

*Case Report*

## **Angiolymphoid Hyperplasia with Eosinophilia of the Nasal Cavity Masquerading Sinonasal Malignancy: An Unusual Presentation- A Case Report**

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### **ABSTRACT**

Angiolymphoid hyperplasia with eosinophilia is a rare benign vascular lesion with unclear etiopathogenesis. The lesions present as red to brown nodules, usually in head and neck region. We report a case of a 37 year old female with mass protruding from left nostril which clinically and on radiology mimicked malignant neoplasm. Left total maxillectomy was performed and histology revealed lobular proliferation of blood vessels lined by plump endothelial cells surrounded by mixed inflammatory infiltrate and lymphoid follicles suggestive of Angiolymphoid hyperplasia with eosinophilia.

**Key words-** Angiolymphoid, eosinophilia, vascular.

### **INTRODUCTION**

Angiolymphoid hyperplasia with eosinophilia (ALHE) is an uncommon benign vascular lesion with unclear etiopathogenetic mechanisms. This term was first proposed by Wells and Whimster in the year 1969. <sup>(1)</sup> On histology, ALHE shows numerous blood vessels lined by prominent endothelial cells with histiocytoid appearance. We describe a case where ALHE was masquerading as sinonasal malignancy clinically and on radiology. Histopathology established the correct diagnosis.

### **CASE REPORT**

A 37 year old female came with complaints of intermittent left side nasal block since 2 years, insidious in onset and gradually progressive in nature. Patient has h/o mass protruding from left nostril since

1 year and nasal bleed which was intermittent and spontaneous in nature for past 1 month. She underwent surgery for the similar complaints 1 year back and her symptoms improved. The histopathology of the lesion was reported as chronic sinusitis. She has normal hematologic and biochemical parameters. There was no eosinophilia on blood smear study. On examination, Anterior rhinoscopy showed hypertrophied polypoidal left inferior turbinate and a pale pink proliferative mass at middle turbinate. Radiology, plain and contrast CT study of paranasal sinuses and neck showed large ill defined mass lesion with heterogenous enhancement in left maxillary sinus measuring 6.6x 4.8x 5 cm. Lesion caused expansion and destruction of inferior, lateral, medial and posterior walls of left maxillary sinus (Fig

1,2,3). There were multiple bilateral enhancing lymph nodes at level II and V. These features were suggestive of malignant neoplasm in the region of left maxilla with lymph node metastasis. Following this left total maxillectomy was done. Grossly the partial maxillectomy specimen showed a nodular growth measuring 4.5x3 cm which on cut section showed grey white areas. On histology lobular proliferation of thick and thin walled blood vessels lined by plump endothelial cells, some with epithelioid appearance were seen. Intervening fibrocollagenous stroma showed scattered lymphocytes, eosinophils, plasma cells and neutrophils along with focal lymphoid follicle formation (Fig 4,5,6,7).

On immunohistochemistry (IHC), CD34 was positive in the endothelial cells. Ki-67 proliferating index was < 2% (Fig 8,9). A diagnosis of Angiolymphoid hyperplasia with eosinophilia was given. Patient is on follow up since 1 year and has no fresh complaints.

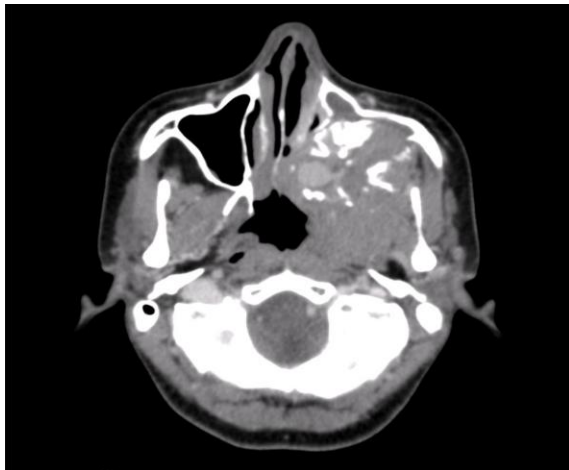


Fig 1 CT scan Axial view-Large ill defined mass lesion with heterogenous enhancement in left maxillary sinus



Fig 3 CT scan Saggital view- Large ill defined mass lesion with heterogenous enhancement in left maxillary sinus



Fig 2 CT scan Coronal View- Lesion caused expansion and destruction of inferior, lateral, medial and posterior walls of left maxillary sinus

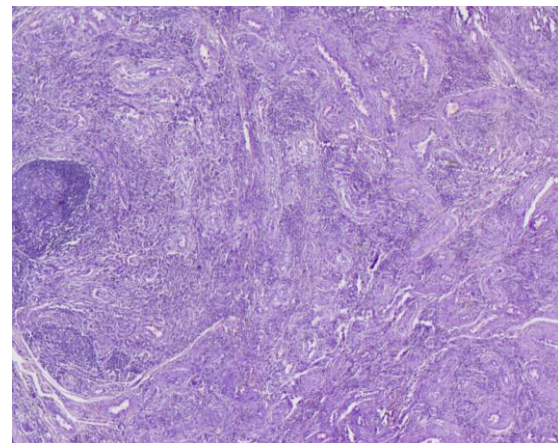


Fig 4- Lobular proliferation of blood vessels, inflammatory cells with focal lymphoid follicle formation H&E X20

