

Keratosis lichenoides chronica - Case report

Queratose liquenoide crônica - Relato de caso

Livia Cibele Gomes Martins¹
 Dário Nunes Moreira Júnior²
 Vitória Regina Pedreira de Almeida⁴

Marina Horne¹
 Ivonise Follador³

Abstract: Keratosis lichenoides chronica or Nekam's disease is a rare mucocutaneous dermatosis characterized by keratinization. It is chronic and progressive usually affecting individuals aged 20-40 years. Around 70 cases have been reported in the literature. Due to the rarity of this condition and the lack of effective treatment, it is a difficult disease to manage. In the case described below we present a 42-year-old patient with violaceous and hyperkeratotic papules in linear, reticular or plaque form, located on the trunk and limbs for five years. Aphthous lesions in the oral cavity and shallow ulcers on the genitalia also formed part of the clinical manifestation. Pathologic examination was suggestive of keratosis lichenoides chronica. Acitretin and dapsone was introduced and the lesions partially improved.

Keywords: Keratosis; Lichenoid eruptions; Skin diseases

Resumo: A queratose liquenoide crônica ou doença de Nekam é uma dermatose mucocutânea rara da queratinização, com curso crônico e progressivo, que acomete geralmente indivíduos entre 20 e 40 anos. Existem, aproximadamente, 70 casos descritos na literatura. Devido à raridade desta dermatose e à ausência de tratamento efetivo, é uma doença de difícil manejo. No caso em questão, apresentamos um paciente de 42 anos com pápulas violáceas, hiperqueratósicas, algumas confluentes, com aspecto linear, rendilhado e em placas, localizadas no tronco e membros há cinco anos. Lesões aftoides na cavidade oral e úlceras rasas na genitália também faziam parte do quadro. O anatomopatológico foi bastante sugestivo de queratose liquenoide crônica. Introduziu-se tratamento com acitretina e dapsona, havendo melhora parcial do quadro.

Palavras-chave: Ceratose; Dermatopatias; Erupções liquenóides

INTRODUCTION

Keratosis lichenoides chronica (KLC), also known as Nekam's disease, is a rare mucocutaneous skin disease of unknown etiology characterized by keratinization with a chronic, progressive course, and usually affecting people aged between 20 and 50. Differences of prevalence between genders and races have been reported.¹ To date, over 60 cases have been reported.² Controversy exists regarding the classification of acne as a distinct nosological entity. Some authors consider it to be the expression of a known

inflammatory disease such as lichen planus, lupus erythematosus or lichen simplex chronicus.² KLC was first described by Kaposi in 1895, who named the disease *lichen planus* and *lichen ruber acuminatus morbilliform* disease.³ In 1938, Nekam observed acrosyringial hyperkeratosis in the case published by Kaposi, which led him to call the disease *Porokeratosis Striate Lichen*, despite the absence of cornoid lamella.² The name *Keratosis Lichenoides Chronica*, introduced in 1972 by Margolis et al., has

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¹ Resident Medical Doctor, Department of Dermatology, Professor at the Edgard Santos University Hospital, Federal University of Bahia (HUPES - UFBA), Salvador, Brazil.

² Resident Medical Doctor, Department of Pathology, Professor Edgard Santos University Hospital, Federal University of Bahia (HUPES - UFBA), Salvador, Brazil.

³ PhD in Medicine and Health Sciences awarded by the Federal University of Bahia (UFBA), Medical Assistant, Department of Dermatology, Professor Edgard Santos University Hospital, Federal University of Bahia (HUPES - UFBA), Salvador, Brazil.

⁴ MA in Medicine and Health Sciences awarded by the Federal University of Bahia (UFBA), Head of the Department of Dermatology, Professor Edgard Santos University Hospital, Federal University of Bahia (HUPES - UFBA), Salvador, Brazil.

been widely employed since then.¹ The following report describes an extensive case of the disease, of difficult diagnosis and involving oral mucosa and genitalia, with a number of atypical features.

