalp for 6 months which had increased in size during the last 3 months. The lesions were not painful but mildly pruritic and there was history of scant bleeding from lesions on repeated pricking by patient. There was no history of trauma or drug intake prior to development of lesions. The patient had similar lesions over the site 1 year back for which she had undergone electrocautery procedure. Cutaneous examination revealed multiple well defined discrete to coalescing, erythematous papules and a single nodule in a clustered pattern present over the 2x2 cm occipital area of scalp predominantly over the left side.(Fig 1) There was no active bleeding or crusting present. There was no palpable regional lymphadenopathy.

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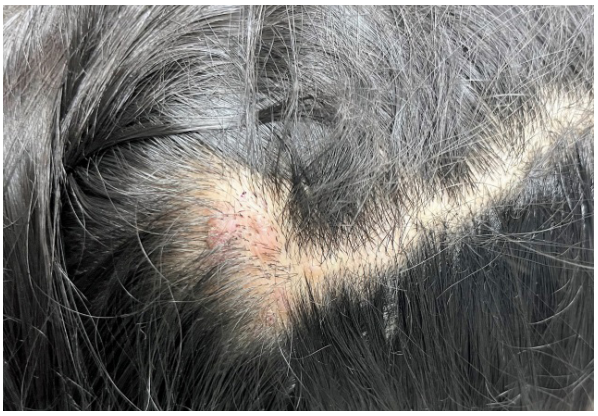

Fig 1- Clustered erythematous papules and a single nodule on occipital scalp

Haematological investigations were remarkable only for mildly elevated absolute eosinophil count. Complete excision biopsy of the lesion revealed normal epidermis with presence of yeast form of malassezia. Dermis showed an ill-defined mass composed of numerous vascular spaces lined by rounded endothelial cells, some of which showed cytoplasmic vacuolation. There was inflammatory infiltrate consisting of lympho- histiocytes along with numerous eosinophils extending to subcutaneous tissue and surrounding major vessels. Foci of granulomatous inflammation with multinucleated giant cells were also noted in upper and mid dermis.

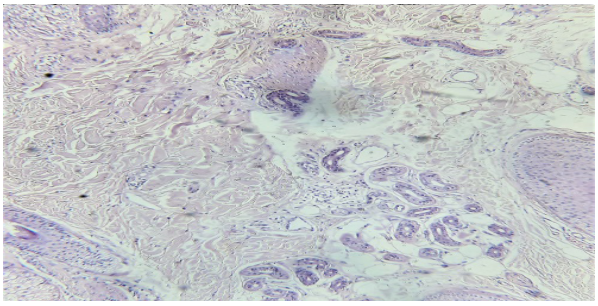

Fig 2- Group of dilated blood vessels lined with plump endothelial cells and perivascular mixed infiltrate (H&E- 40x)

The overall histopathological findings were consistent with angiolymphoid hyperplasia with eosinophilia. Immunohistochemistry could not be performed due to limitation of resources. The patient was reassured about the nature of the disease. There was no recurrence of lesions till 8 months of follow up.

Discussion

There are many hypotheses regarding the etiopathogenesis of ALHE including neoplastic process; reactive process to triggers by viral infections including Human immunodeficiency virus, herpes virus and HTLV1; hormonal imbalance (raised estrogens) and vascular shunt.[2]

The name "epithelioid hemangioma" was coined by Weiss and Enzinger in 1982 supporting the neoplastic hypothesis, but the most accepted theory is it being a reactive vascular response to various triggers, hence the term ALHE is more widely used.

The histopathology is fairly characteristic and shows relatively normal epidermis with marked proliferation of irregular blood vessels which are lined by plump epitheloid endothelial cells having abundant eosinophilic cytoplasm and large vesicular nuclei. They may sometimes protrude into the lumen giving a hobnail appearance and may also have cytoplasmic vacuoles. The other prominent component is the dense perivascular infiltrate consisting of lymphocytes and eosinophils with occasional plasma cells and histiocytes.

There are many differential diagnoses for ALHE including pyogenic granuloma, Kimura's disease, angiosarcoma, cylidroma, trichoepithelioma, and epitheloid hemangioendothelioma. The closest clinical and histopathological differential diagnosis is Kimura's disease. Kimura's disease has a similar predilection for gender, site, clinical appearance of lesions and relative indolent course of disease. It is differentiated from ALHE by presence of regional lymphadenopathy, lesions having a more subcutaneous component, more severe peripheral and tissue systemic eosinophilia, raised serum IgE levels, and absence of epitheloid endothelial cells. It is important to differentiate between these two entities as Kimura's disease has a more chronic course and often requires systemic immunosuppression and long term follow up.[3]

Surgical excision is the choice of treatment with least chances for recurrence. In patients unwilling for surgery, other options which have been tried include topical/ intralesional corticosteroids; cryotherapy; electrocoagulation; LASERs including pulsed dye and carbon dioxide and radiofrequency ablation, but none of these has given consistent results in terms of clearance and prevention of recurrence.[4]

Our case had clustered lesions at the favoured site for ALHE, but had been misdiagnosed as pyogenic granuloma. Incomplete clearance of margins may have led to recurrence which prompted the patient to seek another opinion. The peripheral eosinophilia in our case can be seen in 10-20% cases of ALHE also and the histological findings confirmed the clinical diagnosis. To conclude, it is prudent to take a biopsy of the nodular lesions over scalp and ear area to avoid misdiagnosis and administer proper treatment.

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