Angiolymphoid hyperplasia with eosinophilia
Angiolymphoid hyperplasia with eosinophilia (ALHE) is an uncommon idiopathic condition that manifests in adults as isolated or grouped papules, plaques, or nodules in the skin of the head and neck. Most patients present with lesions in the periauricular region, forehead, or scalp. Rare sites of involvement include the hands, shoulders, breasts, penis, oral mucosa, and orbit. A distinct pathologic entity, ALHE is marked by a proliferation of blood vessels with distinctive large endothelial cells. These blood vessels are accompanied by a characteristic inflammatory infiltrate that includes eosinophils. The lesion is benign but may be persistent and is difficult to eradicate. Whether ALHE represents a benign neoplasm or an unusual reaction to varied stimuli, including trauma, remains unclear.

While ALHE shows some similarity to Kimura disease, it is generally regarded as a separate entity. While ALHE lesions are superficial, Kimura disease involves deeper tissues such as lymph nodes, salivary glands, and the subcutis. However, a 2006 report describes ALHE involving the nail bed and underlying bone. Further, no reports describe a patient with simultaneous tumors, one consistent with Kimura disease and one consistent with ALHE. Such findings challenge whether or not Kimura disease and ALHE represent a spectrum of the same disease.

Pathophysiology

Although ALHE may be a benign tumor, numerous factors suggest that it is an unusual reactive process. Approximately half the patients have multifocal lesions that are grouped anatomically. ALHE has occurred following various forms of trauma or infection. Histologically, most cases 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. Hyperestrogenemia (eg, in pregnancy, with oral contraceptive use) may foster lesion growth. Additionally, the distinctive inflammatory infiltrate in ALHE appears to be an intrinsic (not secondary) component of the lesion. Approximately 20% of patients have blood eosinophilia. However, immunoglobulin E levels are not elevated. Additionally, 3 reported cases describe follicular mucinosis and ALHE occurring in the same biopsy specimen. Interestingly, one report of monoclonality in a patient with ALHE who subsequently progressed to mycosis fungoides raises the question of whether or not ALHE could be an early form of T-cell lymphoma. It should be stressed that most cases are entirely benign.

Patients with angiolymphoid hyperplasia with eosinophilia (ALHE) typically present with an expanding nodule or group of nodules, usually in the vicinity of the ear (see the image below). The lesion(s) may be associated with pain or pruritus. Uncommon symptoms include pulsation and spontaneous bleeding.
Angiolymphoid hyperplasia with eosinophilia (ALHE) typically appears as dome-shaped, smooth-surfaced papules or nodules (see the image below). Approximately 85% of lesions occur in the skin of the head and neck; most of them are on or near the ear or on the forehead or scalp. The extremities are the next most common site. Involvement at other sites is rare. However, case reports have described ALHE affecting the penis and the conjunctiva.

**Treatment**

**Medical Care**

Angiolymphoid hyperplasia with eosinophilia (ALHE) treatment is often challenging. Rarely, spontaneous resolution has been known to occur, obviating the need for medical intervention in some cases. Intralesional corticosteroids and irradiation have been used but are not very effective. Other treatments that have been reported include topical imiquimod, topical tacrolimus, isotretinoin, and interferon alfa-2b.

**Surgical Care**

Simple surgical excision is sometimes used, but the lesions tend to recur. Mohs micrographic surgery has been attempted in order to address ALHE through better margin control. Excisions that include the arterial and venous segments at the base of the lesion prove most efficacious. The pulsed-dye laser and carbon dioxide laser have been used with some success. Cryosurgery and electrosurgery have also been reported.