

Circumscribed palmoplantar hypokeratosis: report of two Brazilian cases*

Hipoqueratose circunscrita palmoplantar: relato de 2 casos brasileiros

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Abstract: Circumscribed palmoplantar hypokeratosis is a recently recognized dermatosis and rarely reported. It was first described in 2002 and is characterized by localized loss of the horny layer in the palmoplantar area. This dermatosis is clinically presented with a sharply circumscribed, reddish and asymptomatic plaque with slightly depressed surface localized on the palms or the soles. The clinical differential diagnosis includes mainly porokeratosis and Bowen's disease. Its pathogenesis remains unknown, but studies have proposed a human papillomavirus induced disease or a localized keratinization disorder in the palmoplantar area. We report herein two cases of patients with lesions clinically and microscopically compatible with the diagnosis of circumscribed palmoplantar hypokeratosis. We also present a brief literature review of the etiopathogenic hypotheses of this dermatosis.

Keywords: Bowen's disease; Keratins; Porokeratosis

Resumo: A hipoqueratose palmar circunscrita é uma condição rara e pouco conhecida que foi caracterizada apenas em 2002. Caracteriza-se por uma perda circunscrita e focal da camada córnea das regiões palmares ou plantares. Clinicamente, observam-se lesões avermelhadas, circunscritas, arredondadas, discretamente deprimidas e assintomáticas, nas palmas ou, mais raramente, nas plantas. A diagnose diferencial clínica é feita principalmente com a poroqueratose palmoplantar e com a disqueratose de Bowen. Sua patogênese é desconhecida, havendo especulações sobre possível origem viral ou de alteração localizada da expressão de citoqueratinas das regiões palmoplantares. Relatamos os casos clínicos de duas doentes femininas com lesões clínicas e alterações histopatológicas características de hipoqueratose palmar circunscrita de evolução crônica, além de uma breve revisão da literatura sobre as hipóteses etiopatogênicas desta dermatose.

Palavras-chave: Doença de Bowen; Poroceratose; Queratinas

INTRODUCTION

Circumscribed palmoplantar hypokeratosis was initially described by Perez et al in 2002 and is characterized by a nummular, circumscribed, depressed, reddish and asymptomatic plaque, commonly located over the thenar and hypothenar eminences of the palm.¹ It rarely occurs in the plantar region. It affects adults or elderly patients, with higher prevalence in females. Histopathology shows sharp decrease in the stratum corneum without

changing the dermis or the surrounding tissue and the absence of local inflammation. While analyzed by electronic microscopy, it typically presents vacuolization of corneocytes.

There are some etiopathogenic hypotheses for the appearance of the lesions, as trauma, papilloma virus infection types 4 and 6, keratinizing disorder localized to the granular and horny layers, reduction of keratin 9 and connexin 26 expression at the lesional

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epidermis and increase of keratin 16 expression associated with reduction of keratin 2e expression.^{2,6}

Palmoplantar hypokeratosis can clinically mimic palmoplantar psoriasis, porokeratosis and Bowen's disease, but there's no report of malignant lesions.

The disease course is chronic and there is no need for treatment.

CASE REPORT

Case 1: Female patient, 68 years old, white, had had lesions on the palms for 30 years. There was no history of trauma or local infection. The dermatological exam showed two reddish plaques of approximately 5x5mm on the right palm and 5x10mm on the left palm. The lesions were circumscribed, nummular, depressed and located symmetrically over the thenar regions (Figures 1 and 2). Her soles were not involved. The general clinical examination was not significant. Direct mycological examination was performed with negative results, and histopathological aspects of skin biopsy showed a focus of sharp decrease in the stratum corneum thickness, mild acanthosis and no change in the basal layer (Figure 3).

The one year follow-up showed no change of clinical lesions.

Case 2: Female patient, 84 years old, white, had had a single lesion on the left palm for over 24 years. The patient had no history of trauma or local infection. The physical examination revealed a reddish, nummular, depressed and circumscribed patch, about 5 mm in diameter, located over the thenar eminence of the left palm (Figures 4 and 5). There were no lesions on contralateral palm or on soles.

Direct mycological examination was performed with negative results. Histologically, it showed an abrupt, well-demarcated decrease in the thickness of the stratum corneum and a diminished granular layer (Figure 6).



