Otolaryngology online journal

ISSN: 2250-0359 Volume 5 Issue 1 2015

ANGIOLYMPHOID HYPERPLASIA

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Abstract

Angiolymphoid hyperplasia with eosinophilia (ALHE) is an uncommon benign idiopathic condition that manifests in adults, characterized by isolated or grouped papules, plaques, or nodules in the skin of the head and neck. Most patients present with lesions in the periauricular region, scalp or forehead. The other rare sites of involvement include the hands, shoulders, breasts, penis, oral mucosa, orbit and scrotum. It is a distinct histopathologic entity, marked by a proliferation of blood vessels with distinctive large endothelial cells, accompanied by a characteristic inflammatory infiltrate that includes eosinophils. Though a few cases of nephropathy have been reported in patients with ALHE there is no strong association. This is in contrast to the related entity, Kimura disease, for which the association with nephrotic syndrome is strong. We present a rare occurance of ALH in a 35 year old post renal transplant male patient which has not been reported so far.

Key words :

angiolymphoid hyperplasia, renal transplant, unilateral

Introduction :

ALHE was originally described by Wells and Whimster[1] in 1969 .They considered the process to be a late stage of Kimura's disease – a disorder described in the Japanese literature 20 years earlier [2] . While ALHE shows some similarity to Kimura disease, it is generally regarded as a separate entity. While lesions are superficial in ALHE, involvement is deeper in Kimura disease such as lymph nodes, salivary glands, and the subcutis. With the advancement of staining techniques , various new differences between the two entities have been unraveled. The condition (ALHE) is clinically characterized by erythematous or skin coloured dome shaped dermal papules or nodules, often associated with spontaneous bleeding, pain, pulsation, pruritus and growth. It has a predilection for head and neck area, especially for the ears. Eosinophilia is present in 20% of cases and lymphadenopathy is uncommon .It is most commonly seen in women in their third to fourth decade. The exact etiology of ALHE is not known. We report a case of ALHE in a post renal transplant patient, for its rare association.

Case Report :

A 35 year old man presented with mildly pruritic, painless papulo-nodules over the back of his left ear of six months duration. It was associated with mild bleeding on scratching which spontaneously stopped. There was no history of trauma prior to the onset of the lesion. He had undergone renal transplantation a year ago and is on systemic tacrolimus. The dermatalogical examination showed multiple erythematous, hyperpigmented, nontender, domeshaped, firm nodules which coalesced to form a plaque of size of 7cm x 4cm over the left retroauricular region (Fig 1). Few discrete papules were seen at the summit of the nodular

plaque. It was warm, non-pulsatile and no bruit was heard. On puncturing the nodule, there was minimal ooze of blood. The regional nodes, salivary glands and other integuments were not involved. Systemic examination was normal. Based on morphology of the lesion, clinical diagnosis of ALHE, Bacilliary angiomatosis and Kaposi's sarcoma were thought of. On investigation, complete haemogram was normal with no peripheral eosinophilia. Mantoux test, screening for HIV were negative. Skiagram chest, USG abdomen was normal. The excision skin biopsy of one discrete papulonodule revealed numerous dilated blood vessels lined with plump endothelial cells resembling epitheloid cells. Heavy infiltrate of mononuclear cells, epitheloid cells especially in mid and lower dermis was seen. There was extravasation of RBCs in one region (Fig 2 , 3) and no eosinophils were seen. Hence the histopathology was aniolymphoid hyperplasia (ALH) instead of angiolymphoid hyperplasia with eosinophilia (ALHE). We could not do Warthin Starry stain due to lack of resources. A therapeutic trial with erythromycin, doxycycline was instituted, with no regression of the lesions. The absence of neutrophils & cytoclasia ruled out bacillary angiomatosis. The differential diagnosis of Kaposi sarcoma was ruled out due to lack of slit like spaces and spindle cells histologically and no systemic disturbances. No mitotic figures and atypicality of endothelial cells were observed to diagnose angiosarcoma. The other histological differential diagnosis for vascular proliferation like pyogenic granuloma (epithelial collarette , lobules of dilated and congested capillaries within myxoid stroma), epitheloid hemangio endothelioma (obvious vascular channels and cells arranged in Indian file pattern in a mucoid stroma), benign angioma or ectasia (inflammatory cell infiltrate) were ruled out. Our final diagnosis based on clinical features (unilateral, intradermal papulonodules with no regional lymphnodes and salivary gland involvement) and histological

findings (hyperplasia of small blood vessels lined by epitheloid endothelial cells surrounded by dense lymphohistiocytic infiltrate. Patient is being treated with CO_2 Laser and he is improving.

