SKIN

BRIEF ARTICLE

A Rare Presentation of Angiolymphoid Hyperplasia with Eosinophilia Involving the Scrotum

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ABSTRACT

Angiolymphoid hyperplasia with eosinophilia (ALHE) is a rare vascular proliferation that typically manifests as pink to reddish brown papules or nodules. These lesions classically affect the face or neck. Involvement of other structures including the tongue, colon, salivary glands, and muscle are rare. Male urogenital involvement for example, had less than 30 cases reported. Here we present the third reported case of ALHE involving the scrotum in a 17-year-old male.

INTRODUCTION

Angiolymphoid hyperplasia with eosinophilia (ALHE) is an uncommon vascular proliferation. The wide histologic variations in presentation have spawned an equally wide degree of nomenclature of ALHE. Epitheliod hemangioma, pseudopyogenic granuloma, inflammatory angiomatous nodule, popular angioplasia, subcutaneous angioblastic lymphoid hyperplasia with eosinophilia and lymphofolliculosis, intravenous atypical vascular proliferation and histiocytoid hemangioma have all been utilized by various authors over the decades.[1] The lesions of ALHE are benign and typically present as papules or nodules on the face or neck.[1] There is a slight predilection of females over males in ALHE with most cases occurring between the ages of 20 and 50.[2] It is likely an underreported condition, therefore, true demographic information may be lacking.

Clinically, ALHE presents as pink to red-brown papules or nodules on the face or neck with a tendency to form in the preauricular area. ALHE lesions often bleed easily and present as either asymptomatic or with pain, pruritis, or pulsations.[3] Renal disease and peripheral eosinophilia occur in a minority of patients.[4] Involvement of the oral mucosa, bone, muscle, parapharyngeal space, tongue, colon, and salivary glands are rare.[5] There are less than 30 cases reported of male urogenital involvement. Here we present the third reported case of ALHE with scrotal involvement.[6,7]

CASE REPORT

A 17-year-old male with a history of refractory verrucous vulgaris presented for asymptomatic nodules on scrotum increasing in size and number over the last 3 years (Figure 1). The patient denied any sexual history, pain, discharge, or trauma to the area. A biopsy was taken to rule out

scrotal syringomas or steatocystoma multiplex. Dense collections of lymphocytes with scattered histiocytes and eosinophils were appreciated in the dermis against a background of increased vasculature. Microbial stains to rule out an infectious trigger were negative. Angiolymphoid hyperplasia with eosinophilia was diagnosed. Treatment options were discussed with the patient, but he has yet to decide upon a course of action. Molecular clonality was not investigated due to cost constraints.

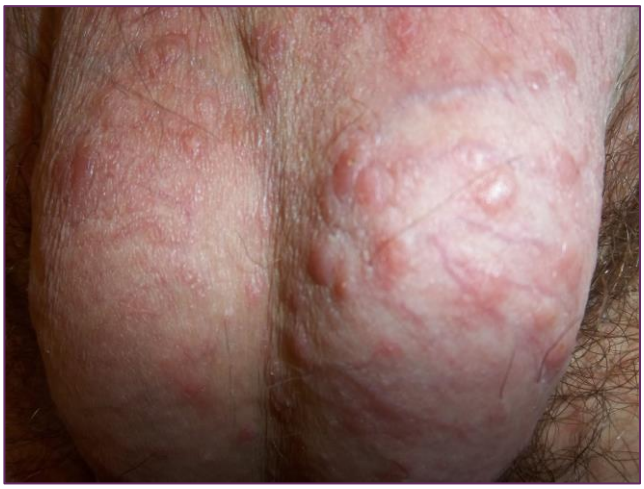

Figure 1.

DISCUSSION

The first description of ALHE was by Wells and Whimster in 1969.[8] Pathogenesis is unknown, but there is speculation that the lesions result from trauma to an artery or vein, infection, or from uncontrolled vascular factors.[9] An estimated less than 10% of patients can recall a history of localized trauma or infection. In these cases, the median interval of lesion manifestation was 30 months.[9] The results from a study of 53 out of 116 patients by Olsen and Helwig suggested that ALHE may be caused by arteriovenous shunts.[9] Additionally, the patient's history of refractory verruca vulgaris may provide further insight into the pathophysiology of ALHE. Since infections have been raised as a possible origin, might there be a possible association between HPV and ALHE?

Our case also raises awareness for the spectrum of presentations ALHE may manifest as when it involves the scrotum. Compared to the other two known reported cases of ALHE with scrotal involvement (Figure 2), our case is the only one that presented bilaterally on the scrotum. Hence, dismantling any notion of using unilateral/bilateral spatial arrangements as a possible differentiating factor when considering scrotal involvement of ALHE. Furthermore, when analyzed alongside previous cases, it appears ALHE of the scrotum may vary in many other characteristics such as tenderness, pruritis and size.

ALHE Cases with Scrotal Involvement	Age	Presentation
Chen et al.	38 years old	Solitary, indurated, non-tender, well-circumscribed, dull red to brownish plaque (1cm in diameter), unilateral scrotum
Park et al.	4 years old	Pruritic, tender, indurated dull red nodules, unilateral scrotum

Figure 2.

Case reports of angiolymphoid hyperplasia with eosinophilia have been inconsistent and underreported due to cases being incorrectly diagnosed as Kimura's disease (KD).[9] While ALHE and KD have similar characteristics, they are now characterized as different disorders.[1,9] Histologically, the main characteristic of ALHE is vascular proliferation sometimes demonstrating cobble stoning with an eosinophil infiltrate.[1] KD is generally deeper with lymphoid follicles, dense fibrosis, and an eosinophil infiltrate.[1] ALHE is a benign primary vascular proliferation, while KD is due to chronic inflammation and is either an allergic or autoimmune disorder.[10] While a

case report by Chong et al presents a patient demonstrating features of both conditions simultaneously, the significance of this occurrence remains unknown.[11]

Many treatment modalities have been used in attempts to clear ALHE lesions, but it is notoriously refractory to current lines of treatment. Surgical excision is often used after other non-invasive options have failed. Other commonly used treatments include intralesional corticosteroids and laser therapy.[3] Lesions after surgical excision have a 33-50% recurrence rate. Mohs micrographic surgery has also been used for treatment with results showing promise of completely resolving lesions with no recurrence and minimal destruction of healthy tissue.[12] The high cost and exclusion criteria however preclude most patients from considering this option. Other treatment modalities have included topical tacrolimus, electrocoagulation, corticosteroids, sclerotherapy, indomethacin, retinoids, cryotherapy, pentoxifylline, intravenous vinblastine sulfate, radiotherapy, interferon α-2b therapy, intralesional bleomycin, and radiofrequency excision.[12]

CONCLUSION

In conclusion, we are reporting only the third known case of scrotal angiolymphoid hyperplasia with eosinophilia. It is a rare but benign vascular proliferation that typically presents on the head or neck area. Due to the high rates of recurrence, many treatment modalities have been used in attempts to clear lesions.

Conflict of Interest Disclosures: None

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