

# Angiolymphoid Hyperplasia with Eosinophilia of Root of Nose: A Rare Phenomenon

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## ABSTRACT

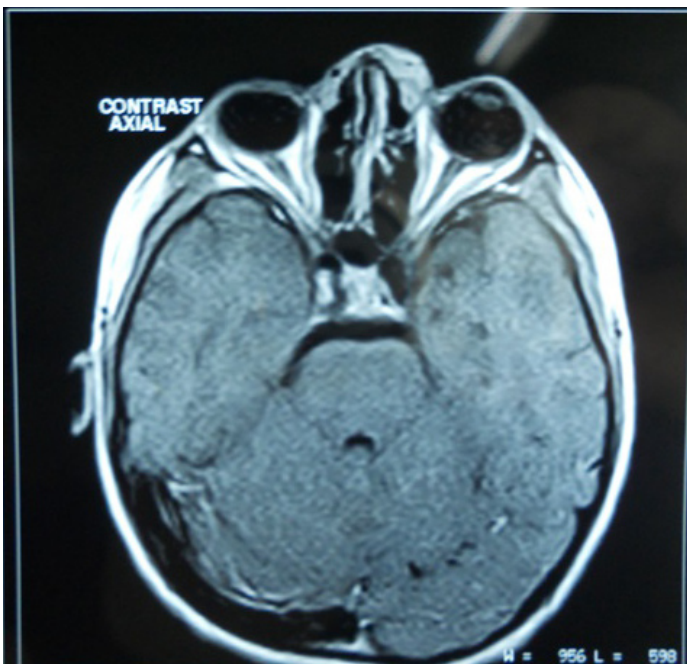
Angiolymphoid Hyperplasia with eosinophilia (ALHE), also known as epithelioid haemangioma, is an atypical vascular tumour which occurs with a predilection for the head and neck region. It is characterized by the presence of solitary or multiple lesions with varying clinical appearances, from intradermal papules to subcutaneous nodules.

**Keywords:** Angiolymphoid hyperplasia with eosinophilia, ALHE, Kimura's disease

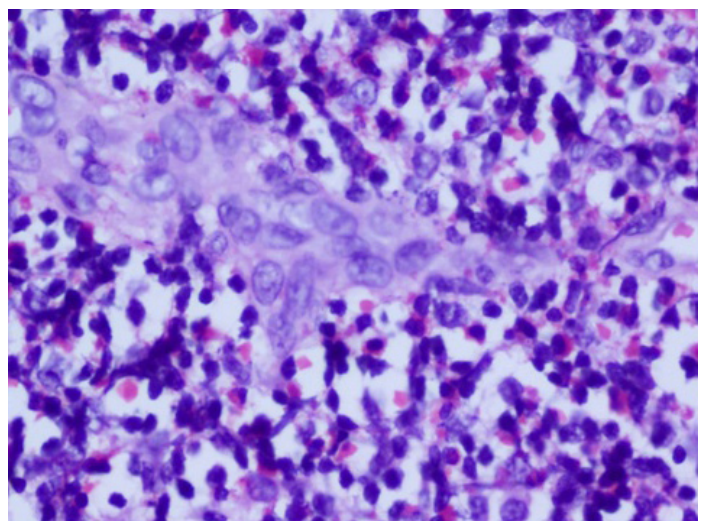
## CASE REPORT

A 12-year-old child from southern coastal India presented with a painless subcutaneous swelling on the left side of the root of nose, which had a duration of 2 years. It was insidious in onset and had gradually progressed over 2 years to attain the size which has been described below. There was no rapid increase or decrease in size of the swelling and no fever. There was no history of bleeding or trauma. On examination, he had a non tender swelling which was 3x2 cm in size, which extended from the glabella superiorly to the left ala inferiorly, medially from the root of the nose to the left inner canthus laterally. The surface was smooth, firm in consistency, with ill-defined borders and an intact nasal bone. There was no evidence of regional lymphadenopathy. MRI showed a swelling in the subcutaneous plane, which extended from the glabella along the left inner canthus to the left ala, with hypointensities on T1 and T2 weighted images, plain and fat suppression with diffuse homogenous contrast enhancements within and few flow voids along the superior and inferior margins [Table/Fig-1]. FNAC done on the swelling was inconclusive. Informed consent was obtained from the patient's guardian for surgery and documentation. The

patient underwent lateral rhinotomy under general anaesthesia and excision of the mass. The mass was vascular, with ill defined margins, without underlying infiltration and it was found to be in the subcutaneous plane with extension as has been described above. The mass was excised in toto and sent for a histopathological examination, which showed angiolymphoid tissue which consisted of proliferating blood vessels which were lined by plump pink epithelioid endothelial cells and a lymphoplasmacytic infiltrate which was admixed with numerous eosinophils, which were suggestive of angiolymphoid hyperplasia with eosinophilia [Table/Fig-2].



**[Table/Fig-1]:** Contrast MRI scan showing mass in the subcutaneous plane with diffuse homogenous contrast enhancements within and few flow voids along the superior and inferior margins



**[Table/Fig-2]:** High power view showing proliferating blood vessels lined by plump pink epithelioid endothelial cells and lymphoplasmacytic infiltrate admixed with numerous eosinophils (H&E X 40)

## DISCUSSION

Angiolymphoid hyperplasia with eosinophilia (ALHE) was first described by Wells and Whimster in 1960s, who reported 9 cases of persistent subcutaneous nodules in the head and neck, which had distinctive histological features [1]. It is a benign vasoproliferative entity with an undetermined pathogenesis, which has also been described as epithelioid haemangioma, histiocytoid haemangioma, papular angioplasia, pseudopyogenic granuloma and inflammatory arteriovenous haemangioma. Although ALHE and Kimura's disease were initially considered as variants of the same disease, it has been confirmed that they are distinct clinical entities which are categorized under eosinophilic dermatoses [2].

