Angiolymphoid hyperplasia with eosinphilia presenting with postauricular swelling

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Angiolymphoid hyperplasia with eosinophilia (ALHE) is an unusual vascular proliferative condition. The entity is almost unknown to the Otolaryngologist though it has a predilection for the head and neck particularly the region around the ear. In this case report ALHE presented as a postauricular swelling.

A 48-year-old Saudi male was referred from General Surgery to the ENT clinic with a painless swelling over the right postauricular region of 2 months duration. The swelling was gradually increasing in size and was associated with itching occasionally. On physical examination there was a superficial swelling on the right mastoid region, 3 x 2 cms in size. The skin over the swelling was free and unremarkable. It was nontender, mobile, firm and rubbery in consistency, with well circumscribed margins. No other swellings were noted in the head and neck region. Ear, nose and throat and systemic examination was normal. Patient was diabetic (non insulin dependant diabetes mellitus).

Routine blood test was carried out. Fine needle aspiration cytology (FNAC) was suggestive of a benign mesenchymal lesion. As FNAC was not conclusive and a 7 day course of antibiotics and anti-inflammatory drugs showed no response, we opted for excisional biopsy under local anesthesia. A well-circumscribed subcutaneous mass was found: the operative field was unduly bloody, there were 2 big vessels supplying the mass which were ligated and small bleeders were cauterized. The mass was completely excised, and the specimen was sent for histopathology. The mass measured 2 x 1.5 x 1 cm and was firm. On section it was grey white with a tan periphery. The histology was characterized by an exuberant proliferation of small blood vessels, and a heavy chronic inflammatory cell infiltrate. Many of the vessels were lined by plump endothelial cells projecting into the lumen like tombstones the cells had an epithelioid appearance with abundant pale eosinophilic cytoplasm and large round vesicular nuclei. The infiltrate consisted of lymphocytes, plasma cells and eosinophils, the last were particularly numerous **(Figure 1)**. The diagnosis was epithelioid hemangioma (ALHE). There was no evidence of arteriovenous malformation. Subsequent follow up for 6 months was uneventful.

Angiolymphoid hyperplasia with eosinphilia is an unusual condition poorly recognized by the Otolaryngologists. The term was first used in 1969 by

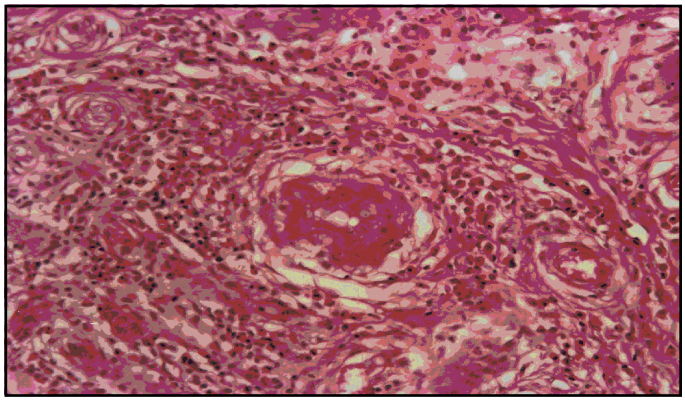

Figure 1 - Histological section shows a blood vessel lined by plump endothelial cells (with characteristic large round vesicular nuceli) protruding into the lumen. The infiltrate around is almost entirely eosinophilic. (Hematoxylin and eosin magnification x 200).

Wells and Whimster[1] to describe certain subcutaneous nodules in the head and neck region. Over the years, many investigators further elaborated and defined its characteristics and a plethora of synonyms including inflammatory angiomatous nodule and atypical or pseudopyogenic granuloma were proposed. Finally, the entity was designated epithelioid hemangioma.[2] In this report; however, we have used the term ALHE for the entity as it is well entrenched in the dermatological and surgical literatures. The case presented here displays many of the clinical and pathological characteristics of ALHE. It is an uncommon disorder occurring in all parts of the globe though it is somewhat more often seen in the Orient. Most of the patients present between the second and fifth decade of life, the condition being extremely rare in the pediatric and elderly populations.

Angiolymphoid hyperplasia with eosinophilia has a predilection for the head and neck, the external ear and the periauricular region being the most frequently affected. Occasionally other skin surfaces, oral mucosa and pharynx may be involved.[3] In the skin, it presents as a discrete slow-growing plum colored papule or nodule sited in the dermis or the subcutis. Nodules are usually single, but in a third of the patients multiple lesions develop in the vicinity. They are usually asymptomatic, with regards one-fifth being pruritic and fewer still showing excoriation and bleeding. Lymphadenopathy is uncommon, and only a fifth of the patients have blood eosinophilia though all nodules, by definition, moderate to abundant eosinophils. The natural history is one of an indolent growth, slowly increasing in size and adjacent lesions gradually coalescing over a period of months and years with persistent symptoms. Due to the multilobulated nature of the lesion, margins are difficult for the surgeon to discern and superficial ablation leaves a deep component that can be the source of recurrence.

Although as many as one third of the lesions recur, virtually none have produced metastasis.

In the postauricular location as in our case, ALHE is an important differential diagnosis after excluding the common conditions like lymphadenopathy, sebaceous cyst and lipoma. Fine needle aspiration cytology is not helpful in the definitive recognition of ALHE, histologic identification of the lesion in the excised tissue being the only basis of diagnosis.[4] Kimura's disease which was formerly thought to be identical to ALHE as of a few histological similarities of the inflammatory component has now been conclusively shown to be an entirely unrelated condition. The plump endothelial cell characteristic of ALHE remains the key to separation of the 2 entities.[5]

Many different forms of treatment have been utilized for the eradication of the lesions of ALHE. However, treatment modalities that do not affect the deep components of the lobulated angiomatous lesions cannot be expected to be curative. Coagulation necrosis may be produced with carbon dioxide or Argon laser, but their tissue effects decrease with the depth of penetration, and the deeper aspects of ALHE are not affected by these laser treatments.[6] There is a consensus among the present day investigators that the best results are obtained with complete surgical excision of the lesion. Our patient's lesion was subjected to complete excision and follow-up for 6 months showed no signs of recurrence. In cosmetically sensitive areas, after a punch biopsy confirmation of the diagnosis, the newer long pulsed tunable dye laser is being employed in the treatment of superficial lesions with less scarring and ablating deeper blood vessels.[7]

Despite its rarity ALHE is an important entity for the Otolaryngologist to recognize. Though its behavior is benign, it is a persistent disorder and its presence results in irritating symptoms and local disfigurement it has been aptly summed up in the phrase "Persistent pruritic plaque of the ear and the periauricular region." Definitive diagnosis depends on surgical excision and histologic examination. Effective treatment depends on the adequacy of such excision.

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