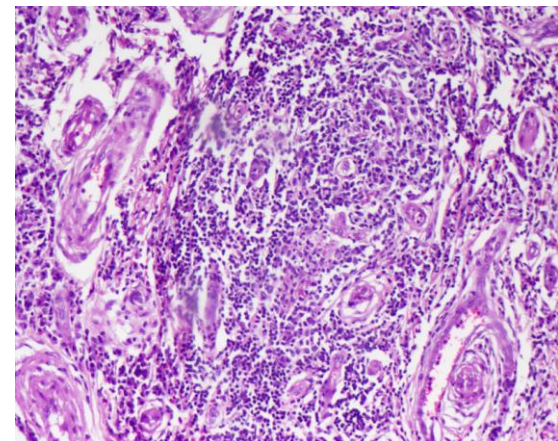


Fig 5- Stroma with lymphocytes, eosinophils, plasma cells and neutrophils H&E X100

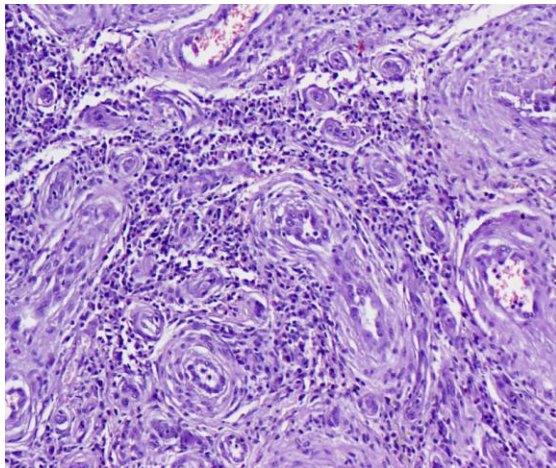


Fig 6- Thick and thin walled blood vessels H&E X100

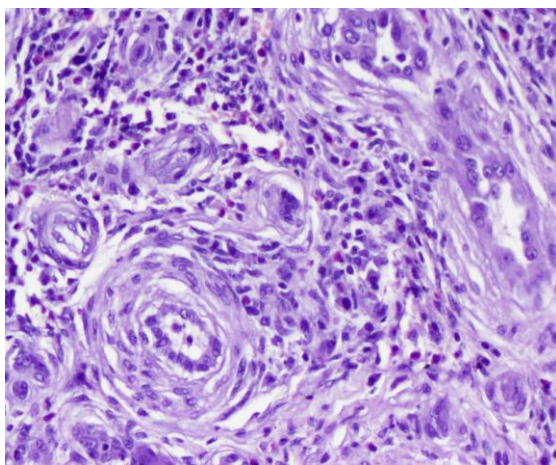


Fig 7- Plump endothelial cells with epithelioid appearance H&E X200

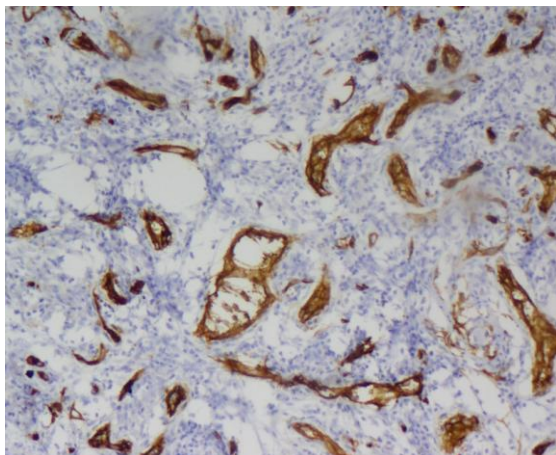


Fig 8- CD34 positivity in endothelial cells X100

## DISCUSSION

ALHE is a rare, benign however locally proliferating vascular lesion. Earlier been denoted with various names like epithelioid haemangioma, pseudopyogenic granuloma, inflammatory angiomatous nodule, papular angioplastia,

intravenous atypical vascular proliferation and histiocytoid haemangioma in literature. <sup>(2)</sup>

