Portuguese Journal of Dermatology and Venereology

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CASE REPORT

Chronic lichenoid keratosis with good response to oral acitretin: a case report

Queratose liquenoide crônica com boa resposta a acitretina oral: relato de um caso

Mariana S. Bernardes[1], Jéssica C. N. Araújo[1], Luísa L. Macias[1], Laísa E. De Hollanda[1], Joelly T. L. Levermann[1], Alcidarta R. Gadelha[2], Patricia C. B. De Melo[1], and Luciana M. Dos Santos[1]*

[1]Department of Dermatology, Hospital Universitário Getúlio Vargas, Manaus; [2]Department of Dermatology, Fundação de Medicina Tropical Doutor Heitor Vieira Dourado. Manaus, Brazil

Abstract

Keratosis lichenoides chronica or Nekam's disease is a rare disease of unknown etiology and pathogenesis, with few cases described in the literature. The lichenoid-like lesions as well as their histopathological findings require a wide range of differential diagnoses. Its chronic, progressive course and refractoriness to topical and systemic therapies are hallmarks of this dermatosis. The aim of this report is to describe the case of a patient with typical cutaneous manifestations and compatible histopathological findings with a good response to acitretin.

***Keywords*:** Keratosis. Lichenoid eruptions. Skin diseases. Retinoids.

Resumo

A queratose liquenóide crônica ou doença de Nekam é uma doença rara, de etiologia e patogênese desconhecida, com poucos casos descritos na literatura. As lesões de aspecto liquenóide assim como seus achados histopatológicos requerem um amplo leque de diagnósticos diferenciais. O seu curso crônico, progressivo e a refratariedade às terapias tópicas e sistêmicas são características marcantes dessa dermatose. O objetivo deste relato é descrever o caso de um paciente com manifestações cutâneas típicas e achados histopatológicos compatíveis com boa resposta à acitretina.

***Palavras-chave*:** Queratoses. Erupções liquenoides. Dermatopatias. Retinoides.

Introduction

Keratosis lichenoides chronica (KLC) or Nekam's disease is a rare disease of unknown etiology, a chronic course, and progressive character, with less than 150 cases published to date. Clinically, it presents with lichenoid papules arranged linearly or in a reticulated pattern on the extremities and trunk, together with facial erythema in a seborrheic distribution, oral erosions, nail, and genital involvement[1,2]. Due to the rarity of the cases and unknown pathophysiology, it becomes a therapeutic challenge.

The aim of this report is to describe the case of a patient with typical cutaneous manifestations and compatible histopathological findings, with a good response to acitretin.

***Corresponding author:**
Mariana Santiago Bernardes
E-mail: marianasanber@gmail.com

Received: 14-09-2022
Accepted: 03-10-2022
DOI: 10.24875/PJDV.22000018

Available online: 09-12-2022
Port J Dermatol and Venereol. 2022;80(4):317-321
www.portuguesejournalofdermatology.com

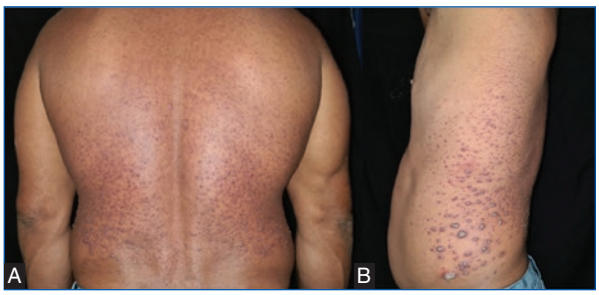

Figure 1. A: violaceous hyperkeratotic papules forming plaques on the dorsum. **B:** linear distribution of lesions with the confluence of some generating a reticulated appearance.