FIGURE 2: Detail of the right palm lesion (Case 1)

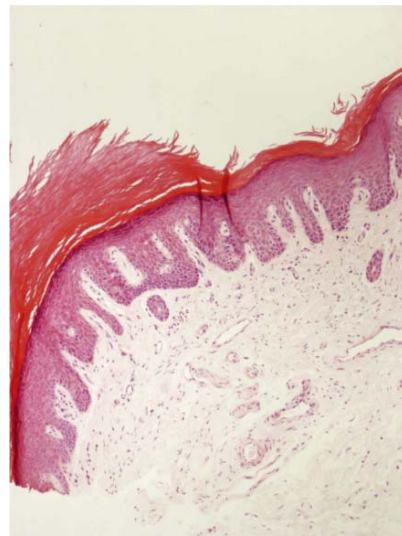


FIGURE 3: Histopathological aspects (Case 1) - Focus of sharp decrease in the stratum corneum thickness - HE



FIGURE 1: Circumscribed and reddish plaque located on the thenar eminences (Case1)



FIGURE 4: Reddish, nummular and depressed lesion, located on the thenar eminence of the left palm (Case 2)



FIGURE 5: Detail of the lesion (Case 2)

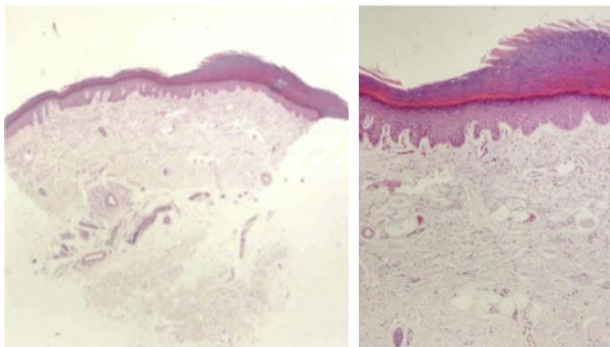


FIGURE 6: Histopathological aspects (Case 2) - Abrupt decrease in the stratum corneum thickness - HE

DISCUSSION

The circumscribed palmoplantar hypokeratosis is a benign dermatosis first described by Perez et al. Thus far, little more than 50 cases have been reported in the literature.¹ The cases reported in this study are probably the first reports of this disease in Brazil.

There is a predominance of cases among women (4:1), mean age 65 years. Clinically, lesions are typically nummular, circumscribed, reddish and asymptomatic plaques, commonly located over the thenar and hypothenar eminences of the palms and the medial portion of the plantar region.

The main disorders considered in clinical differential diagnosis are palmoplantar psoriasis, Bowen's disease and palmoplantar porokeratosis. We must also remember dermatophytoses and palmoplantar syphilids. Histopathologically, the palmar hypokeratosis focus is characterized by abrupt reduction of the stratum corneum, forming a "step" between the lesion and the perilesional normal skin. The decrease of the focal stratum corneum is responsible for the clinical aspect of the depressed lesion; the reddish

color is caused by visualization of dermal blood circulation in the area of injury due to corneal thinning. For this reason erythema shall not be considered true, since there is no vasodilatation.

The pathogenesis of palmoplantar hypokeratosis is unknown. Some authors argue that the disease is caused by local trauma, being a "forme fruste" of epidermolysis bullosa, due to an alteration in keratinocytes maturation.² However, only a minority of the previously reported cases had a history of local trauma.

Some authors have detected within palmoplantar hypokeratosis lesions the presence of the human papillomavirus (HPV) 4 and 6, arguing that the infection with these viruses may be a (co) factor in the pathogenesis of this dermatosis.⁴ However, in some other reports where immunohistochemical techniques and polymerase chain reaction were used, there was no evidence of the presence of HPV in lesions.^{1,4,7,8}

Urbina et al showed a reduction of hyaline granules and decrease in number of these keratinocyte cells by electron microscopy, suggesting a primary disturbance of keratinization.⁹

Ishiko et al immunohistochemically demonstrated, in 2007, increased expression of keratin 16 and Ki-67 in skin lesions, speculating that the circumscribed hypokeratosis may be a consequence of epidermal hyperproliferation associated with fragility of keratinocytes.⁶

In 2009, Tanioka M et al proposed the existence of two subtypes of the disease. In the first subtype, the most common lesion is characterized by palmar "erythema", with immunohistochemical reduction of keratin 6 and 16 and normal expression of keratin 9.⁵ The second subtype is typically found in the plantar region with little or no "erythema". In this subtype the expression of keratin 26 and connexin 9 was reduced. There were also non-significant changes of keratins 6 and 16.⁵

Recently, Kanitakis et al showed abnormal expression of some antigens associated with epidermal differentiation and proliferation of keratinocytes, as corneodesmosin, LEKTI (Lymphoepithelial Kazal type inhibitor) and kallikrein 5, concluding that in this dermatosis there is an alteration of the regulation of the cutaneous desquamation process.¹⁰

The circumscribed palmoplantar hypokeratosis does not require treatment and there is no description of malignancy in the literature. Only one case was reported with the presence of alterations such as actinic keratosis limited to hypokeratosis lesions and that may be a consequence of relevant sun exposure.¹⁰

Attempts to treat this disease were made with surgery, cryotherapy, photodynamic therapy and topical therapy with calcipotriol, corticosteroids and retinoids, with poor results¹⁰.

The two cases presented here are fairly typical of circumscribed palmar hypokeratosis. They consti-

tute the first report of this rare condition in Brazil. Familiarity with this illness is relevant for dermatologists in order to properly distinguish it from other palmoplantar dermatosis avoiding unnecessary treatment. □

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