

## Case for diagnosis \*

### Caso para diagnóstico

Felicidade Santiago<sup>1</sup>

José Pedro Reis<sup>2</sup>

#### CASE REPORT

A 22-year old male patient presented with skin lesions on his face that had been becoming progressively worse over the previous four years. The patient had no relevant personal history of atopy, infection or trauma associated with the appearance of the skin lesions. At objective examination, multiple erythematous, purplish, shiny, hard papules and nodules were found, some with superficial erosion and crusting. They were situated unilaterally on the right side of his face in the preauricular area, tragus and external auditory canal (Figures 1 and 2). The remainder of the objective examination was normal, with no adenopathies or locoregional masses being found. The lesions were asymptomatic, albeit sporadically friable.

The skin biopsy showed a dense infiltrate in the dermis associated with capillary proliferation with hyperplasia of the endothelial cells, which were swollen and protruding towards the vascular lumen (Figure 3).

Laboratory evaluation, which included full blood count with differential white blood cell count, serum biochemical tests, measurement of immunoglobulin levels and urinalysis, was normal.

Various forms of treatment were implemented, albeit unsuccessfully: tacrolimus ointment, topical and oral corticotherapy and oral isotretinoin; however the best results were obtained with electrocoagulation and CO<sub>2</sub> laser.



FIGURE 1: Skin lesions in the preauricular region



FIGURE 2: Details of the lesions in the external auditory canal

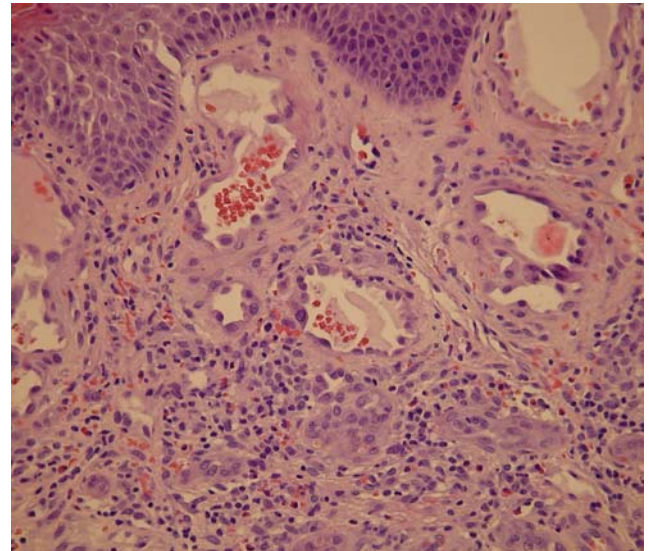


FIGURE 3: Histopathological examination showing capillary proliferation with hyperplasia of the endothelial cells accompanied by inflammatory infiltrate consisting of eosinophils and mononuclear lymphocytes

Received on 10.08.2010

Approved by the Editorial Board and accepted for publication on 25.08.2010.

\* Study conducted at the Teaching Hospitals of the University of Coimbra, Coimbra, Portugal.

Conflict of interest: None / *Conflito de interesse: Nenhum*

Financial funding: None / *Suporte financeiro: Nenhum*

<sup>1</sup> Internship in Dermatology, Dermatology Department, Teaching Hospitals of the University of Coimbra, Coimbra, Portugal.

<sup>2</sup> Assistant physician, Dermatology Department, Teaching Hospitals of the University of Coimbra, Coimbra, Portugal.

## DISCUSSION

The clinical and histopathological data are consistent with a diagnosis of angiolymphoid hyperplasia with eosinophilia (ALHE). ALHE was first described by Wells and Whimster in 1969 as a late stage of Kimura's disease (KD).<sup>1</sup> Currently, these conditions are recognized as distinct clinicopathological entities and ALHE is considered a benign form of vascular hyperplasia.<sup>2</sup> It is an unusual disease, albeit not rare, affecting individuals of 20 to 50 years of age (mean 30-33 years), principally women. It typically presents as angioma-tous-appearing papules and nodules that are almost always superficial and range in size from 0.5 to 2 cm. In 50% of cases, the lesions are multiple, clustered together at the same anatomical site. As a rule, they are asymptomatic but may occasionally cause pain and pruritus, become pulsatile or suffer sporadic spontaneous hemorrhages. They are located predominantly on the head and neck (85%), with a predilection for the periauricular region, the scalp and forehead.<sup>2,3</sup>