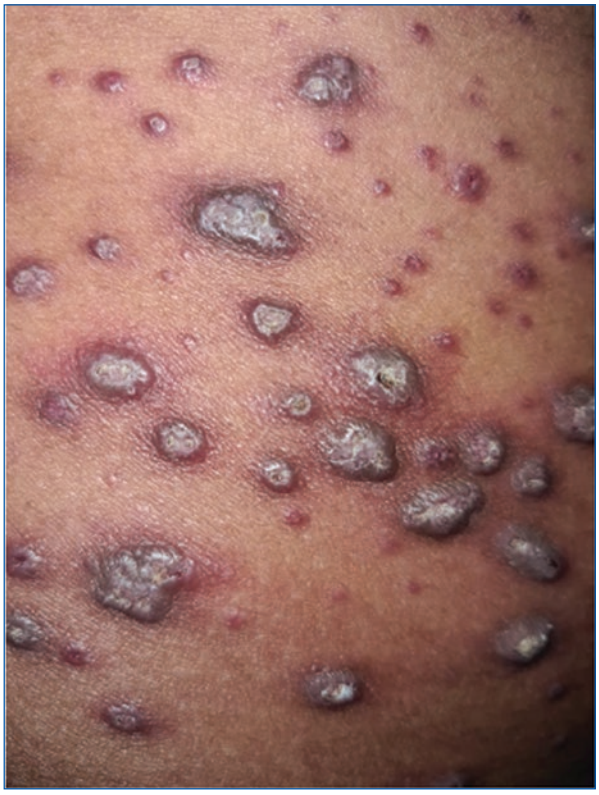

Figure 2. Detail of violaceous, confluent, hyperkeratotic papules in a linear and reticulated pattern.

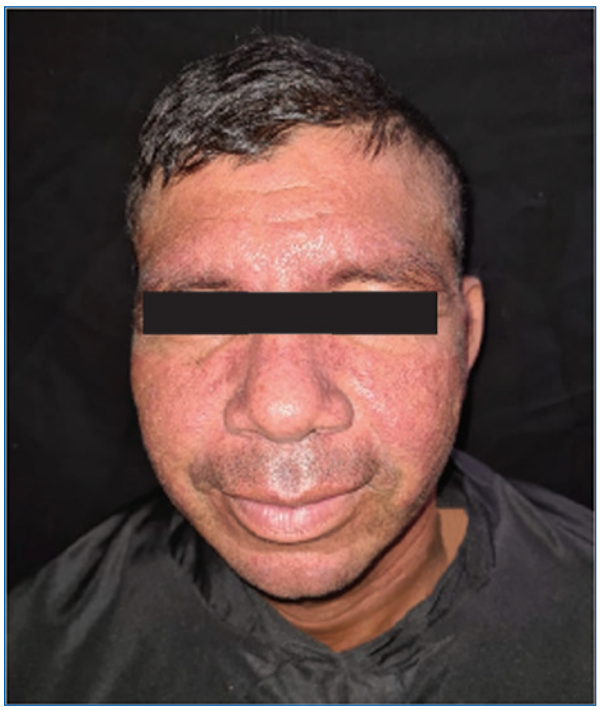

Figure 3. Diffusely infiltrated facial lesions with diffuse erythema and telangiectasia affecting predominantly convex areas.

Case report

A 42-year-old male patient, born and living in the municipality of Tefé, in the interior of Amazonas State (Brazil), complained for approximately 12 years of hyperkeratotic, pruritic, erythematous papules and plaques, some arranged in a parallel linear pattern and others in a reticular shape, symmetrically distributed on the back (Figures 1 and 2). Dermatological examination also showed erythema, edema, and facial telangiectasia with a rosacea-like appearance (Figure 3). Laboratory tests, including blood count, liver and kidney function, rheumatologic markers, serologies for human immunodeficiency virus (HIV), syphilis, hepatitis B and C, and urinalysis, were unaltered. An incisional biopsy was performed, and the anatomopathological examination revealed a hyperkeratotic epidermis, with foci of parakeratosis, large ortho- and parakeratotic corneal plugs, moderate irregular acanthosis, marked vacuolar degeneration of the basal layer, and rare individually necrotic keratinocytes (Figure 4). In the dermis, there were numerous lumps of melanin, free and within macrophages, and moderate perivascular, perifollicular, and interstitial infiltrate composed of lymphocytes, histiocytes, melanophages, and macrophages (Figure 5). Within the differential diagnoses of a lichenoid eruption, lichen planus (LP), and chronic cutaneous lupus were investigated and excluded. Once the hypothesis of KLC was raised, acitretin 30 mg/day (approximately 0.3 mg/Kg/day) was introduced, with progressive improvement of erythema and pruritus in 90 days of treatment (Figure 6).

