Angiolymphoid hyperplasia with eosinophilia presenting with postauricular swelling

Ibrahim S. Akram, FACHARTZ, Syed S. Raza, MCPS,
Latifi Asrar, MS DLO(Lon), Mohammed Faizuddin, MD(Path).

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 eosinophilia is an unusual condition poorly recognized by the Otolaryngologists. The term was first used in 1969 by Wells and Whimster 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. 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. In the skin, it presents as a discrete slow-growing plum colored 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. 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.
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. 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.

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 angiomatic 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. 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.

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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From the Department of ENT (Akram, Raza, Asrar), Department of Pathology (Faizuddin), Buraidah Central Hospital, Buraidah, Kingdom of Saudi Arabia. Address correspondence and reprint requests to Dr. Latifi Asrar, PO Box 2290, King Fahad Specialist Hospital Campus, Buraidah, Kingdom of Saudi Arabia. Tel. +966 (6) 3238314. Fax. +966 (6) 3238314. E-mail: latiftiasrar@hotmail.com

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