CASE REPORT

Auricular Angiolymphoid Hyperplasia with Eosinophilia

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ABSTRACT

Angiolymphoid hyperplasia with eosinophilia is an uncommon, benign, angioproliferative cutaneous disorder. Previously, angiolymphoid hyperplasia with eosinophilia and Kimura's disease were regarded as identical conditions, however, recent work identifies them as separate entities. It is clinically manifested by solitary or multiple, red to brown firm papules and nodules, occurring in the head and neck region with a predilection for the peri-auricular area. It usually occurs during the early and mid-adult life. It is more common in Caucasians with equal gender involvement. We report this condition in a 32 years old adult Pakistani male having red-brown papules in left ear.

Key words: *Angiolymphoid hyperplasia. Eosinophilia. Peri-auricular. Angioproliferative disorder.*

INTRODUCTION

Angiolymphoid hyperplasia with eosinophilia (ALHE) is a rare disorder presenting as solitary or multiple, red-brown coloured papules or nodules. It occurs in both the dermal and subcutaneous tissue. The site of predilection is the head and neck region.[1] Rarely, it has been reported on other body sites such as hands, feet, oral mucosa, orbit and parapharyngeal spaces. Both genders are equally affected and the disease is more prevalent in young to middle-aged adults than in children or elderly. It is a benign condition of unknown etiology, though it has been associated with inflammation, infection, trauma, pregnancy and hormonal changes.

We report a case of an adult Pakistani male having these lesions on left ear.

CASE REPORT

A 32 years old male reported to the Dermatology Department of Combined Military Hospital, Malir Cantonment, Karachi with 3 months history of mild pruritus and pain in left ear canal. For a similar duration, he had also noticed a cluster of skin coloured growths at the site, gradually increasing in size. There were no systemic complaints.

On examination, there were multiple, firm, red-brown papules on the upper part of the concha of left ear and extending to the crus of helix, as shown in Figure 1. The surrounding skin was normal and there was no regional lymphadenopathy. His systemic examination was unremarkable. Skin biopsy was performed and histopathological examination revealed an unremarkable epidermis. Dermis showed mixed inflammatory infiltrate with numerous lymphocytes and eosinophils. There was proliferation of dermal blood vessels, which were lined with prominent endothelial cells, exhibiting abundant eosinophilic cytoplasm and large nuclei. There was no lymphoid follicle formation (Figures 2 and 3). This picture was consistent with angiolymphoid hyperplasia with eosinophilia. His routine laboratory workup included complete blood count, urine analysis, liver and kidney function tests; they did not reveal any abnormality. There was no blood eosinophilia and his IgE levels (48 IU/ml) were within normal limits.

The patient was counselled about the benign nature of the cutaneous lesions and possible treatment options were discussed. He was advised intra lesional corticosteroids but the patient denied treatment and did not come for follow-up.

DISCUSSION

Angiolymphoid hyperplasia with eosinophilia is a benign entity of unknown pathogenesis, though it has been associated with inflammation, trauma, infection, pregnancy, hormonal changes and AV malformation. Vaccination has possibly induced ALHE in a 2 years old child.[2] Recently, there is some debate as to whether angiolymphoid hyperplasia with eosinophilia is a reactive or neoplastic disease. Chen *et al.* have suggested that ALHE may be a low-grade T-cell lymphoproliferative disorder, based on immuno-histochemistry and T-cell receptor (TCR) gene re-arrangement.[3]

Considerable controversy exists about its relationship with Kimura's disease. Previously, angiolymphoid hyperplasia with eosinophilia and Kimura's disease

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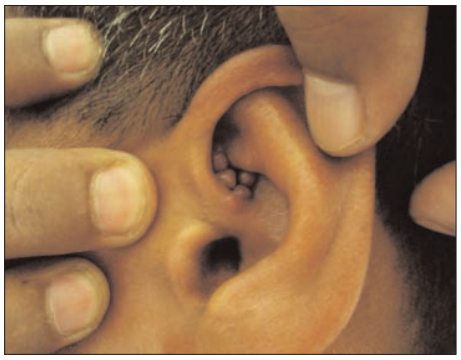

Figure 1: Dome shaped red-brown papules in the concha of ear and crus of helix.

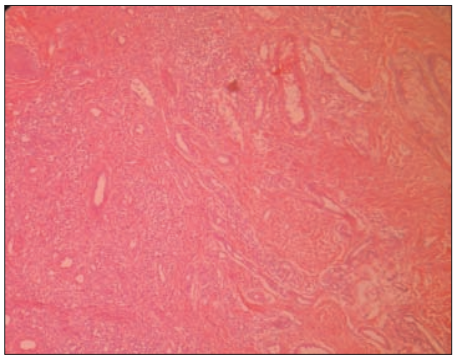

Figure 2: Histopathological examination demonstrating dermal vessels proliferation and infiltration of lymphocytes and eosinophils (H&E, x10).