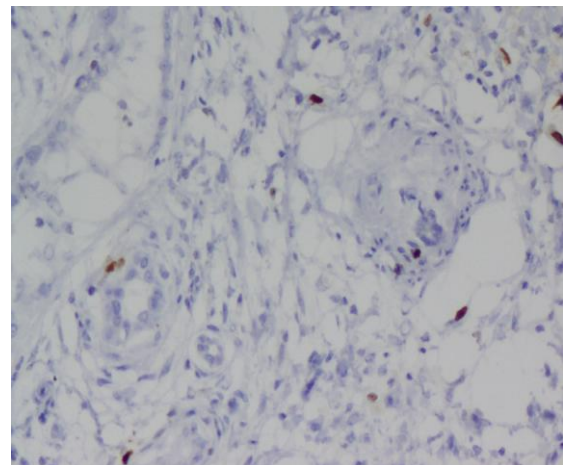


Fig 9- Ki-67 index <2% X200

The precise etiopathogenesis is yet to be established, however proposed mechanisms include a neoplastic process developing from endothelial cells, a hypersensitivity reaction, an inflammatory vascular reaction involving immunologic processes or a tissue reaction due to prior trauma. Hyperestrogenemia is been reported to enhance the growth. The distinctive inflammatory infiltrate seen in this lesion appears to be an intrinsic component. <sup>(3,4)</sup> A history of local trauma or infection is found in 9% of patients. <sup>(2)</sup> A case of ALHE developing after the diagnosis of peripheral T-cell lymphoma with T-cell gene rearrangements studies showing monoclonality in both the lymphoma and vascular lesions <sup>(5)</sup> has been described which supports the neoplastic pathogenetic hypothesis.

ALHE is commonly seen in Asian population and has a female preponderance. It presents in patients aged 20-50 years, with a mean onset of 30-33 years. <sup>(2,3)</sup> The lesion appears as dome-shaped, erythematous to brown, smooth-surfaced papules or nodules. It may be solitary in nearly 80% patients or present as multiple subcutaneous nodules. Lesions range between 1-10 cm, mostly fall within 0.5-2 cm diameter. <sup>(3)</sup> Lesions may be

asymptomatic, painful on palpation or even pruritic. Regional lymphadenopathy and arteriovenous shunts have occasionally been reported but are found to be inconsistent findings. <sup>(2)</sup> Systemic eosinophilia ranging from 6 to 34% is reported and seen in 20% of the cases, although eosinophilia is inconstant and is not mandatory to make the diagnosis of ALHE. <sup>(4)</sup> Our case had regional lymphadenopathy but no eosinophilia. Predilection for site is seen for the head and neck region especially around the ear, forehead or scalp which is seen in nearly 85% cases. However, ALHE has also been reported in the lip, tongue, orbits, conjunctiva, liver, spleen, heart, bone and blood vessels. <sup>(2,6,7)</sup>

ALHE on histology shows characteristic features, including prominent proliferation of small sized blood vessels lined by plump, epithelioid looking endothelial cells. The vessels typically have an immature appearance and may lack a well-defined lumen. These are however well formed vessels with single cell layer of endothelium and an intact smooth muscle layer. These distinctive endothelial cells have amphophilic or eosinophilic cytoplasm with a single large nucleus, open chromatin and a central nucleolus. A perivascular and interstitial infiltrate composed of lymphocytes, plasma cells and eosinophils is present. Eosinophils typically comprise 5-15% of an infiltrate. Rarely, these may comprise even 50% of the inflammatory infiltrate. Role of cytokines in this eosinophil rich inflammatory milieu of ALHE has been described. However, occasionally the infiltrate may be even devoid of eosinophils. Lymphoid aggregates with or without follicle formation are characteristic feature. <sup>(3,4)</sup>

Characteristic histologic features of ALHE help to distinguish it from other more aggressive vascular tumors. Epithelioid hemangioendothelioma

described as an intermediate-grade malignant vascular neoplasm can be distinguished by the appearance of its vessels. The vascular differentiation here is more primitive than seen in ALHE. The vessels are arranged in cords or nests without lumina compared to the lobulated organization seen in ALHE. Also the stroma here has a hyalinized or myxoid appearance, which is absent in ALHE. Epithelioid angiosarcoma is a rare high-grade malignant vascular tumor occurring in older male patients. It may be distinguished from ALHE primarily by its architecture, high mitotic activity, presence of necrosis and high degree of cytologic atypia. Also the epithelioid endothelial cells in this lesion almost always grow in large sheets and exhibit primitive branching vascular channels. Metastatic carcinomas are usually multifocal and have characteristic cytologic atypia associated with the carcinoma. <sup>(3,8)</sup> ALHE and Kimura's disease (KD), although share many common features but represent different entities. KD is a systemic disease and occurs as deep nodules or tumors with lymphadenopathy and a marked peripheral blood eosinophilia. On histology, it shows prominent cellular areas with lymphocytes forming follicles surrounded by an inflammatory infiltrate with an eosinophilia and fibrosis. <sup>(3,6)</sup>

The most successful and recommended treatment of ALHE is deep surgical excision. Other modalities include intralesional corticosteroids, oral retinoids, intralesional chemotherapy, radiotherapy, cryotherapy electrodesiccation, pulse-dye laser and carbon dioxide laser. <sup>(2,3)</sup> Rarely, the lesion may regress spontaneously. Malignant transformation almost never occurs. Recurrences documented in 30 percent cases after incomplete excision as the surgical margins in ALHE are difficult to determine. <sup>(4,9)</sup>

## CONCLUSION

ALHE is a rare, benign vascular tumor commonly seen in Asian population with female preponderance. It poses challenge in diagnosis and must be considered as an important differential diagnosis in head and neck tumors. Surgical removal is the treatment of choice. In spite of the benign course, it causes a therapeutic dilemma because it may be associated with cosmetic defects.

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