From a histopathological point of view, ALHE has two components: vascular and inflammatory. The vascular component is characterized by an anomalous capillary proliferation with lumens of irregular morphology lined with swollen, protruding endothelial cells. These hyperplastic endothelial cells, which develop epithelioid or histiocytoid morphology, are the principal histological characteristics of ALHE. The vascular alterations are associated with an interstitial and perivascular inflammatory infiltrate consisting of lymphocytes, mastocytes and eosinophils, the latter of which are generally present in percentages that range from 5 to 15%, rarely reaching 50%, and may even be

completely absent.<sup>2</sup>

The principal differential diagnosis to bear in mind is Kimura's disease, which affects younger, male patients of Asian's origin. KD presents as deep, painless, subcutaneous masses covered by normal skin, located on the head and neck, ranging from 2 to 10 cm in size and associated with locoregional adenopathies that sometimes affect the salivary glands. Histologically, there is a predominance of lymphoid follicles with well-formed germinative centers and a high percentage of eosinophils that occasionally form abscesses. From an analytical point of view, eosinophilia and high IgE levels are characteristic features of KD, whereas in ALHE these analytical alterations are generally absent.<sup>2,5</sup>

The treatment of ALHE, reserved for symptomatic and disfiguring cases, is considered a challenge, with multiple possible forms of management of varying success, ranging from topical treatments (tacrolimus, imiquimod), intralesional treatments (interferon, corticotherapy) and oral treatments (corticotherapy, retinoids).<sup>4,6</sup> Cases of spontaneous regression have been reported. Surgical excision is considered one of the most effective options; however, recurrence of the lesions occurs in around one-third of cases. Therefore, destructive treatments such as electrocoagulation, cryotherapy and pulsed dye laser or CO2 laser are among the most commonly used forms of therapy and those with the highest success rates.<sup>5</sup>

In conclusion, ALHE is a persistent disease that is difficult to treat; however, there is no known risk of malignant transformation. □

**Abstract:** This report describes a patient with clinical and histopathological findings compatible with angiolymphoid hyperplasia with eosinophilia. This benign vascular hyperplasia should be recognized and differentiated from Kimura's disease.  
**Keywords:** Angiolymphoid hyperplasia with eosinophilia; Endothelial cells; Inflammation.

**Resumo:** Descreve-se um paciente com características clínicas e histopatológicas típicas da hiperplasia angiolinfoide com eosinofilia. Trata-se de uma hiperplasia vascular benigna que importa reconhecer e distinguir da doença de Kimura.  
**Palavras-chave:** Células endoteliais; Hiperplasia angiolinfoide com eosinofilia; Inflamação

## REFERENCES

1. Wells GC, Whimster IW. Subcutaneous angiolymphoid hyperplasia with eosinophilia. *Br J Dermatol.* 1969;8:1-14.
2. Requena L, Sangua OP. Cutaneous vascular proliferations. Part III. Malignant neoplasms, other cutaneous neoplasms with significant vascular component, and disorders erroneously considered as vascular neoplasms. *J Am Acad Dermatol.* 1998;38:143-75.
3. Trindade F, Haro R, Requena L. Giant angiolymphoid hyperplasia with eosinophilia on the chest. *J Cutan Pathol.* 2009;36:493-6.
4. Carlesimo M, Mari E, Tammaro A, Persechino S, Camplone G. Angiolymphoid hyperplasia with eosinophilia treated with isotretinoin. *Eur J Dermatol.* 2007;17:554-5.
5. Marcoux C, Bourlond A, Dcroix J. Angiolymphoid hyperplasia with eosinophilia. Remission with acitretin. *Ann Dermatol Venereol.* 1991;118:217-21.
6. Nogueira A, Accioly Filho JW, de Castro MCR, Maceira JP, Ramos-e-Silva M. Hiperplasia angiolinfoide com eosinofilia - relato de dois casos. *An Bras Dermatol.* 2003;78:79-85.

*MAILING ADDRESS / ENDEREÇO PARA CORRESPONDÊNCIA:*

*Felicidade Santiago  
 Serviço de Dermatologia e Venereologia  
 Hospitais da Universidade de Coimbra  
 Praceta - Mota Pinto  
 Coimbra, Portugal 3000-075  
 E-mail: felicidadessantiago@hotmail.com*

How to cite this article/Como citar este artigo: Santiago F, Reis JP. Case for diagnosis. Angiolymphoid hyperplasia with eosinophilia. *An Bras Dermatol.* 2011;86(4):815-824.