What's your diagnosis?

Erythematous nodules of the ear and the retro-auricular area in a Moroccan woman. What's your diagnosis?

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CASE REPORT

A 54-year-old Moroccan patient, without any notable pathological history, who consulted us for slightly pruritic lesions on the scalp and periauricular area evolving for 2 years.

At our examination, we noted violaceous polypoid nodules at the number of 20, of different sizes ranging from 2 mm to 5 mm, with a slightly infiltrated base located at the auricular, retro auricular and left temporal level (Fig. 1). Salivary glands and cervical lymph nodes examination were free and no other lesions throughout the body were seen. Dermoscopic examination showed a pale pink background with some dotted vessels (Fig. 2). We performed an excisional biopsy of a nodule which revealed a moderately dense dermal mixed inflammatory infiltrate of lymphocytes with many eosinophils (Fig. 3a). Prominent vascular proliferation, the dermal vessels are lined by plump endothelial cells (cobblestone appearance) (Fig. 3b). The epidermis demonstrated irregular hyperplasia and hyperkeratosis with no atypia. A peripheral blood sample showed hyper eosinophilia.

WHAT'S YOUR DIAGNOSIS?

Angio-Lymphoid Hyperplasia With Eosinophilia (ALHE)

Angio-lymphoid hyperplasia with eosinophilia (ALHE) is a benign vascular tumor proliferation, first described in 1969 by Wells and Whimster. It, etiology, remains unknown until now. Some authors consider ALHE as a benign neoplastic disease, others still consider it as a low-grade neoplasm owing to its progressive nature, high rate of local recurrence while others think that traumatic stimuli and hormonal factors (pregnancy) can produce the disease [1]. ALHE occurs most often in young or middle-aged adults, and more frequently in women, giving rise to theories that estrogen sensitivity is a component of pathogenesis. More frequent than Kimura disease, of which it is the main differential diagnosis [2]. It may present as one or multiple erythematous, purple, angiomatous papules, nodules or plaques with a predilection for the head and neck region, especially the auricular area. Other rarer localizations have been described, such as the upper limbs and the buttocks, trunk, lower extremities, penis and oral mucosa. Generally, these lesions are asymptomatic, sometimes they can be pruritic or painful, crusty and bleeding [3]. Dermoscopy may show a polymorphic vascular pattern composed of irregular, linear and dotted vessels, evenly distributed on a pale reddish to pale pink background [4]. Histological examination of ALHE shows well-defined lesions, mainly confined to the dermis, but sometimes also in the hypodermis. The lesions show an abundant proliferation of small capillary-sized vessels that are typically lined with epithelioid endothelial cells, with a characteristic (cobblestone appearance) and an inflammatory infiltrate consisting mainly of eosinophils [5]. Blood eosinophilia is noted in 10-20% of cases. While the IgE level is often normal [1]. The main differential diagnosis is Kimura's disease, an entity long confused with HALE but now recognized as quite distinct. It is rarer than HALE and prefers to affect males of Asian descent with a history of atopy. Clinically, it presents as one or more

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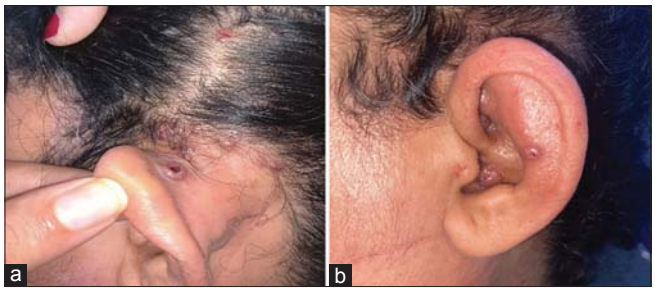

Figure 1: (a and b) Clinical image, showing multiple erythematous angiomatous nodules in the ear and the the retro-auricular area.