Discussion :

ALHE is a benign proliferating lesion composed of vascular channels lined by endothelial cells with abundant pink cytoplasm and vesicular nuclei. The terms epitheloid hemangioma and pseudo-pyogenic granuloma have been used as synonymously . The cause is unknown. But more studies suggest a reactive process. Some regard ALHE essentially as a malformation of blood vessels caused by an underlying AV shunt [3]. Affected individual are commonly young adults who present with a cluster of small translucent, pruritic papules, nodules and plaques at or around the external ear. The lesions can be limited to one side. Occlusion of the external auditory canal can bring patient to the attention of ENT specialist. Spontaneous regression is seen in majority cases after a variable period of time. ALHE may also rarely occur in the oral cavity, lymph nodes, bone, testis, ovarian teratoma [4], lung [5] and orbit [6]. Association with Hepatits C antibodies[7] and follicular mucinosis[8] have been reported. Kimura's disease, now a distinct entity is a subcutaneous variant. Differences between them have been illustrated in the table. Exceptionally, ALHE may co-exist with Kimura's disease [9].

Histopathologically, in ALHE, dermis and/or subcutis exhibit hyperplasia of small blood vessels lined by plump endothelial cells which line and protrude into lumina of blood vessels resulting in 'tombstone' appearance surrounded by dense infiltration of lymphocytes, plasma cells, histiocytes and eosinophils . Eosinophils are typically prominent, but absent in some cases[10]. Thus we labelled our case as angiolymphoid hyperplasia rather as ALHE. The cause of ALH is unknown in our case, however scratching of the ear might have led to a reactive vascular proliferation or the persistent antigenemia in immunosuppression should have played a

role[11]. Recently, it has been hypothesized that the pathology may represent a T cell lymphoproliferative disorder of benign or low-grade malignant nature[12]. Also malignant transformation has been observed in a young patient with ALHE who developed peripheral T-cell lymphoma [13]. Multiple treatment optionshave been reportd to be effective, including curettage and desiccation, cryotherapy, radiotherapy, steroids (oral, topical, and intralesioal) , retinoids,excision, various lasers [14].

The role of immunosuppressive therapy becomes inevitable following organ transplantation, for which the benefit of preventing graft rejection, against the risk of their adverse effects. The long term consequences of immunosuppressive therapy like predisposition to infections, metabolic effects, cutaneous effects and development of benign and malignant tumours like seborrhoeis keratosis, actinic keratosis, squamous cell carcinoma, basal cell epithelioma, Kaposi's sarcom, bacillary angiomatosis, malignant melanoma, Merkel cell carcinoma have frequently been seen and reported. The unusual occurrence of ALH in our patient post renal transplantation might be of the possible etiology of a reactive vascular proliferation secondary to epithelial damage or a persistent antigenemia due to immunosuppression.

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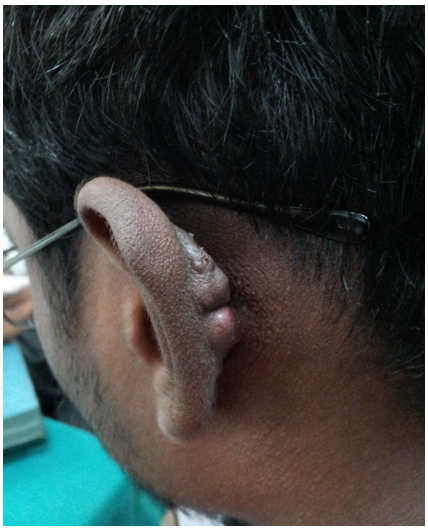

Fig 1

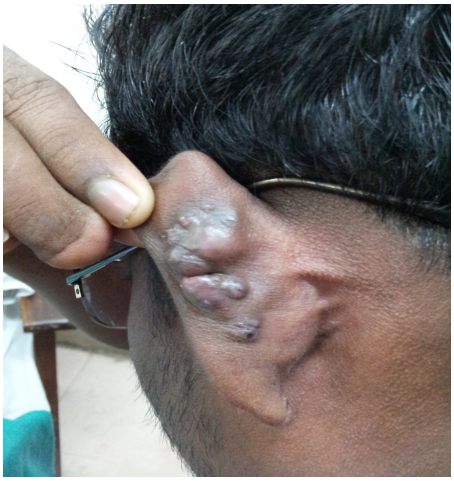

Fig 2

Auricle shaped Erythematous pigmented nodular plaque with dome shapedpapules at summit. Puncture mark at the lower end of the lesion.