CASE REPORT

42-year-old male patient, chef, married, born in Agua Fria in the state of Bahia and resident in the state capital Salvador. On admission, he reported the appearance of "rash-like" lesions on the trunk over the previous 5 years. The lesions were non-pruritic, which had partially involuted after some unspecified topical treatment. Three years later the symptoms became more exuberant when he was treated with dapsone over a period of four months, with some slight improvement. The patient reported having also used topically a salicylic acid and betamethasone dipropionate combination, as well as calcipotriol and emollients, but without response. He also reported the emergence of oral aphthous lesions and genital ulceration 2 years after onset of the symptoms, experiencing periods of remission and relapse ever since. He reported no comorbidities or similar cases in his family, or alcoholism/smoking. The dermatological examination detected violaceous hyperkeratotic papules, confluent in some areas, in a linear form with tracery lines and plaques, located predominantly on the trunk and the upper and backs of the patient's lower limbs (Figures 1 and 2). Also aphthous lesions in the oral cavity and shallow ulcers on the penis and scrotum, with an erythematous base (Figures 3 and 4). Laboratory tests (blood count, liver and kidney functions, urinalysis and rheumatologic markers, etc) were normal. The patient was submitted to the first



FIGURE 2: Violaceous hyperkeratotic papules, confluent, linear aspect; tracery lines

anatomopathological examination in March 2007, which suggested psoriasiform dermatitis associated with folliculitis. In November 2008 a new biopsy concluded folliculitis. At that point we started treatment with dapsone 100mg/day, resulting in regression of genital ulceration and mild improvement of skin lesions. In February 2009 the two biopsies were reviewed, both of which maintained the previous findings of acute abscess folliculitis. In November 2009 a new anatomopathological examination showed marked parakeratosis and epidermal acanthosis, with areas of vacuolar alteration of the basal layer and corneous plugs (Figure 5). In the superficial dermis we observed a dense linfhistoplasmosis infiltration. We also noted a well-defined dermoepidermal papule with parakerato-



FIGURE 1: Violaceous hyperkeratotic papules forming plaques on the back and upper limbs



FIGURE 3: Aphthous lesion in upper lip mucosa



FIGURE 4: Shallow ulcers on the penis and scrotum, with an erythematous base

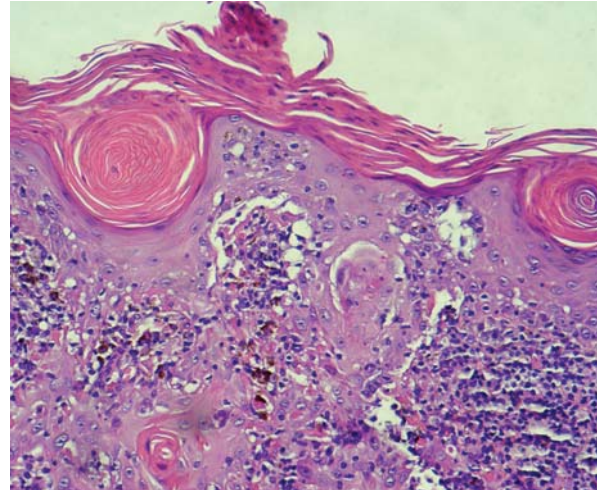


FIGURE 5: Parakeratosis and acanthosis, with areas of vacuolar alteration of basal layer and cornal plugs and with dense lymphohistoplasmosis infiltrate in the superficial dermis

sis and moderate epidermal hyperplasia with multiple apoptotic bodies and vacuolar alteration of the basal layer. Conclusion: Keratosis lichenoides chronica. On that occasion we opted for the introduction of acitretin 25mg/day, which produced significant improvement of the skin lesions. However, the mucosal lesions relapsed after discontinuation of dapsone, which has since been reintroduced and maintained in conjunction with acitretin. NB-UVB has been recently added in order to optimize the therapy.