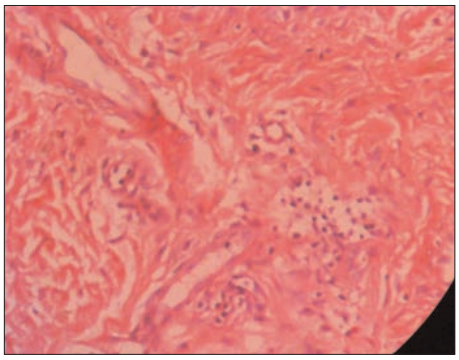

Figure 3: Dermal blood vessels lined by enlarged endothelial cells with a histiocytoid appearance, protruding into the lumen. A perivascular inflammatory infiltrate composed predominantly of lymphocytes as well as eosinophils (H&E. x40).

were regarded as same conditions, however, recent work has clearly identified them as separate entities.[4] Both are rare, and benign with chronic inflammatory conditions. Kimura's disease commonly occurs among the Oriental population. Some cases have been reported in the Caucasians and in the African population. Kimura's disease presents with painless subcutaneous nodules or plaques commonly located in the head and neck region particularly in the pre- or postauricular region and rarely in the oral mucosa, orbit, and the scalp. It may involve the parotid or other salivary glands and lymphadenopathy is present in more than 50% cases. It is often associated with peripheral blood eosinophilia, raised ESR and serum IgE levels. ALHE usually occurs during early and mid-adult life (20 - 40 years). There is a female predominance in white population. It is characterized by persistent and recurrent erythematous or hyperpigmented, dome-shaped, papules or nodules on the head and neck region, especially peri-auricular region. Other sites such as oral mucous membranes, paraphyrangeal space, and orbit have been involved rarely.[5] ALHE has been reported on hand.[6] The lesions are chronic with little tendency for spontaneous resolution. Systemic spread is extremely rare. Rarely, peripheral blood eosinophilia has been reported. There is no evidence of malignant transformation as yet.

Histologically, ALHE is characterized by a vascular and an inflammatory component. There is proliferation of small-sized blood vessels lined by enlarged endothelial cells with a histiocytoid appearance, protruding into the lumen. There may also be aggregates of non-canalized, plump endothelial cells. A perivascular inflammatory infiltrate composed of lymphocytes as well as eosinophils and mast cells is seen.

There are many methods of treatment but the final result is not always satisfactory to the patient. The lesions regress spontaneously in the majority of cases, after a variable time. It is reasonable to observe a small lesion for 3-6 months and await spontaneous regression.[7] Simple procedures like topical and intralesional steroid injection and cryotherapy are easy, safe and, at times, successful method of treatment. Laser therapy, photodynamic therapy, electrodessication and radiation are alternative treatment modalities. Resection of ALHE using Mohs micrographic surgery enables complete resolution of lesion with maximum conservation of tissue.[8]

ALHE has been sparsely reported in the local literature Muzaffar *et al*. have described the histopathological aspects of ALHE. Their patient had been clinically misdiagnosed as having sebaceous cyst of the scalp.[9] Badami *et al*. have reported a case of Kimura's disease, a condition related with ALHE but having its own distinct features. Their patient presented with pruritus and generalized lymphadenopathy. He had markedly elevated serum IgE levels and blood eosinophilia.[10]

REFERENCES

1. Chen AY, Janik MP, Moad JC, Rubin MB. Multiple papules and nodules of the scalp. Angiolymphoid hyperplasia with eosinophilia (ALHE). *Arch Dermatol* 2010; **146**:911-6.
2. Jeon EK, Cho AY, Kim MY, Lee Y, Seo YJ, Park JK, *et al*. Angiolymphoid hyperplasia with eosinophilia that was possibly induced by vaccination in a child. *Ann Dermatol* 2009; **21**:71-4. Epub 2009 Feb 28.
3. Chen JF, Gao HW, Wu BY, Tsai WC, Chiang CP. Angiolymphoid hyperplasia with eosinophilia affecting the scrotum: a rare case report with molecular evidence of T-cell clonality. *J Dermatol* 2010; **37**:355-9.
4. Seregard S. Angiolymphoid hyperplasia with eosinophilia should not be confused with Kimura's disease. *Acta Ophthalmol Scand* 2001; **79**:91-3.
5. Sánchez-Acosta A, Moreno-Arredondo D, Rubio-Solornio RI, Rodríguez-Martínez HA, Rodríguez-Reyes AA. Angiolymphoid hyperplasia with eosinophilia of the lacrimal gland: a case report. *Orbit* 2008; **27**:195-8.
6. Krapohl BD, Machens HG, Reichert B, Mailänder P. A rare vasoproliferative lesion: angiolymphoid hyperplasia with eosinophilia of the hand. *Br J Plast Surg* 2003; **56**:168-70.
7. Calonji E. Soft tissue tumours and tumour-like conditions. In:

Burns DA, Breathnach SM, Cox N, Griffiths CE, editors. Rook's textbook of dermatology. 8th ed. Oxford: *Blackwell Science;* 2010. p. 56.27-8.

8. Miller CJ, Ioffreda MD, Ammirati CT. Mohs micrographic surgery for angiolymphoid hyperplasia with eosinophilia. *Dermatol Surg* 2004; **30**:1169-73.
9. Muzaffar M, Mamoon N, Mushtaq S, Tahir M. Angiolymphoid hyperplasia with eosinophilia. *Pak Armed Forces Med J* 1997; **47**: 30-2.
10. Badami MA, Mahmood K, Uzma G, Ahmed A. Kimura`s disease with atypical presentation. *J Coll Physicians Surg Pak* 2001; **11**: 575-7.