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Case Report

Angiolymphoid hyperplasia with eosinophilia: improvement in a recurrent case with isotretinoin

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Abstract Angiolymphoid hyperplasia with eosinophilia (ALHE), characterized by dermal and subcutaneous nodules in head and neck areas, is often very disfiguring. There are different modalities of treatment but surgery seems to be the best. We report a 31-year-old man with recurrent ALHE showing considerable improvement with isotretinoin.

Key words
Angiolymphoid hyperplasia with eosinophilia, isotretinoin.

Introduction

Angiolymphoid hyperplasia with eosinophilia (ALHE) is a benign but potentially disfiguring vascular lesion. It is usually characterized by dermal and subcutaneous nodules, primarily in the head and neck region. Mostly lesions are present in periauricular region,[1] forehead or scalp. Spontaneous regression is common, but persistent lesions may require treatment. Surgery seems to be the best modality of treatment but often may be disfiguring and difficult in periauricular region. Often the lesions are recurrent and may require several treatments.

We report a 31-year-old man presenting with multiple nodules on the cheeks, preauricular region since last 3 years. The lesions which were recurring after intralesional injections of corticosteroid and surgical excision showed considerable improvement with systemic isotretinoin.

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Case report

A 31-year-old male presented with a 3-year history of multiple, asymptomatic, smooth, dome-shaped, dull erythematous to bluish compressible, soft, papulonodular lesions of 5-25 mm size, grouped in and around the left ear (**Figures 1**). A few of the lesions were pedunculated. The overlying skin was intact. The lesions had developed spontaneously. General health of the patient was unaffected. There was no regional lymphadenopathy. Routine laboratory tests were normal. There was no peripheral eosinophilia. The IgE level was normal. Excision of the lesions attempted twice in the past led to recurrence within a period of about 2 months. Intralesional injections of

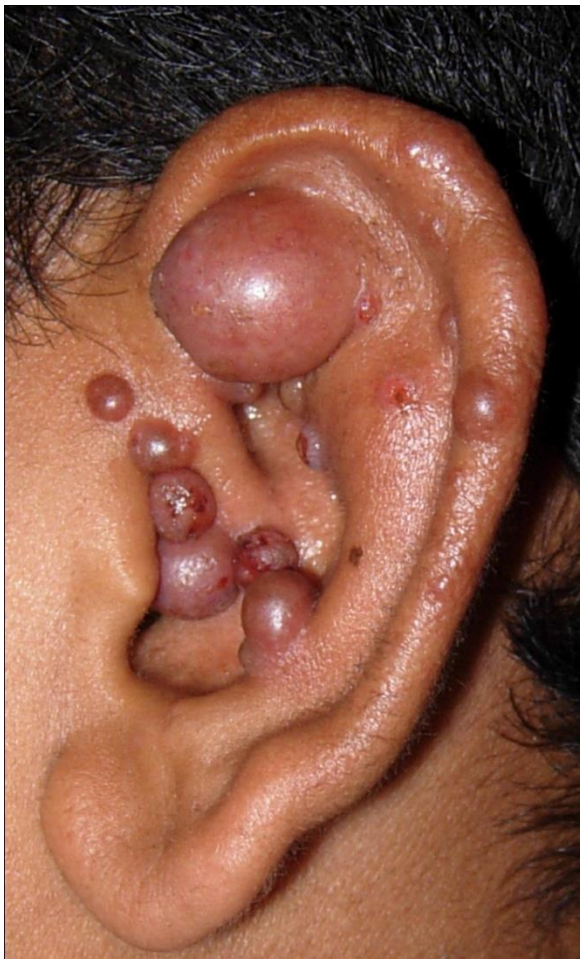

Figure 1 Dome-shaped, dull erythematous, papulonodular lesions of 5-25 mm size, grouped in and around left ear.

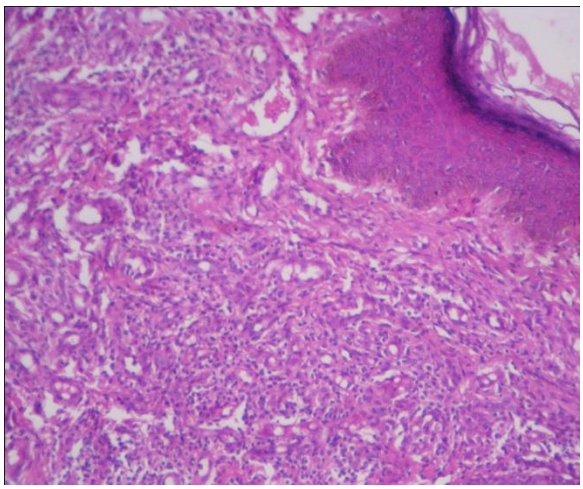

Figure 2 Polymorphous inflammatory infiltrate rich in lymphocytes and eosinophils in the dermis with prominent blood vessels lined by plump like histiocytic endothelial cells (H and E, X40).

