

Angiolymphoid Hyperplasia with Eosinophilia: A Case Report

Paola Esteves^a Marcella Barbalho^a Tiago Lima^a Leonardo Quintella^b
João Paulo Niemeyer-Corbellini^a Marcia Ramos-e-Silva^a

^aSector of Dermatology and Post-Graduation Course and ^bSector of Pathology, University Hospital and School of Medicine, Federal University of Rio de Janeiro, Rio de Janeiro, Brazil

Key Words

Angiolymphoid hyperplasia with eosinophilia · Kimura's disease · Vascular malformation

Abstract

Angiolymphoid hyperplasia with eosinophilia is a rare disease considered as being a vascular malformation resulting from a subjacent arteriovenous shunt. It affects mostly the head of women between 20 and 40 years old and may present spontaneous involution.

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Introduction

Angiolymphoid hyperplasia with eosinophilia (ALHE) is a rare vascular disorder described in 1969 by Wells and Whimster [1]. It preferably affects women between 20 and 40 years old, being of benign character. Underlying vascular malformation or local trauma are suggested as possible causes [2–4]. ALHE is often mistaken for Kimura's disease [2, 3, 5].

ALHE is characterized by erythematous or violaceous papulonodular lesions located on the head and neck [5]. Occurrence of lesions on the trunk, upper limbs and genital region is rare. The lesions may be asymptomatic, painful or pruriginous [4]. Lymphadenomegaly and peripheral eosinophilia may be present. In some older reports ALHE and Kimura's disease were even described as the same disease with spectral difference [6]. Spontaneous involution may occur, but lesions can persist indefinitely [5].

Case Report

Our patient was a 52-year-old married woman, a manicurist, born and living in the State of Rio de Janeiro. She complained about appearance of pruriginous papules on her scalp about 6 months before. She reported ulcerations and discrete bleeding after scratching occasioned by pruritus. She denied constitutional symptoms or local trauma. She had a history of hypothyroidism. Her serum eosinophilic count was normal.

At dermatological examination three grouped erythematous papulonodular lesions were observed, with a diameter of 1 cm each, located on the occipital region (fig. 1). A biopsy was carried out on one of the lesions, and histopathological examination revealed proliferation of vessels of varied calibers as well as inflammatory infiltrate comprising lymphocytes and eosinophils in the dermis (fig. 2). The patient refused surgical procedure and opted for application of imiquimod once a day, 5 times a week for 12 weeks, without any improvement of the clinical picture.

Discussion

ALHE is a benign slowly growing tumor with a still not totally clarified etiopathogenesis. It can be self-limited and characterized by intense vascular proliferation [7]. Much confusion exists in the literature regarding ALHE and Kimura's disease. Both present nodules, preferably on the head and cervical region, but in ALHE they tend to be much more erythematous as opposed to Kimura's disease, where the lesions are normochromic. They also share histopathological similarities, such as involvement of the dermis and subcutaneous infiltrate comprising lymphocytes and eosinophils, proliferation of endothelial cells and the absence of adnexal structure involvement [3, 7].

Men of Asian origin are more prone to Kimura's disease, which features a triad of painless subcutaneous masses, usually unilateral, in the head and neck region. There is peripheral eosinophilia and significant increase in immunoglobulin E levels in the tissues. Salivary gland enlargement may also occur. In histopathology, Kimura's disease differs from ALHE for being mainly a disorder of the lymphoid follicles, without irregular blood vessels and with non-protruding endothelial cells in the vascular lumen, but always presenting an increased number of eosinophils that may extend to the muscular fascia [8].

The treatment of choice of ALHE is surgical excision, but due to the high rate of recurrence, other less invasive therapeutic modalities may be employed [5]. Intralesional corticoid therapy is effective with recurrence rates similar to surgical excision, however with better aesthetic result [8]. Other treatments such as cryotherapy [5], photodynamic therapy [9] and application of imiquimod are described in literature with good results. The latter acts by induction of the production of interferon- α and induction or inhibition of certain cytokines, mainly interleukin-5, implicated in the genesis of ALHE [10].

Conclusion

In the present case, clinical and histopathological examination were typical of ALHE [11]. The disease did not present spontaneous remission 1 year after imiquimod cessation. The patient is in follow-up every 6 months in order to evaluate possible progression or appearance of new lesions and possible therapeutic changes if the patient consents.

Statement of Ethics

The authors state that the patient gave her informed consent to have her case report published.

Disclosure Statement

The authors have no conflict of interest for this publication and there was no funding.

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Fig. 1. Grouped erythematous nodules in the occipital region.

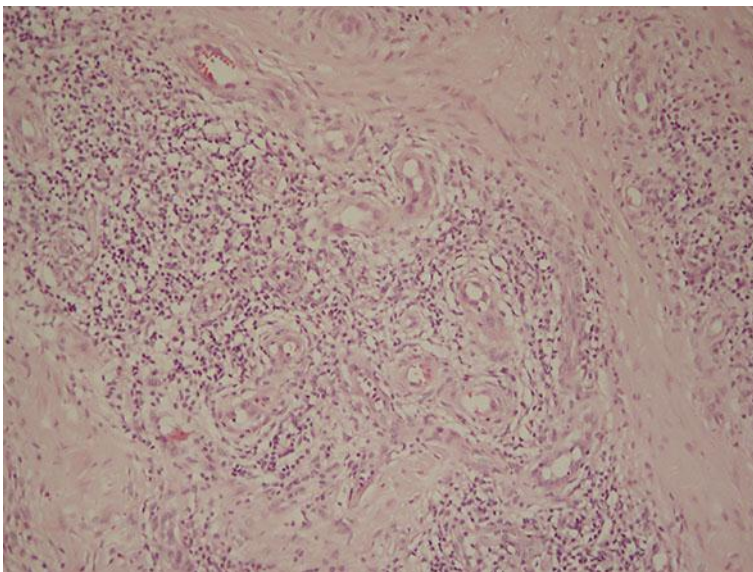


Fig. 2. Vessels of different calibers and inflammatory infiltrate rich in lymphocytes and some eosinophils (hematoxylin and eosin, $\times 40$).