

Asymptomatic nodules over ear in a young female

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This is a case of a 22-year old female who presented to outpatient department with the complaint of swelling over her right ear since 2 years associated with itching. Lesion started over the right ear lobule gradually progressed upwards involving the antihelix. On enquiring about history of trauma prior to the onset of lesions, patient gave history of ear piercing 3 years back. There is no history of any ear discharge, hearing difficulty, neither was there any complaints of oozing, scaling, pus discharge, pain, or bleeding from the lesion. Patient cannot recollect any history of insect bite, fever, or chronic cough. There is no history suggestive of keloidal tendencies in the past or in the family. On examination, there is a solitary erythematous nodule over right ear lobule extending upwards to cover lower part of helix, measuring 3x 3 cm, firm in consistency, with no tenderness or bleeding on palpation (**Figure 1 and 2**). Skin over the lesion is fixed and was not pinchable and two piercings are present over the ear lobule as “prima facie” of the trauma probably leading to the growth. There was no involvement of external auditory canal, or enlargement of cervical lymph nodes, and the respiratory system as well as ear, nose and throat

is found to be normal. A clinical differential of keloid, lobomycosis, deep fungal infection and cutaneous tuberculosis was kept. Complete blood count, liver function test, chest X-ray does not show any abnormality. Mantoux test is found to be negative. Biopsy for histopathology stained with hematoxylin and eosin reveals keratinized epithelium overlying dermal nodules composed of proliferating capillaries, lined by plump endothelial cells, surrounded by dense infiltrate of lymphocytes, few histiocytes, eosinophils, plasma cells forming lymphoid aggregates (**Figure 3-5**).

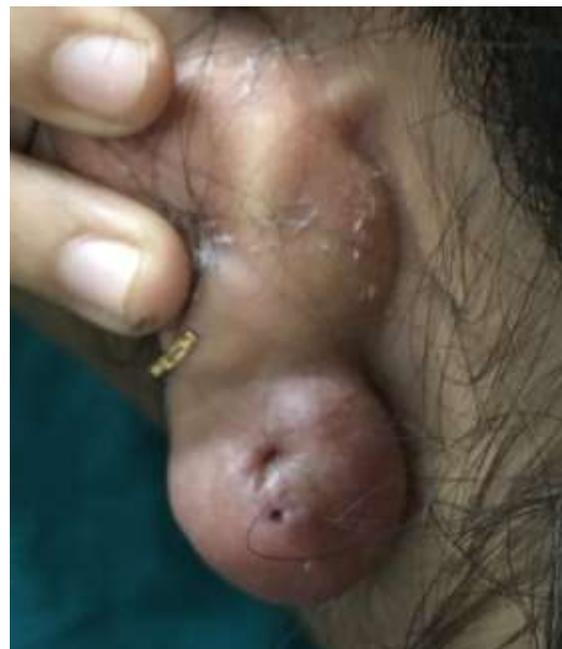


Figure 1 Single erythematous nodule measuring 3 X 3 cm over lower pole of right ear.

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Figure 2 Anterior aspect of the lesion, nodularity extending on the tip of lobule.

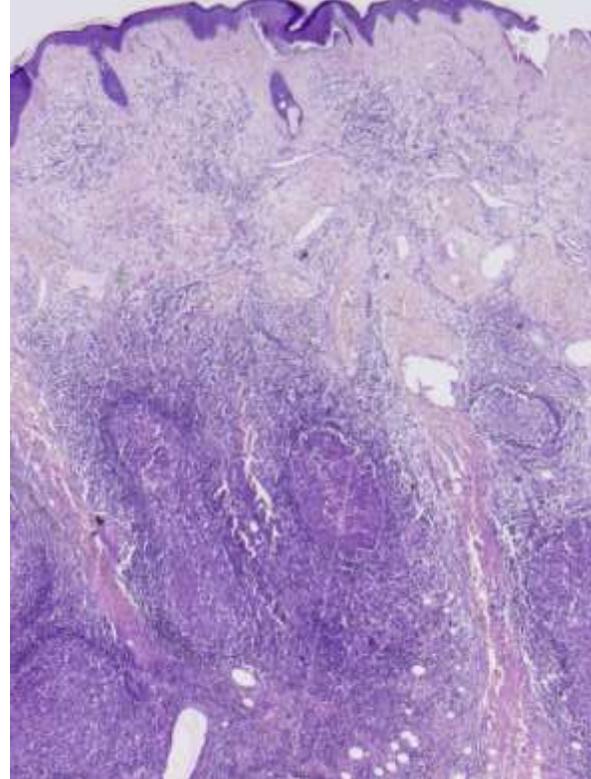


Figure 3 HPE 200X – keratinized epithelium overlying dermal nodules.

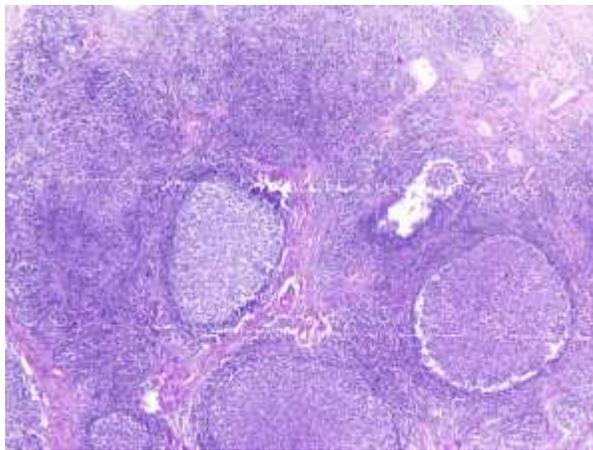


Figure 4 HPE 400X – dermal nodules composed of blood vessels lined by plump endothelial cells and surrounded by lymphocytes, histiocytes, eosinophils.