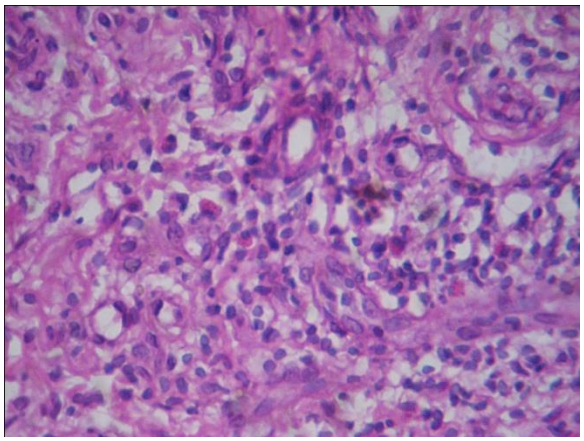

Figure 3 Histological picture under further magnification (H & E, X 100).

triamcinolone were given thrice with same sequele. Histology of one of the lesions showed abundant proliferating blood vessels with prominent plump like histiocytic endothelial cells. There were lymphocytic and eosinophilic infiltrates (**Figures 2** and **3**). The clinical features as well as histology were consistent with diagnosis of ALHE.

Patient's weight was 62 kg and he was put on tablet isotretinoin 40mg (approximately 0.75 mg/kg body weight) daily with considerable improvement after 1 month. His fasting lipid profile was normal after 1 month and the prescribed medications were continued for one more month. Then it was decreased to 20 mg per day. After two months of further treatment, the lesions had almost subsided..

Discussion

The pathogenesis of ALHE is unknown. While some thought ALHE to be a benign tumor, numerous factors suggest that it is an unusual reactive process.[2] Improvement after topical or intralesional steroids (as in our case earlier on) also speaks against its malignant etiology. ALHE has occurred following various forms of trauma or infection. Histologically, most cases

Table 1 Differences between angiolymphoid hyperplasia with eosinophilia and Kimura's disease.

	Angiolymphoid hyperplasia with eosinophilia	*Kimura's disease*
Presentation	Superficial papules or nodules Multiple lesions	Large subcutaneous nodules Usually one lesion
Population	Older age Caucasian Female	Younger age Asian Male
Duration	Shorter	Longer
Regional lymph nodes	No	Possible
Blood eosinophilia	Mostly absent	++
Elevated serum IgE	Infrequent (20% cases)	Frequent
Blood vessels	Thick walled and concentric with prominent plump 'histiocytiod' endothelial cells	Thin walled
Eosinophils in tissue	+	++
Lymphoid follicles	-/+	+
Origin of the disease	Vascular origin (endothelial cell)	Chronic inflammatory process

of ALHE show damaged and/or tortuous arteries and veins at the base of the lesion, suggesting that arteriovenous shunting may play a role in the pathogenesis.

Differential diagnoses of ALHE include granuloma faciale, persistent insect bite reaction, injection site granuloma, cutaneous lymphoma, cavernous hemangioma, pyogenic granuloma, angiomatous lymphoid hamartoma, pseudolymphoma (lymphocytic infiltrate of Jessner, lymphocytoma cutis), sarcoidosis and bacillary angiomatosis. ALHE and Kimura's disease were earlier considered to be same. Features common to both include predilection for head and neck and infiltration in dermis by lymphocytes and eosinophils. But now, they are thought to be separate clinicopathological entities.[3] The differences between the two entities are shown in **Table 1**.

Patients with solitary or few small lesions may benefit from excision or Mohs' surgery.[4] Surgery may be difficult, especially in the case of periauricular lesions. Here radiotherapy, curettage, shave excision with electrodessication, cryotherapy, are good options. Different other modalities are systemic and intralesional steroid administration, interferon therapy, cryotherapy, topical application of tacrolimus, imiquimod[5] and laser therapy.[6] The pulsed dye laser has an edge over continuous-wave carbon dioxide and argon lasers as it does not cause post-treatment scarring.

There are few reports of treatment with systemic retinoids in the literature. There was complete resolution of scalp nodules after 4 months of treatment with acitretin (1 mg/kg/day).[7] There is a report of recurrence when isotretinoin was used for short period of two months.[8] However, isotretinoin was used successfully when it was used for long duration.[9,10] So we used a trial of isotretinoin 40mg per day (0.75mg/kg body weight) with considerable improvement after 2

months. Then we tapered the dose to 20 mg per day with almost subsidence of the lesions in 4 months of starting treatment. We planned to continue the dose for further 2 months and then further reduce the dose to 20 mg alternate day for at least more 6-8months.

In our case, isotretinoin was used successfully to control the disease and lesions completely subsided. This effect may be due to the capacity of retinoids to inhibit angiogenesis. Retinoids inhibit the anti-activator protein 1 (AP1) pathway by down-regulating the expression of vascular endothelial growth factor (VEGF) genes.[11]

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