DISCUSSION

Around 70 cases of dermatitis lichenoides chronica (DLC) have been reported.¹ It occurs most commonly in adolescents and young adults, with a slight predominance in males (1.35:1).³ Clinically, the disease is characterized by violaceous hyperkeratotic papules and plaques, generally symmetrically arranged in a linear or reticular pattern on the trunk and extremities.¹ In 75% of cases, rash-like or rosacea-like facial seborrheic dermatitis is present which facilitates diagnosis.^{1,4} Ungueal dystrophy^{1,4} may be seen in 30% of these patients and palmoplantar keratoderma in about 40%.^{3,5} The most common nail changes are yellowing, thickening and hyperkeratosis of the nail bed.^{3,6} This skin problem can also be associated with oral manifestations in 50% of cases presenting as recurrent aphthous ulcers and lesions.⁶ In relation to genital lesions, keratotic papules can be found on the scrotum and penis as well as chronic balanitis and phimosis. The eyes can also be affected, with the most common problems being blepharitis, conjunctivitis, uveitis and iridocyclitis.⁶ Reports exist of association with glomerulonephritis and lymphoproliferative dis-

orders. In rare cases hoarseness, due to infiltration of the epiglottis, can be observed.⁷ The histopathological findings of keratosis lichenoides chronica vary. These typically refer to the presence of lichenoid dermatitis (lymphocytic infiltration in striped bars), associated with numerous necrotic keratinocytes, focal parakeratosis, neutrophils remaining in the corneal layer, irregular acanthosis and corneal plugs.^{1,2} The lichenoid infiltrate is often perinfundibular or around the acrosyringial areas. There may be vacuolar alteration of the basal layer along the dermoepidermal junction. The granular layer may increase or decrease: the first phenomenon is linked to areas of acanthosis and the second to areas of hyperkeratosis. Atrophy or erosion of the epithelium may be present, along with necrotic keratinocytes.⁸ KLC is chronic and progressive. Its treatment represents a significant challenge because it is extremely resistant to available treatments. Topical treatments are usually ineffective, as well as treatment with systemic steroids, sulfones, methotrexate, anti-malarial agents, radiation therapy and cyclosporine.^{5,6,9} Acitretin, isotretinoin, etretinate, psoralen and UVA (PUVA), Re-PUVA and topical calcipotriol have all been used, with different results.⁶ A few reports exist of good results with re-PUVA, acitretin-UVB + NB and also with efalizumab, but further studies are called for regarding these therapies.^{5,6,9} The patient in question is a rare case, difficult to diagnose, because of the lack of pathologic substrate compatible with the disease in the first biopsies carried out and the lack of facial involvement of the disease as is normally present in most other cases. However the other clinical and, subsequently,

histopathological characteristics, together with the exclusion of possible differential diagnoses (lichen planus, pityriasis rubra pilaris, mycosis fungoides and folliculotropic lichenoid eruption), make it essential to diagnose KLC.^{2,8} Moreover, it is worth noting that our patient developed genital ulcerations, an atypical manifestation of the genital tract which has not yet been recorded in the literature. Despite being a rare skin disease, it is important to become familiar with KLC, which involves different diagnosis to that of the very prevalent lichen

planus. Some authors have considered that this could even be a rare presentation of lichen planus that could evolve in the same way as Graham-Little-Lassuer syndrome.¹⁰ The clinical and histopathological characteristics are similar in both diseases and the linear pattern of KLC can be confused with the Koebner phenomenon of lichen planus.^{3,5} However the symmetry, the absence of intense pruritus and the resistance to treatment of KLC should call for a keratosis lichenoides chronica diagnosis. □

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MAILING ADDRESS / ENDEREÇO PARA CORRESPONDÊNCIA:

Livia Cibele Gomes Martins
Ambulatório Magalhães Neto
Rua Padre Feijó, 240 – 3º andar, Canela
40110-170 Salvador (BA) - Brazil
E-mail: liviacibele@botmail.com

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