CASE REPORT

Hyperkeratosis lenticularis perstans *

Hiperqueratose lenticular persistente

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Abstract: Flegel's disease, also known as hyperkeratosis lenticularis perstans, is a rare skin disease characterized by small reddish-brown asymptomatic hyperkeratotic papules usually located on the lower extremities. The histopathological features are hyperorthokeratosis, epidermal atrophy and band-like inflammatory infiltrate in the superficial dermis. Treatment is generally ineffective. We report a case of hyperkeratosis lenticularis perstans that improved following excisional biopsy of the lesions.
Keywords: Biopsy; Keratinocytes; Keratosis; Leg dermatoses; Skin diseases, papulosquamous; Treatment outcome

Resumo: Doença de Flegel ou hiperqueratose lenticular persistente é uma doença rara, caracterizada por pequenas pápulas hiperceratóticas, assintomáticas, localizadas preferencialmente nos membros inferiores. Histologicamente, há hiperortoceratose, atrofia epidérmica e infiltrado inflamatório em banda. Os tratamentos, em geral, são ineficazes. Relatamos um caso em que as lesões biopsiadas não recidivaram após dois anos de seguimento.
Palavras-chave: Biópsia; Ceratose; Dermatopatias papuloescamosas; Dermatoses da perna; Queratinócitos; Resultado de Tratamento

INTRODUÇÃO

Hyperkeratosis lenticularis perstans (HLP) is a rare disease first described by Flegel in 1958.[1] It is characterized by small erythematous or brownish hyperkeratotic papules distributed symmetrically on the limbs, particularly the dorsal feet and lower third of the legs. Curettage of the hyperkeratotic components of the lesion causes pinpoint bleeding.[2] Although the lesions in the case described here were not abundant, this case report is justified by the rarity of the condition, the resolution of the biopsied lesions and the dearth of articles in Brazilian journals (Scielo, PUBMed, LILACS, Medline and ABD) observed following a review of the literature in electronic format.

CASE REPORT

A 41-year-old white male presented with a 28-year-old complaint of asymptomatic reddish papules between 1 and 5 mm in diameter covered with light-colored or brownish scales in the medial and lateral regions of the feet but not on the plantar surfaces or upper parts of the dorsal feet (Figure 1). Patient history and clinical examination did not reveal any other diseases. Family medical history was unknown because the patient was adopted. During follow-up we observed that the more recent lesions were more erythematous and broader than the older ones. Two excisional biopsies were performed. The first, on a new lesion, revealed hyperkeratosis, parakeratosis, epidermal atrophy and dense lymphohistiocytic infiltrate in the upper dermis (Figure 2A). The second, on an old lesion, revealed discrete atrophy and a discrete infiltrate. Treatment with betamethasone, salicylic acid, calcipotriol, tretinoin, cryotherapy and 5-fluorouracil produced little or no effect. There was no recurrence of lesions in the biopsied areas after two years of follow-up.

DISCUSSION

The etiology of hyperkeratosis lenticularis perstans is unknown. However, according to some authors, it may be the result of a keratinization disorder and may be triggered by ultraviolet radiation. It

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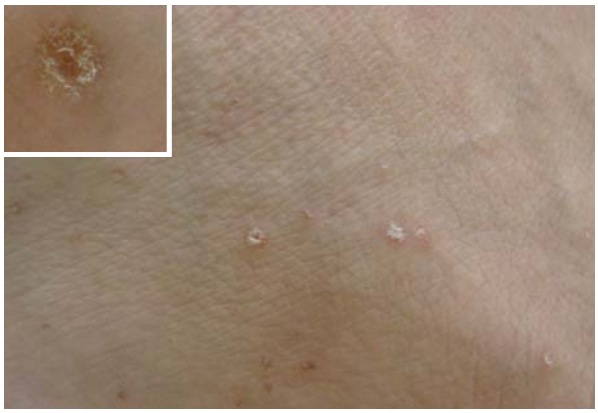

Figure 1: Hyperkeratosis lenticularis perstans – erythematous hyperkeratotic papules a few millimeters in size located in the lateral and medial regions of the dorsal foot. The older lesions are brownish and flatter. The inset in the top left shows a close-up of a more recent lesion. The lesion is an inflammatory papule with a light scale