ALHE is a self limiting but a prolonged disorder with its distribution restricted to the head and neck region, with a predilection for the

external ear, especially the external auditory canal. It may rarely involve the orbit, lacrimal gland, oral mucosa and arm [3].

Clinically, it presents as itchy, solitary or multiple nodules which bleed easily, following mild trauma. The lesions vary from intradermal papules to subcutaneous nodules. Regional lymphadenopathy may be present, which may regress upon treatment of the primary lesion [4]. Pregnancy can exacerbate the nodular growth and trauma has been reported as an occasional preceding event [5]. Systemic eosinophilia is rare. Radiological evaluations such as MRI or angiography may be required to determine the extension of the lesion [2].

Histologically, it is characterized by the proliferation of vessels, a rich perivascular infiltrate of lymphocytes and plasma cells. The distinctive feature is the presence of a large nucleus and abundant eosinophilic cytoplasm in the endothelial lining of the blood vessels. The endothelial lining in ALHE is plump and epithelioid [6].

Immunohistochemistry which is done for B- and T-cell markers might show a diffuse T cell infiltrate with a prominent B cell follicle centre, which stains strongly with BCL-2 and MT2. Endothelial markers like Factor VIII and CD34 will show numerous blood vessels and their varying and irregular luminal calibres from muscular arterioles to capillaries, which permeate the lymphoid tissue [7].

Differential diagnosis which has to be considered for ALHE are Kimura's disease, extranodal lymphoma, pyogenic granuloma, insect bite and angiosarcoma [6]. Kimura's disease is usually generalized and often solitary lesions of more than 2 cms are commonly seen, with the presence of systemic lymphadenopathy and marked peripheral eosinophilia. ALHE is usually localized and often multiple, without the presence of systemic lymphadenopathy and peripheral eosinophilia. The characteristic histological endothelial lining of ALHE is absent in Kimura's disease, which is characterized by the presence of a flat endothelial lining. ALHE is a stage of histiocytoid or epithelioid haemangioma, which is a true vascular neoplasm, whereas Kimura's disease is a localized manifestation of a systemic inflammatory reaction [3].

Treatment modalities for ALHE nodules include intra-lesional and systemic steroids, curettage, simple surgical excision, cryotherapy,

laser excision, injection of cytotoxic drugs, micrographic surgery, phototherapy, alpha-2a interferon and radiation therapy. However, recurrences are quite common [6,8].

Recently, an interleukin-5 based therapy has been found to be effective. This interferes with the production and activation of eosinophils and it thereby stops their production, which is the key step in the pathogenesis of ALHE [9].

ALHE is a benign process with no tendency to metastasize and it can undergo remission spontaneously without any intervention [4].

## CONCLUSION

ALHE is a rare affliction of the head and neck which poses a diagnostic dilemma for the head and neck surgeons and dermatologists. It is often misdiagnosed as Kimura's disease and hence, it requires a careful histopathological examination. Treatment usually varies in different centres and recurrence of the lesion is common. Due to the presence of the lesion in the head and neck, care should be taken to provide cosmetics to the patient and adequate treatment should be administered to ensure complete remission of ALHE.

## REFERENCES

- [1] Wells GC, Whimster IW. Subcutaneous angiolymphoid hyperplasia with eosinophilia. *Br J Dermatol*. 1969;81:1-14.
- [2] Zaraq I, Mlika M, Chouk S, Chelly I, Mokni M, Zitouna M, Ben Osman A. Angiolymphoid hyperplasia with eosinophilia: A study of 7 cases. *Dermatology Online Journal*. 2011;17(2).
- [3] Ingrams DR, Stafford ND, Creagh TM. Angiolymphoid hyperplasia with eosinophilia. *J Laryngol Otol*. 1995;109:262-4.
- [4] Vallis RC, Garfield Davies D. Angiolymphoid hyperplasia with eosinophilia. *J Laryngol Otol*. 1988;102:100-01.
- [5] Don DM, Ishiyama A, Johnstone AK, Fu Y, Abemayor E. Angiolymphoid hyperplasia with eosinophilia and vascular tumors of the head and neck. *Am J Otolaryngol*. 1996;17(4):240-5.
- [6] Sharp JF, Rodgers MJC, MacGregor FB, Meehan CJ, McLaren K. Angiolymphoid hyperplasia with eosinophilia. *J Laryngol Otol*. 1990;104:977-9.
- [7] Backhouse SS, Shone GR, Douglas-Jones AG. Angiolymphoid hyperplasia with eosinophilia of the parapharyngeal space. *J Laryngol Otol*. 1998;112:802-4.
- [8] Shenefelt PD, Rinker M, Caradonna S. A case of angiolymphoid hyperplasia with eosinophilia treated with intralesional interferon alfa-2a. *Arch Dermatol*. 2000;136:837-9.
- [9] Braun-Falco M, Fischer S, Plotz SG, Ring J. Angiolymphoid hyperplasia with eosinophilia treated with anti-interleukin-5 antibody (mepolizumab). *Br J Dermatol*. 2004;151:1103-04.

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