ANGIOLYMPHOID HYPERPLASIA WITH EOSINOPHILIA OVER CHEEK - A CASE REPORT

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ABSTRACT

BACKGROUND
Angiolympoid hyperplasia with eosinophilia (ALHE) is an uncommon benign, reactive vasoproliferative disease which is not accompanied by regional lymphadenopathy, peripheral blood eosinophilia and elevated IgE levels. It commonly affects patients aged 20–50 years and presents as vascular nodules in the dermal and subcutaneous tissues of the head and neck, particularly around the ear. Initially, it was thought to be related to Kimura’s disease but recent studies indicate that Kimura disease differs from ALHE in several clinical and histopathologic features. Complete local excision and follow-up are the optimal management for ALHE. A 36-year-old female diagnosed as ALHE on histopathology is presented here.

KEYWORDS
Angiolympoid Hyperplasia, Eosinophilia, Kimura’s Disease.


BACKGROUND
Angiolympoid hyperplasia with eosinophilia (ALHE) is an uncommon benign, reactive vasoproliferative disease. It presents with painless, vascular nodules in the dermal and subcutaneous tissues of the head and neck, particularly around the ear but has also been reported in the scalp, lip, tongue, orbits and conjunctiva. It commonly affects patients aged 20–50 years, with a mean onset of 30–33 years. It should be distinguished from Kimura’s disease which is a chronic inflammatory condition, producing large subcutaneous nodules on the head and neck with normal overlying skin. Both share histopathological similarities, such as involvement of dermis and subcutaneous infiltrate comprising lymphocytes and eosinophils, proliferation of endothelial cells and absence of adnexal structure involvement.

Surgical excision and intralesional steroids are the main treatment options. Local recurrence is reported to occur in up to one-third of patients. ALHE rarely regresses spontaneously; however, malignant transformation does not occur.

Case Report
A 36-year-old female presented with swelling over the left cheek since 2 years accompanied by itching over the lesion occasionally. Patient was relatively asymptomatic before 2 years when she developed papular lesions over the left cheek which were hard in consistency and increased in size over a period of one year. No history of trauma was elicited. There was no history of fever, pain over the lesion or any infective foci in the body. General examination showed no lymphadenopathy, clubbing, icterus and cyanosis. Patient had past history of similar complaint for which surgery was done, but the lesion reappeared after a period of 6 to 8 months.

On clinical examination, swelling of 3 x 4 cm in size which was skin coloured and soft in consistency was present over left cheek. [Figure 1]. Few scars of past surgery were also seen. Oral cavity, genitals, nails and anal region were normal.

Routine investigation including haemogram showed no eosinophilia. Patient had no other comorbid condition. Once again, the mass was removed and sent for histopathology which showed haphazard nodular vascular proliferation in the subcutaneous plane, accompanied by a dense lymphoid infiltrate, rich in eosinophils in addition to histiocytes and plasma cells. The vascular component has a variable size that ranged from capillary-sized to medium sized vessels lined by plump endothelial cells, many with hyperchromatic nuclei, copious eosinophils in cytoplasm and inconspicuous nucleoli. [Figure 2a & b]. Changes were suggestive of ALHE.

Figure 1. Skin coloured soft swelling over left cheek
The aetiopathogenesis of ALHE is not well known. Trauma, hormonal changes and infections, Human T-cell lymphotropic virus or Human Herpes virus 8 have been suggested to play a role in the pathogenesis. Associations of ALHE with nephrotic syndrome and pregnancy have been described. Over-expression of oestrogen and progesterone receptors were detected in pregnant women. Interleukin and vascular endothelial growth factors were also found to be increased in some cases.

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. There is not a single reported case of ALHE with aggressive behaviour. Hence, ALHE lesions are benign tumoural conditions.

Initially, ALHE was thought to be related to Kimura’s disease but recent studies indicate that Kimura disease differs from angiolymphoid hyperplasia with eosinophilia in several clinical and histopathologic characteristics including male predominance, striking lymphadenopathy, higher incidence of peripheral blood eosinophilia and lack of the distinctive endothelial cell as a marker.

KD is characterised by lymphoid nodules with germinal centres which may extend from the dermis to the underlying fascia and muscles. Lesions show a distinct eosinophilic infiltrate with microabscesses. Vascular proliferation is not always present but many canalised capillaries lined by flat endothelial cells may be seen. Systemic eosinophilia is almost always present, in approximately 98% of cases in comparison to 20% in ALHE.

Differential diagnoses of ALHE include cutaneous lymphoma, cavernous haemangioma, pyogenic granuloma, Kaposi’s sarcoma, angiomatous lymphoid hamartoma, granuloma faciale, polyarteritis nodosa, pseudolymphoma (Lymphocytic infiltrate of Jessner, lymphocytoma cutis), persistent insect bite reaction, injection site granuloma and bacillary angiomatosis.

Epithelioid haemangioma are characterised by a proliferation of small, capillary-sized vessels lined by plump, epithelioid endothelial cells. The vessels typically have an immature appearance and lack a well-defined lumen, but are well formed with single cell layering of the endothelium and an intact myopericytic/smooth muscle layer. Aggregation of lymphocytes is typical with a tendency to form lymphoid follicles. Lack of sinus structure indicates that these follicles arise in the subcutaneous tissue.

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.

The treatment of choice for ALHE is surgical excision, but due to high rate of recurrence, other less invasive therapeutic modalities may be employed. Intralesional corticosteroid therapy is effective with recurrence rates similar to surgical excision, but with better aesthetic result. Other treatments such as cryotherapy, photodynamic therapy and application of imiquimod have good results. The latter acts by induction of the production of interferon-α and induction or inhibition of
certain cytokines, mainly interleukin, implicated in the genesis of ALHE.[13]

REFERENCES