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CLINICAL REPORT

Angiolymphoid Hyperplasia with Eosinophilia of the External Auditory Canal

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Abstract

Angiolymphoid hyperplasia with eosinophilia is a rare, benign, vaso-proliferative disease with a dense eosinophilic inflammatory infiltrate seen in the intradermal or subcutaneous region. It is also termed an epitheloid hemangioma, and it mostly affects females rather than males. It is seen as reddish to brown, fleshy, proliferative, papular, or nodular lesion affecting the head and neck region, most commonly the auricular and periauricular regions. The aetiology is unknown, but the most likely causes are trauma, infection, and hyperestrogenic condition like pregnancy or the use of oral contraceptive agents. It usually measures about 2–3 cm in size. It should be differentiated from Kimura disease, which is characterized by chronic inflammation and large subcutaneous nodules in the head and neck region. Here is a rare case report of angiolymphoid hyperplasia in a middle-aged male patient involving the external auditory canal of the left ear.

Keywords Angiolymphoid hyperplasia · Eosinophilia · Epitheloid hemangioma · Kimura · Auricular region

Introduction

Angiolymphoid hyperplasia was first described by Wells and Whimster in 1969 and was thought to be a late-stage form of Kimura disease [1]. Now it is described, differentiated from Kimura and considered a separate entity [2]. The tumour is rare, benign, and characterised by the proliferation of blood vessels lined by plump endothelial cells admixed with a dense inflammatory infiltrate of eosinophils, mast cells, and lymphocytes. Weiss and Enzinger wanted to differentiate it from malignant vascular tumours and epithelial hemangioendothelioma. For this, they have introduced the term "epitheloid hemangioma." The aetiology is idiopathic [3], and the likely cause is trauma [2, 3] infection, and hyperestrogenic conditions like pregnancy [4], oral contraceptive agents and immunisation. It is more commonly seen in middle-aged women than in men. Clinically, it is seen as reddish to brown, single or multiple fleshy proliferative, papular, or nodular lesions, mostly affecting the intra-dermal or subcutaneous layer of the head and neck region, most commonly the auricular or preauricular area, with a size measuring about 2–3 cm [4]. The diagnosis is made by biopsy and histopathological examination. Treatment options include surgical excision, intralesional steroids, and topical tacrolimus. Surgical excision is the treatment of choice. Recurrence is most commonly seen in 33–50% of individuals.

Case Report

A 40-year-old male patient presented with insidious onset and gradually progressive swelling in the left ear. He reported occasional episodes of left ear bleeding but denied any history of ear discharge, trauma, itching, pain, or hearing loss. No history of similar complaints in the past. On clinical examination, there were multiple, reddish brown, fleshy,

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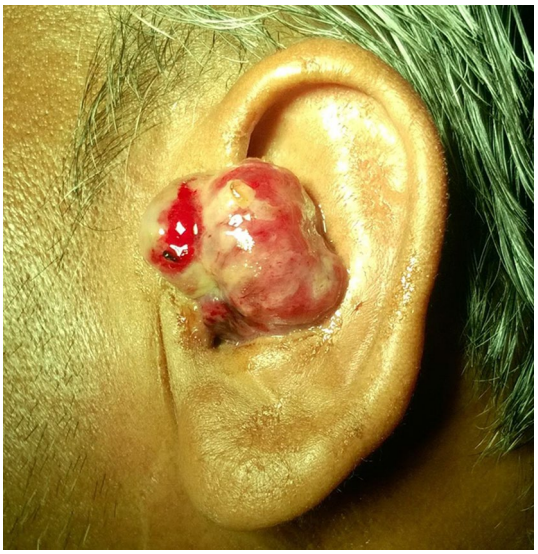