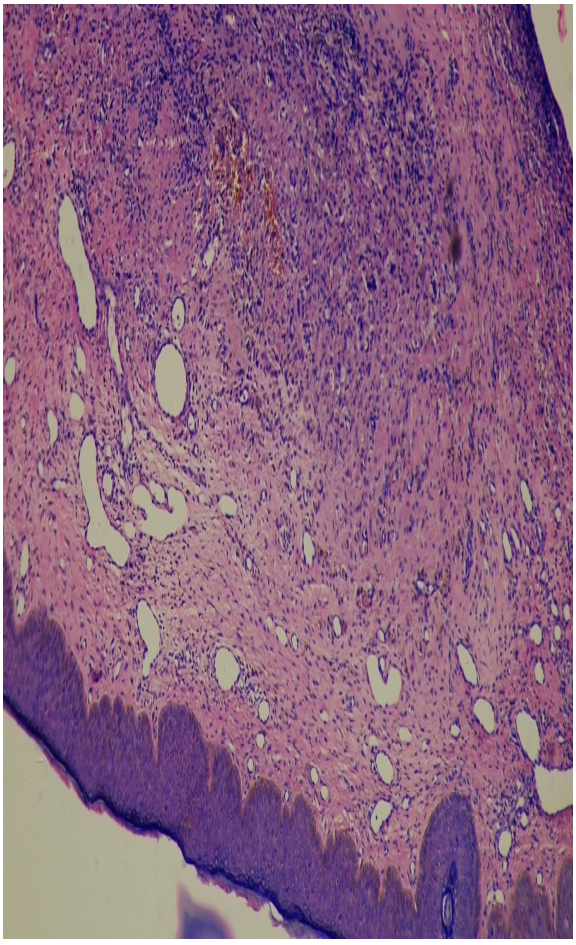

Figure 3: Psoriasiform acanthosis with prominent endothelial cells, lymphohistoid infiltrate. (H and E, ×10)

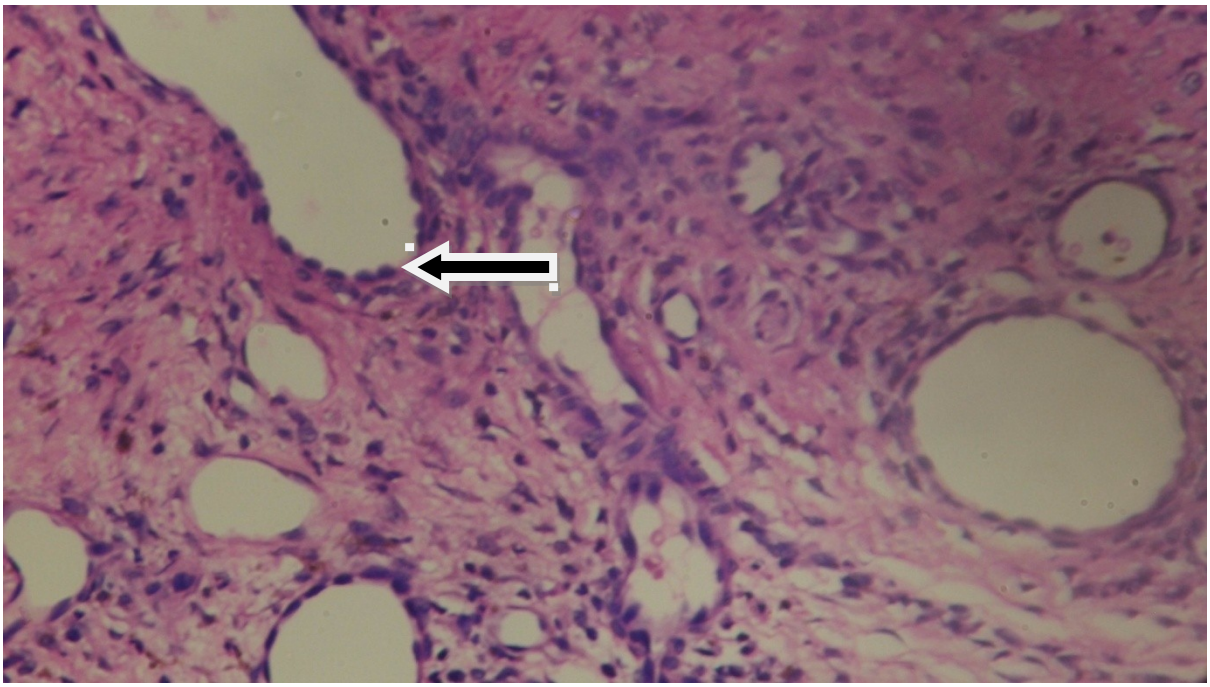

Figure 4: Tombstone appearance of endothelial cells (H and E, ×40)

Differences and similarities between Kimura's disease and angiolymphoid hyperplasia with eosinophilia

Features	Kimura's disease	Angiolymphoid hyperplasia with eosinophilia
CLINICAL:		
Race	Oriental	Any
Sex	Males	Males and females
Age	20-40 yr	All ages
Lesion	Subcutaneous mass	Dermal papulonodules
Location	Head, neck	Multiple locations
Pruritis	No	May be severe
Lymphadenopathy	Always	Rare
Glomerulonephritis	Occasional	Rare
HISTOLOGY:		
Depth	Subcutaneous, muscle	Dermis, subcutaneous
Infiltrate	Nodular	More diffuse
Germinal centres	Always	Uncommon
Fibrosis	Usually marked	Mild
Eosinophilia	Massive	Mild-marked
Edema	Often marked	Minimal
Blood vessels	Well formed	Variable
Endothelial cytology	Occasional atypical	Often atypical
IMMUNOHISTOCHEMISTRY		
Lymphoid markers	T and B cells in lymphoid follicles	Variable pattern
INFLAMMATORY CELL GRANULE PROTEINS		
Eosinophill MBP and ECP	IC	IC and EC
Mast cell tryptase	IC	IC
Neutrophil elastase	IC	IC

[EC- Extracellular staining pattern; ECP-Eosinophil cationic pattern; IC- Intracellular staining pattern; MBP- Major basic protein]

[Reference : Journal of cutaneous pathology, Vol 22 No.4, August 1995, pg 319-326]