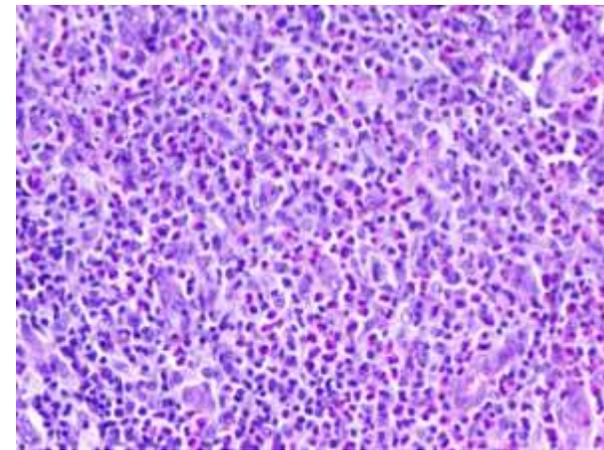


Figure 5 HPE 1000X – infiltrate surrounding the nodules composed of eosinophils, histiocytes and lymphocytes.

What is the diagnosis?

Diagnosis

Angiolymphoid hyperplasia with eosinophilia.

Discussion

Angiolymphoid hyperplasia with eosinophilia is an uncommon benign or reactive angioproliferative lesion presenting with cutaneous and less often with extracutaneous manifestation. It is more commonly seen in Caucasian females in 3rd to 5th decade of life, with a predilection for head and neck area especially auricular area, appearing as erythematous to violaceous firm nodules varying in size from 2-3 cm in diameter. It was first described by Whim and Whimster in 1969, they were also the ones who linked ALHE to Kimura's disease, though the association is now controversial and not certain.¹ ALHE has also been labelled by various other names, pseudopyogenic granuloma, atypical pyogenic granuloma, papular angioplasia, epitheloid hemangioma. Ackerman and colleagues proposed to replace the name with 'angiolymphoid hyperplasia with eosinophils' instead of eosinophilia, which appears to be quite substantiated by the fact that only 20% of patients with ALHE actually show blood eosinophilia while eosinophils in histopathology is a characteristic feature.²

Origin or pathogenesis of ALHE is still a mystery, several theories have been proposed. One school of thoughts proposes it to be a neoplasia or vascular malformation while others consider it to be proliferative change in response to various triggers like trauma, insect bite, infections (association with HHV8 has been mentioned) and hormones. ALHE is now classified under reactive vascular hyperplasias along with pyogenic granuloma and bacillary angiomatosis. Association of atopy and ALHE has been made recently, especially on the basis

of discovery of reticulated IgE pattern observed on immunostaining in ALHE patients. Association of ALHE to underlying renal glomerulonephritis, cryoglobulins, undifferentiated connective tissue disorders has been proposed.^{2,3}

Histopathology of ALHE demonstrates both vascular and inflammatory component, with proliferation of dilated small to medium sized capillaries in the dermis amidst mixed infiltrate of eosinophils, plasma cells, histiocytes, and lymphocytes. Blood vessels appear tubular or elongated lined by plump, hobnail shaped endothelial cells. The plump endothelial cells protruding into the lumen of blood vessels provides a 'tombstone' appearance. Kimuras disease bears a lot of similarity to ALHE histologically, both showing angiomatous proliferation with surrounding infiltrate rich in eosinophils, but Kimuras disease can be differentiated by presence of large number of germinal follicles with lymphocytic infiltration whereas ALHE characteristically shows histiocytoid shaped endothelial cells which are rarely seen in Kimuras disease. Clinically, Kimuras presents as subcutaneous swelling in neck rather than erythematous papules or nodules akin to ALHE and peripheral eosinophilia is seen in almost all patients of Kimura's disease which is seen in only 20% of ALHE patients. Immunohistochemical analysis reveals abundance of B cells in Kimuras with presence of IgE in germinal follicles whereas ALHE shows T cell predominance.^{3,4}

Treatment of ALHE mainly relies on physical modalities or surgical excision, though recurrences are common. Topical or intralesional steroids, cryotherapy, radiofrequency, ablative laser, imiquimod have all been tried with variable rates of success. Few cases of spontaneous resolution have also been reported.

Key features of angiolymphoid hyperplasia

- Benign or reactive angioproliferative lesion.
- Common in Caucasian females in 3rd to 5th decade of life.
- Manifest as erythematous to violaceous firm nodules.
- Localization - predilection for head and neck area especially auricular area.
- Investigations - 20% of patients with ALHE actually show blood eosinophilia while eosinophils in histopathology is a characteristic feature.
- HPE shows vascular (proliferation of blood vessels lined by hobnail appearance/ 'tombstone' appearance) with inflammatory component (vessels surrounded by eosinophils (predominantly), lymphocytes, histiocytes and plasma cells.

References

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