Fig. 1 Mass extrusion from external auditory canal

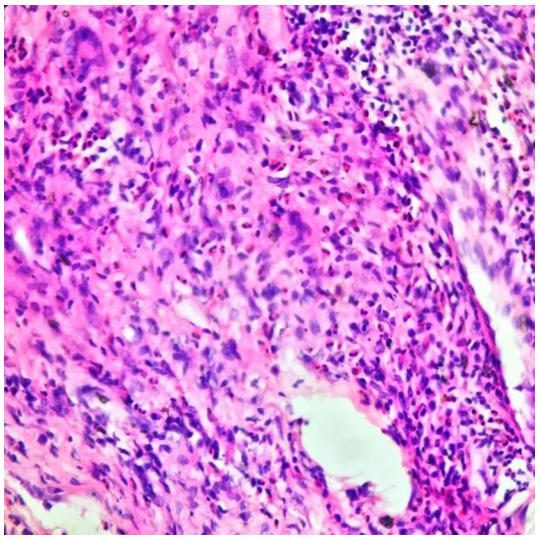

Fig. 2 Histopathological examination of the specimen

smooth-surfaced, nodular, non-tender, non-pulsatile, non-compressible lesions seen arising in the floor of the lateral end of the left external auditory canal and mass extruding out of the left external auditory canal (Fig. 1). The tympanic membrane appeared normal on microscopic examination in both ears. Other local examination findings appeared normal in both ears. No lymphadenopathy was noted. Other systems were normal. The patient underwent comprehensive blood investigations, which revealed mild eosinophilia (Absolute Eosinophil count of 600/microlitre) along with normal IgE levels. Audiological examinations showed mild conductive hearing loss of 30 dB air–bone gap in the pure-tone audiometry of the left ear and normal findings in the right ear. A CT scan ruled out middle ear and mastoid involvement. Surgical excision was performed under local anaesthesia, and a biopsy specimen was sent for histopathological examination. The histopathological findings confirmed the diagnosis of angiolymphoid hyperplasia with eosinophilia, characterized by the proliferation of small vessels with plump endothelial cells surrounded by a high density of eosinophils and lymphocytes (Fig. 2). Postoperative follow-ups were uneventful, with no complications. Postoperatively, the patient received a tapering dosage of oral steroids (prednisolone) to prevent disease recurrence. The steroid regimen consisted of 60 mg of prednisolone (1 mg/kg/day) in three divided doses for the first 5 days, 40 mg in two divided doses for the next 5 days and 20 mg once daily for the last 5 days. The patient underwent regular follow-up visits every 2 weeks for the initial 2 months, followed by monthly visits for the first six months, and then every 6 months for 2 years. Repeated postoperative blood investigations demonstrated normal eosinophil count (Absolute eosinophil count of 350/microlitre) with normal Ig E levels and audiological examination revealed normal findings in both ears. To date, no recurrence has been observed.

Discussion

Angiolymphoid hyperplasia with eosinophilia is characterised by the florid proliferation of blood vessels lined by endothelial cells and admixed with a dense inflammatory infiltrate of eosinophils, lymphocytes, and mast cells. Many theories have been proposed to explain the aetiology of angiolymphoid hyperplasia, which may be a traumatic, neoplastic, hyperestrogenic condition with an infectious mechanism and a possible association with HIV [5]. Some cases of reported angiolymphoid hyperplasia with peripheral T-cell lymphoma are noted [6]. T-cell receptor gene (TCR) rearrangement and monoclonality association have been detected in angiolymphoid hyperplasia [5]. 20% of cases show eosinophilia and elevated IgE levels are a rare finding in Angiolymphoid hyperplasia [6]. According to the literature, the prevalence of the disease is seen in a range of 0.7 months to 91 years, and the mean age of presentation is 37.6 years [7]. Early onset, longer duration, and multiple lesions are associated with a high recurrence rate after the excision of this disease [7]. It is most commonly seen in women. The ear is the most common site of the disease, while involvement in the nose is rare. It presents as reddish to brown, single to multiple, fleshy papular to nodular lesion.

Differential diagnoses of tumours in the external auditory canal include benign lesions like Kimura disease, squamous papilloma, sarcoidosis, exostosis, seborrheic keratosis, keratoacanthoma, hemangioma, angiolymphoid hyperplasia, and

malignant lesions like squamous cell carcinoma, Kaposis sarcoma and Basal cell carcinoma.

Treatment of angiolymphoid hyperplasia is challenging because of its high rate of recurrence [33–50%] [8]. Surgical excision is the treatment of choice with good disease-free margins. Other surgeries include laser surgery using a carbon dioxide laser or an Nd: YAG laser [9]. The disadvantage of laser surgery is the risk of scarring. A few cases have been reported for cryotherapy, Red light photodynamic therapy [10], corticosteroids [8], chemotherapy agents, Interferon -alpha and isotretinoin treatment. In our case, surgical excision was the treatment of choice with oral corticosteroids, and no recurrence has been noted in the follow-up of the case till now.

Conclusion

Our case is peculiar because it involves a male patient with Angiolymphoid hyperplasia, which is most commonly seen in women. Because of the disfiguring nature of the disease and its high rate of recurrence, effective treatment should be chosen for a high success rate. It is important to distinguish Angiolymphoid hyperplasia from a similar disease called Kimura disease by its size, eosinophil count, and IgE levels.

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Declarations

Conflict of interest The authors declare that they have no conflict of interest.

Ethical Approval Before starting the study ethical clearance was taken from the institutional ethical committee as per the Declaration of Helsinki.

Informed Consent Informed consent was obtained from the patient and their relatives involved in the study.

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