

An Unusual Cause of Papules on the Face

Dear Editor,

Angiolymphoid hyperplasia with eosinophilia (ALHE) is a rare, locally proliferating disorder that affects predominantly the head and neck region (1,2). There seems to be a higher incidence in middle-aged Caucasian women (2,3).

A 28-year-old female patient with no relevant personal or family medical history and only taking an oral contraceptive, presented to our department with multiple, well delimited, infracentimetric erythematous papules with a smooth surface on the left frontal, temporal, and preauricular regions (Figure 1). The lesions had appeared 7 months earlier, with progressive growth in number and dimensions since. The patient reported pruritus and denied previous trauma, topical application of any sort, insect bite at these locations, and any other accompanying symptoms. A thorough physical examination revealed no additional abnormalities. An excisional biopsy of one of the left temporal papules revealed a prominent lymphoid component, with a dense multinodular in-



Figure 1. Clinical aspect of the lesions: well delimited, erythematous, tumid papules in the left frontal and temporal regions.

filtrate in the superficial and deep dermis, with reactive germinative centers of considerable dimensions (Figure 2). Large and atypical lymphocytes were confined to the germinative centers, with reactive characteristics. Lymphocytes surrounding the germinative centers were predominantly small, accompanied by a significant number of scattered eosinophils. CD3 and CD20 immunohistochemical staining revealed B-cells predominantly in the nodular areas corresponding to the germinative centers, while T-cells displayed a diffuse peripheral distribution. There was severe neovascularization, with thick-walled vascular channels lined by enlarged plump endothelial cells with an "epithelioid" appearance. These findings supported the diagnosis of angiolymphoid hyperplasia with eosinophilia (ALHE). Laboratory workup did not show any abnormalities, including eosinophilia or elevation of immunoglobulin E levels. Due to pruritus and aesthetic concerns, surgical excision of the larger and most symptomatic papules was performed. The patient was assured of the benign nature of the disease and informed about the possible development of new lesions. Kept under clinical surveillance, the patient remained free of new lesions at 6-month follow-up.

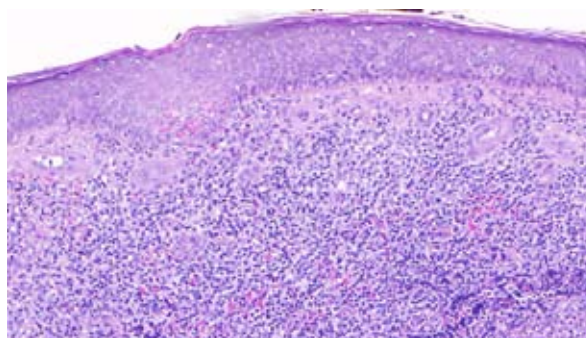


Figure 2. Histologic examination: lymphocytic infiltrate with numerous eosinophils and increased vascularity with enlarged and epithelioid endothelial cells (hematoxylin and eosin, $\times 200$).

ALHE generally presents as solitary or multiple erythematous or hyperpigmented dome-shaped papulonodules. Lesions can be pruritic or painful and do not tend to resolve spontaneously (4). The pathogenesis of ALHE remains controversial, although some theories have been suggested. The most widely accepted hypothesis is that it is an angioproliferative process, accompanied by an inflammatory infiltrate, reactive to several stimuli (3). Some authors believe it is an allergic reaction, but no specific sole agent has been identified (5). Others claim ALHE may represent a T-cell lymphoproliferative disorder of benign or low-grade malignant nature (6). Some recent studies suggest that ALHE pathogenesis may be related to a vascular malformation secondary to a subcutaneous arteriovenous shunt (1-3). Histologically there are both vascular and inflammatory components, with an abnormal vascular proliferation and diffuse lymphocytic infiltrates with eosinophils. The vascular component is formed by capillaries clustered around arterial or venous vessels, dilated and atypical, with a protruded endothelium (3). The main differential diagnosis of ALHE is Kimura's disease, and there has been some discussion regarding the relationship between these two entities due to their clinical and histopathological similarities. However, most studies currently agree that they are distinct diseases. The differential diagnosis also includes angiosarcoma, particularly the epithelioid variant, epithelioid hemangioendothelioma, Kaposi sarcoma, pyogenic granuloma, and cutaneous metastasis (3). ALHE usually requires treatment as spontaneous regression, although reported in the literature, is rare (1,3). Many options have been suggested, with variable levels of success, but there is no definitive treatment for this condition (2). Surgical excision is the preferred choice, but recurrence may happen if the excision is incomplete (1). Mohs micrographic surgery with excision of abnormal vessels at the base of the lesion may be more effective in reducing recurrences (4). Other treatments reported include laser therapy (pulsed dye, CO₂, copper vapor), systemic or intralesional corticosteroid injection, cryotherapy, imiquimod, tacrolimus, isotretinoin, radiotherapy, interferon alfa 2a, anti-interleukin-5 antibody, photodynamic therapy, and methotrexate (1).

In the present case the diagnosis of ALHE was established through the conjunction of clinical and histological findings. Although a rare entity, its predominantly facial involvement in young adults and the absence of a satisfactory treatment can produce a significant impact that can include the quality of life of the patients.

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