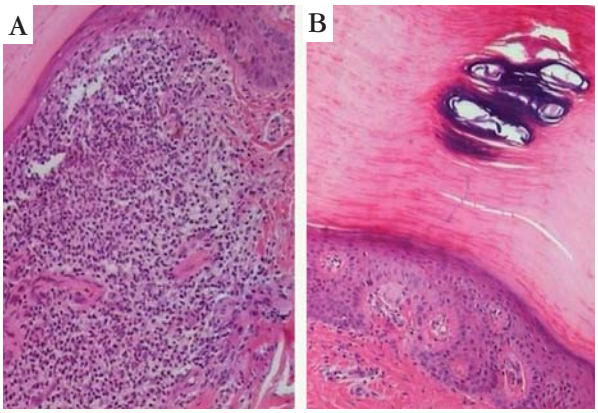

Figura 2: A. Pathological examination - Optical micrograph - hyperkeratosis, epidermal atrophy, dense band-like inflammatory infiltrate in the papillary dermis with lymphocytes and some histocytes in new lesions (hematoxylin-eosin [HE] stain, 100x). **B.** In the old lesions the infiltrate and atrophy are discrete (HE, 60x)

may also be hereditary, with autosomal dominant transmission.[2]

Histologically, the condition is characterized by hyperkeratosis, a thinning or absence of the granular layer, epidermal atrophy and a band-like infiltrate in the upper dermis. These characteristics vary between new and old lesions.[3] In new lesions there is more atrophy and inflammation, in agreement with our findings. There is a lack of consensus among ultrastructural studies; some suggest that there is a reduction in the number of Odland bodies, while others disagree.[4,5]

Differential diagnosis is with stucco keratosis, actinic keratoses, focal acral hyperkeratosis, Darier's disease and Kyrle's disease. Diagnosis is confirmed by histological examination and the clinical evolution of the disease.[6] It has been reported that there is an association between basal cell and squamous cell carcinomas and HPL. [7] The association between HLP and hyperthyroidism, diabetes and gastrointestinal cancer is not significant. [8] Treatment is challenging and yields irregular results. While there are reports of improvements with corticosteroids, vitamin D analogs and 5-fluorouracil, there have also been reports of failure to respond to treatment with the first two of these.[9,10] For our patient, treatment with these topical medicines was ineffective. Interestingly, however, the lesions that were biopsied healed completely. We believe there was no recurrence because this procedure removed the defective keratinocytes. Ablative treatments that replace the defective keratinocytes with new cells should therefore be investigated. Such treatments include CO2 laser, curettage and electrocoagulation. These treatments were not carried out as the patient did not give his consent. ❑

REFERENCES

1. Flegel H. Hyperkeratosis lenticularis perstans. Hautarzt. 1958;9:363-4.
2. Price ML, Jones EW, MacDonald DM. A clinicopathological study of Flegel's disease (hyperkeratosis lenticularis perstans). Br J Dermatol. 1987;116:681-91.
3. Ando K, Hattori H, Yamauchi Y. Histopathological differences between early and old lesions of hyperkeratosis lenticularis perstans (Flegel´s Disease). Am J Dermatopathol. 2006;8:122-6.
4. Langer K, Zonzits E, Konrad K Hyperkeratosis lenticularis perstans (Flegel's disease). Ultrastructural study of lesional and perilesional skin and therapeutic trial of topical tretinoin versus 5-fluorouracil. J Am Acad Dermatol. 1992;27:812-6.
5. Tidman MJ, Price ML, MacDonald DM. Lamellar bodies in hyperkeratosis lenticularis perstans. J Cutan Pathol. 1987;14:207-11.
6. Cunha Filho RR, Almeida Jr HL. Hiperceratose focal acral associada à hipocromia de dermatóglifos. An Bras Dermatol. 2008;83:441-3.
7. Beveridge GW, Langlands AO. Familial hyperkeratosis lenticularis perstans associat ed with tumours of the skin. Br J Dermatol. 1973;88:453-8.
8. Ishibashi A, Tsuboi R, Fujita K. Familial hyperkeratosis lenticularis perstans. Associated with cancers of the digestive organs. J Dermatol. 1984;11:407-9.
9. Blaheta HJ, Metzler G, Rassner G, Garbe C. Hyperkeratosis lenticularis perstans (Flegel's disease) - lack of response to treatment with tacalcitol and calcipotriol. Dermatology. 2001;202:255-8.
10. Sterneberg-Vos H, van Marion AM, Frank J, Poblete-Gutierrez P. Hyperkeratosis lenticularis perstans (Flegel's disease) - successful treatment with topical corticosteroids. Int J Dermatol 2008;47:38-41.

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