

## Angiolymphoid Hyperplasia with Eosinophilia of the Scalp: A Case Report

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**Abstract:** A 38 year old female presented with a painless swelling in the scalp. Clinical diagnosis was epidermoid cyst. Fine needle aspiration cytology showed sheets of lymphoid cells along with numerous eosinophils. Subsequent histopathological examination showed eosinophilic plump endothelial cells in nests and sheets in a fibrous background with lymphoid aggregates and plenty of eosinophils, suggestive of angiolymphoid hyperplasia with eosinophilia.

**Keywords:** Angiolymphoid hyperplasia, eosinophilia, scalp

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### I. Introduction:

Angiolymphoid hyperplasia with eosinophilia, also known as epithelioidhemangioma, most commonly occur in the subcutaneous tissue of the head and neck.<sup>[1],[2]</sup> This entity occurs predominantly in patients in the third to fifth decade.<sup>[2]</sup>

### II. Case Report:

A 38-year-old female presented with a painless swelling in the scalp. On examination, there was a lump measuring 2 x 2 cm. The mass was firm, mobile, and non-tender. The clinical diagnosis was epidermoid cyst. Fine needle aspiration cytology showed sheets of lymphoid cells along with numerous eosinophils [Figure 1]. The mass was excised and sent for histopathological examination.

Histopathology showed eosinophilic plump endothelial cells in nests and sheets in a fibrous background with lymphoid aggregates and eosinophils [Figure 2]. Peripheral blood did not show eosinophilia.

### III. Discussion:

Angiolymphoid hyperplasia with eosinophilia (ALHE), also known as epithelioidhemangioma, is a uncommon benign vaso-proliferative disease. It presents most commonly in patients aged 20-50 years, with a mean onset of 30-33 years.<sup>[3]</sup> ALHE affects women more often than men.<sup>[4]</sup> The condition is more commonly encountered in the Asian population followed by Caucasians. It is rare in the black population.<sup>[4]</sup>

The majority of patients present with a mass of a year or less in duration.<sup>[6]</sup> Systemic eosinophilia of 6-34% is seen in 20% of patients with ALHE.<sup>[5]</sup> Serum hypereosinophilia is inconstant and is not required to make the diagnosis of ALHE.<sup>[7]</sup>

Despite the benign nature of ALHE, considerable controversy still exists as to the basic nature of these lesions. Some authors consider ALHE as a neoplasm developing from endothelial cells; others suggest that it is secondary to an inflammatory vascular reaction secondary to complex immunologic mechanisms. Many other hypotheses have been reported implicating environmental factors such as insect bite, trauma, and infections. Some authors consider that arterio-venous shunt is the main etiopathogenetic mechanism. The predominance of T lymphocytes and a rearrangement of TCR receptor in some cases made some authors suppose that ALHE is a low-grade neoplastic disease secondary to various stimuli.<sup>[8],[9]</sup>

More recently, Kempf *et al.* evaluated seven patients with ALHE and found that five of the seven patients showed a clonal T-cell population and proliferative T-cell activity, suggesting that a subset of these lesions might represent T-cell lymphoproliferative disorders of benign or low grade malignancy. Regardless of the anatomic site involved, there is not a single reported case of ALHE with aggressive behavior. Hence, it is fair to assume that these lesions are benign tumoral conditions.<sup>[10]</sup>

Grossly, these are usually circumscribed lesions of subcutis or dermis measuring 0.5-2.0 cm in size, with rare examples exceeding 5 cm. Less frequently they involve deep soft tissue, vessels, and parenchymal organs. Apart from the size, gross characteristics of this process are not well described. Many of them have a rather nonspecific nodular appearance and some with retained blood may have an appearance suggestive of a hemangioma. Occasionally, subcutaneous examples may resemble a lymph node because of its circumscription and a peripheral lymphoid reaction.<sup>[11]</sup>

Epithelioidhemangioma are characterized by a prominent proliferation of small, capillary-sized vessels lined by plump, epithelioid endothelial cells. The vessels typically have an immature appearance and they may lack a well-defined lumen, but they are well formed with single cell layering of the endothelium and an intact myopericytic/smooth muscle layer. The endothelial cells have amphophilic or eosinophilic cytoplasm that is sometimes vacuolated and they contain a single, relatively large nucleus with an open chromatin pattern and often, a central nucleolus.<sup>[11]</sup>

ALHE is usually well demarcated from the surrounding soft tissue, and commonly, it is associated with (sometimes centered around) a larger vessel, usually a muscular artery. An inflammatory milieu rich in eosinophils, plasma cells, and lymphocytes is present in the overwhelming majority of cases. Eosinophils usually account for 5-15% of the infiltrate, though in rare cases eosinophils may account for up to 50%.<sup>[3]</sup> Cytokines like interleukin-5 which interfere with the production and activation of eosinophils are supposed to play a key role in the pathogenesis of eosinophil rich inflammatory milieu in ALHE.<sup>[12]</sup>

Aggregation of lymphocytes is typical with a tendency to form lymphoid follicles. The lack of sinus structure indicates that these follicles arise in the subcutaneous tissue.<sup>[13]</sup>

It is also common to encounter epithelioid endothelial cells within the lumen of the larger vessel, either replacing part of the normal endothelial lining or "coating" fibrin fronds, as seen in papillary endothelial hyperplasia.<sup>[11]</sup>

Two types of ALHE lesion are described by Wells and Whimster. The early lesion demonstrates a predominance of rapidly proliferating atypical vasculature. However, late lesions illustrate maturation of these atypical blood vessels with thickened walls along with a high prevalence of lymphoid follicles seen towards the periphery of the lesion.<sup>[14]</sup>

Complete local excision and follow-up are optimal management for epithelioidhemangioma. Local recurrence is reported to occur in up to one-third of patients.<sup>[15]</sup> ALHE rarely regresses spontaneously; however, malignant transformation does not occur.<sup>[16]</sup>

#### **IV. Conclusions:**

Angiolymphoid hyperplasia with eosinophilia, a benign vascular tumor, commonly presents in third decade with a female preponderance. Surgical removal is the treatment of choice. There is no evidence of malignant transformation.

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