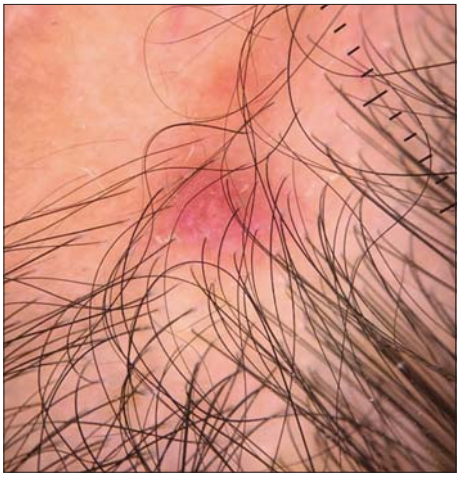

Figure 2: Dermoscopic image showing a pale pink background with some dotted vessels

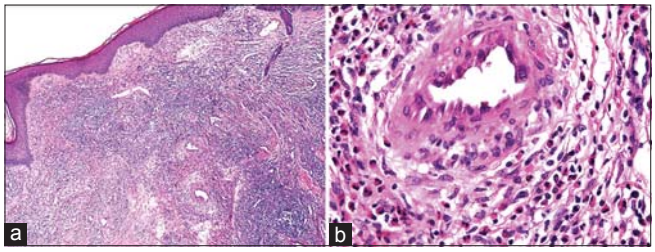

Figure 3: (a) Histological picture showing, vascular hyperplasia of variable size in the dermis + polymorphic inflammatory infiltrate rich in eosinophils. (b) Histological picture showing endothelial cells protruding like (cobblestone appearance) in endoluminal + Inflammatory infiltrate rich in eosinophils.

deep cutaneous or subcutaneous nodules located in the cervicofacial region, often with locoregional adenopathy and extension to the salivary glands. The biology shows a hypereosinophilia and an elevation of total IgE. Renal involvement with proteinuria is associated in 10 to 60% of cases. Histologically, there is a lymphocytic infiltrate with lymphoid follicles between which there are numerous eosinophils, as well as venous hyperplasia. Many approaches to treat ALHE have been tried, such as steroids (local and systemic), methotrexate, pentoxifylline, propranolol, isotretinoin, cryotherapy, surgical excision, as well as newer cytokine-targeted therapies including topical imiquimod, interleukin-5, and interferon alpha-2a. Laser therapy has also been used to treat ALHE lesions such as pulsed dye laser, carbon dioxide laser, and argon laser [6]. Although treatment failure rates are high for all modalities, surgical excision is the most effective treatment. Further research is needed on the origin and treatment of ALHE to have a codified and optimal management [3].

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

REFERENCES

1. Adler BL, Krausz AE, Minuti A, Silverberg JI, Lev-Tov H. Epidemiology and treatment of angiolymphoid hyperplasia with eosinophilia (ALHE): A systematic review. J Am Acad Dermatol. 2016;74:506-512.e11.
2. Bahloul E, Amouri M, Charfi S, Boudawara O, Mnif H, Boudawara T, et al. Angiolymphoid hyperplasia with eosinophilia: report of nine cases. Int J Dermatol. 2017;56:1373-8.
3. Menicanti C, Nazzaro G, Cavalli R, Paolucci A, Berti E, Passoni E. Transarterial embolization and surgical excision as treatment for angiolymphoid hyperplasia with eosinophilia. Dermatol Ther. 2020;33:e14138.
4. Rodríguez-Lomba E, Avilés-Izquierdo JA, Molina-López I, Parra-Blanco V, Lázaro-Ochaita P, Suárez-Fernández R. Dermoscopic features in 2 cases of angiolymphoid hyperplasia with eosinophilia. J Am Acad Dermatol. 2016;75:e19-e21.
5. Sagi L, Halachmi S, Levi A, Amitai DB, Enk CD, Lapidoth M. Combined pulsed dye and CO2 lasers in the treatment of angiolymphoid hyperplasia with eosinophilia. Lasers Med Sci. 2016;31:1093-6.
6. Baghestani S, Firooz A, Ghazisaidi MR. A refractory case of angiolymphoid hyperplasia with eosinophilia successfully treated by surgery. J Dermatolog Treat. 2011;22:49-51.

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