Angiolymphoid Hyperplasia with Eosinophilia on the Lower Extremity: Case Report with Brief Review

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Abstract

Angiolymphoid hyperplasia with eosinophilia (ALHE), also known as epithelioid hemangioma (EH), is an uncommon, benign vascular proliferation characterized by an inflammatory infiltrate composed primarily of eosinophils. In the current literature, there is little evidence of ALHE occurring on the lower extremities. We describe a rare case of ALHE occurring in a 48-year-old Caucasian female who presented with a pruritic, tender, palpable lesion on the right posterior thigh. Histopathological examination revealed increased dermal vasculature comprised of thick-walled blood vessels with protuberant hobnail endothelium. The lesion was diagnosed as angiolymphoid hyperplasia with eosinophilia, and the patient was referred to a Mohs surgeon for local excision.

Introduction

Angiolymphoid hyperplasia with eosinophilia (ALHE), also known as epithelioid hemangioma (EH), is a rare, benign vascular proliferation first described by Wells and Whimster in 1969. ALHE is characterized by a proliferation of epithelioid cells in thick-walled blood vessels. Clinically, ALHE appears as smooth, violaceous, red-brown papules or nodules in the head and neck region, most commonly near the ears.1-3 The lesions can be solitary or numerous and range in size from 1 cm to 10 cm.3-4 Generally, the lesions are asymptomatic, though patients occasionally present with a pulsatile sensation, pruritus, or bleeding. ALHE rarely causes systemic complications, unlike the related entity Kimura disease, which is strongly linked to nephropathy and the resulting nephrotic syndrome with proteinuria.3,4,5 Both conditions are eosinophilic dermatoses, but Kimura's disease is associated with increases in serum eosinophil count and immunoglobulin E (IgE) level. Approximately 20% to 21% of patients with ALHE have blood eosinophilia, but without elevated IgE.3,5,6 As a result, peripheral eosinophilia is not required to make the diagnosis of ALHE.

There are several theories on the pathogenesis of ALHE, though the specific etiology remains elusive. Current literature suggests ALHE is a reactive process, proposing trauma, infection or hormonal/autoimmune disorders as causes.5,8,9 In many cases, the base of the lesion shows evidence of tortuous vasculature on histology, suggesting arteriovenous shunting plays a role in the pathogenesis of ALHE.8-10,11,12,13

ALHE is reported most frequently in females between 20 years and 40 years old.5,6 The most common age at presentation is between 30 years and 33 years.1,2 Children and the elderly are rarely afflicted with ALHE. Along with clinical findings, histology is the principal diagnostic component. The fundamental histopathologic finding in ALHE is vascular proliferation lined by epithelioid endothelial cells.8,12,15 The endothelial cells are enlarged, with abundant eosinophilic cytoplasm and large nuclei with intermittent hobnailing or a cobblestone appearance.7,8 ALHE is also characterized by an inflammatory infiltrate often comprised of histiocytes, mastocytes and, most notably, eosinophils.3,4 Eosinophils have been found to account for up to 50% of the ALHE infiltrate.4,5 Ordinarily, ALHE does not resolve spontaneously, and the mainstay of treatment is surgical excision.1,7,9,17

Case Description

A 48-year-old Caucasian female presented with concerns about her psoriasis and a pruritic, tender, palpable lesion on the right posterior thigh. Histopathological examination revealed increased dermal vasculature comprised of thick-walled blood vessels with protuberant hobnail endothelium. The lesion was diagnosed as angiolymphoid hyperplasia with eosinophilia, and the patient was referred to a Mohs surgeon for local excision.

Following punch biopsy, histopathological examination revealed parakeratosis with psoriasiform epidermal hyperplasia and an inflammatory interstitial infiltrate extending from the dermis into the subcutaneous tissue (Figure 2). Also present was increased dermal vasculature comprised of thick and thin-walled blood vessels with protuberant hobnail endothelium (Figures 3, 4), and a mixed inflammatory infiltrate comprised of lymphocytes, histiocytes, plasma cells and many eosinophils (Figure 5). CD34 immunohistochemical stain identified dermal vessels with endothelial hyperplasia within the
proliferation surrounded by an inflammatory infiltrate comprised primarily of eosinophils. The most common location for lesion development is the head and neck area. We recommend clinicians reassure patients and families that the vast majority of ALHE cases are entirely benign, with no malignant transformation reported.1,3

In our case, ALHE involved the lower extremity, an unusual clinical presentation. Histopathological findings, however, were typical of ALHE. The patient underwent Mohs excision of the lesion and is following up routinely to evaluate for new lesions and receive continued treatment of her psoriasis.

Discussion
Angiolymphoid hyperplasia with eosinophilia is an uncommon, benign vascular disease commonly presenting as subcutaneous nodules on the head and neck. Current literature describes very few cases of ALHE involving the lower extremities. A literature search turned up only three reports: The first involved the distal femoral epiphysis; the second was a presumed but unconfirmed case of ALHE in the posterior tibial artery; and the third was a single lesion on the thigh and inconclusive histologically, displaying features of both Kimura’s disease and ALHE.4,8,18

ALHE and Kimura’s disease can be differentiated on pathology. ALHE is characterized by the occasional lymphoid follicle, whereas Kimura’s disease is known for well-formed lymphoid follicles in addition to the eosinophilic infiltration.3,9,14

Since the first reported cases of ALHE, most cases have occurred within the cephalic region. The predilection for the head and neck, specifically the periauricular region, is well noted throughout the literature.3,5,6,9 All nine of Wells and Whimster’s first reported cases involved the head and neck.3 Additional sites of involvement include the hands, shoulders, breasts, oropharynx, orbit, ocular adnexa, upper extremities and genitalia.1,5,9,13 There are rare reports of extracutaneous involvement, including of the lungs, bone, colon and heart.3,10,19

ALHE rarely undergoes spontaneous resolution, and the mainstay of treatment is surgical excision. Recurrence is observed in one-third of cases.7,8,20 Lembo et al. reports local recurrence may be as high as 50 percent after standard surgical excision.17 Due to recurrence, other, less-invasive therapeutic modalities have been explored, including topical tacrolimus, isotretinoin, imiquimod and interferon alpha.1,21-24 Intralesional corticosteroid therapy has shown better aesthetic results than topical, with recurrence rates comparable to surgical excision.6,47 Surgical excision, however, remains the treatment of choice, and when the lesion is completely excised, recurrence is rare.3,1,3,12,16,17,25,26

Conclusion
Angiolymphoid hyperplasia is an uncommon lesion characterized by endothelial cell lumina (Figure 6). A PAS stain was negative for fungal organisms. The lesion was diagnosed as angiolymphoid hyperplasia with eosinophilia, and the patient was referred to a Mohs surgeon for local excision.

References


