

Angiolymphoid Hyperplasia with Eosinophilia in The External Auditory Canal - A Rare Presentation

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Abstract

Angiolymphoid hyperplasia with eosinophilia is an uncommon idiopathic condition that manifests as isolated or grouped papules or nodules in the head and neck region, especially in the periauricular region. It is therefore necessary for an ENT and Head Neck surgeons to be aware of this condition and its management.

Introduction

Angiolymphoid hyperplasia with eosinophilia is a benign uncommon skin disease with approximately 85% cases; most of them are in peri-auricular region or on the forehead or scalp. While ALHE shows some similarity to Kimura's disease, it is separate entity.^{1,2} We report this rare entity in a 56 year old male, presented with external auditory canal nodules that was treated surgically.

Case Report

A 56 year old male came to ENT OPD with left external auditory canal nodules of 6 months duration. He gave history of painless gradually enlarging nodules in left ear with occasional pruritus of overlying skin. He had no history of ear discharge, trauma or constitutional symptoms. On physical examination he was averagely built and nourished. Examination of ear showed nodular masses in left external auditory canal and in external auditory meatus. On otoscopic examination tympanic membrane was not completely seen. Overlying skin was normal. No cervical lymph node was palpable. The systemic examination was unremarkable. Haematological and urine examinations were normal. Renal function tests were within normal limits. X-ray mastoid was suggestive of pneumatic air cells.

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Whole nodular masses were removed surgically from the external auditory canal and external auditory meatus and sent for histopathological evaluation. Biopsy showed increased number of thick walled and dilated blood vessels (capillaries and venules) involving the whole of reticular dermis. Around these vessels and at times infiltrate in their walls is an infiltrate of lymphocytes and eosinophils. The lining of these vessels showed thick endothelial cells projecting into the lumina giving a 'hobnail' appearance.

The diagnosis of ALHE was made on characteristic histological features.

It has been 6 months post surgery, patient is well without any local recurrence.

Discussion

ALHE is a rare skin disease occurring slightly more frequently in females in the 3rd and the 4th decade with no racial predominance. It manifests as isolated or grouped papules, plaques or nodules in the skin of the head and neck region. Most patients present with lesions in the periauricular region, forehead or scalp. Rare sites of involvement include the hands shoulders, breasts, penis oral mucosa and orbit.^{3,4} ALHE is idiopathic, whether it is neoplastic or reactive state is uncertain, but a reactive cause is favoured. The clinical presentation of papules may suggest ALHE but a biopsy is required to establish the diagnosis. ALHE shows characteristic histological features including a proliferation

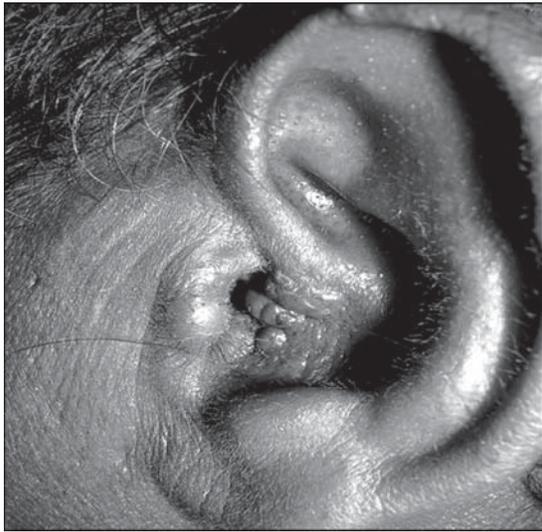


Fig. 1 : Showing nodules in the external auditory canal.



Fig. 2 : Slide showing histological features of ALHE.

of small blood vessels, many of which are lined by enlarged endothelial cells with uniform ovoid nuclei and intracytoplasmic vacuoles.

These distinctive endothelial cells have been described as having a 'Hobnail' or 'Cobblestone' appearance. In addition the perivascular and interstitial infiltrate composed primarily of lymphocytes and eosinophils. Eosinophils typically comprise 5-15 % of the infiltrate; rarely they can account for as much as 50% of the infiltrate. Occasionally the infiltrate is devoid of the eosinophils. The lymphoid aggregate with or without follicle formation are typical.

Differential diagnosis includes

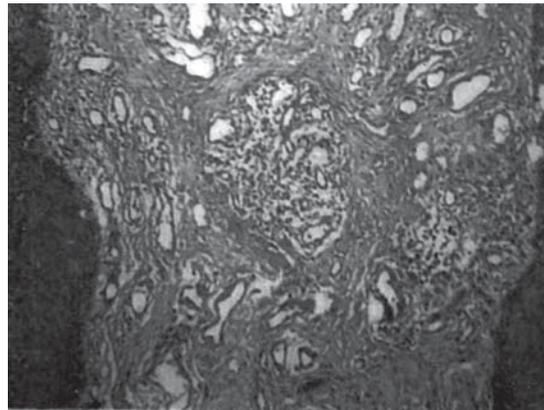


Fig. 3 : Slide showing histological features of ALHE.

angiosarcoma, haemangioendothelioma, kimura's disease, Kaposi's sarcoma; dermatofibroma etc. ALHE should be distinguished from Kimura's disease. Kimura's disease is associated with markedly elevated peripheral eosinophilia and elevated serum IgE levels as well as lymphadenopathy. Histologically Kimura's disease is characterized by marked lymphoid hyperplasia with an inflammatory infiltrate rich in eosinophils, eosinophilic microabscesses and fibrosis. In Kimura's disease association with nephritic syndrome is strong.

Treatment

- a) *Medical*: Intralesional steroids⁵ and irradiation have been used but not very effective. Other treatments that have been reported include topical imaquimod, topical tacrolimus and interferon alpha 2b.⁶
- b) *Surgical*: Local excision with a safe margin of healthy tissue is the treatment of choice, but insufficient removal can lead to recurrence. Pulse dye laser and the CO₂ laser have been used with some success.

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TENOFOVIR AS PRE-EXPOSURE PROPHYLAXIS

'The severity of the HIV epidemic and the potential benefits of pre-exposure prophylaxis should lead us to begin planning for implementation as soon as possible'

Great advances have been made in the treatment of HIV infection, but traditional methods to prevent transmission such as condom use do not stop 11000 people becoming infected every day. However, trials of vaccines and other preventive strategies have had mixed results. Lynn Paxton and colleagues discuss the potential use of the antiretroviral drug tenofovir as pre-exposure prophylaxis. Many trials for this use are underway in countries with high HIV prevalence. Yes, if tenofovir proves successful as a prophylactic, many issues will need to be addressed, such as to whom it should be given, the possibilities of adverse reactions to the drug, and the development of viral resistance if a person becomes infected with HIV while taking prophylactic tenofovir. Other important topics include the possibility of increases in risky behaviours, and the public-health programmes that might need to be planned to educate at risk groups such as men who have sex with men.

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