Discussion

Nekam's disease or KLC is a relatively rare disease, that was first described by Kaposi in 1886 as "lichen ruber moniliformis." However, in 1895, two patients with

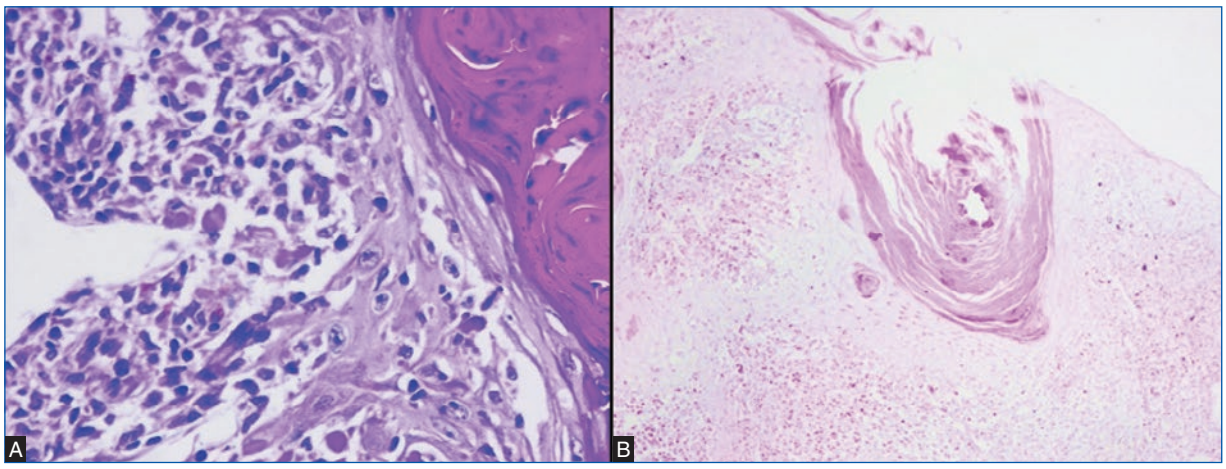

Figure 4. **A:** intense vacuolization of the basal layer (HE, ×400). **B:** large corneal stoppers (HE, ×100).

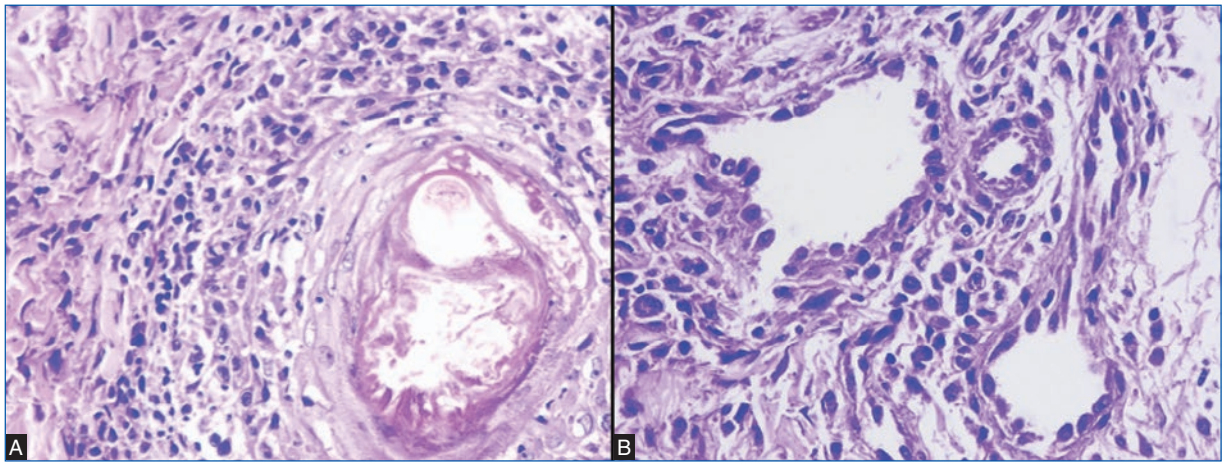

Figure 5. **A:** perifollicular inflammatory infiltrate, invading the follicle wall, with vacuolar degeneration of the basal layer (HE, ×400). **B:** infiltrate with plasma cells around dilated vessels and entombed endothelium (HE, ×400).

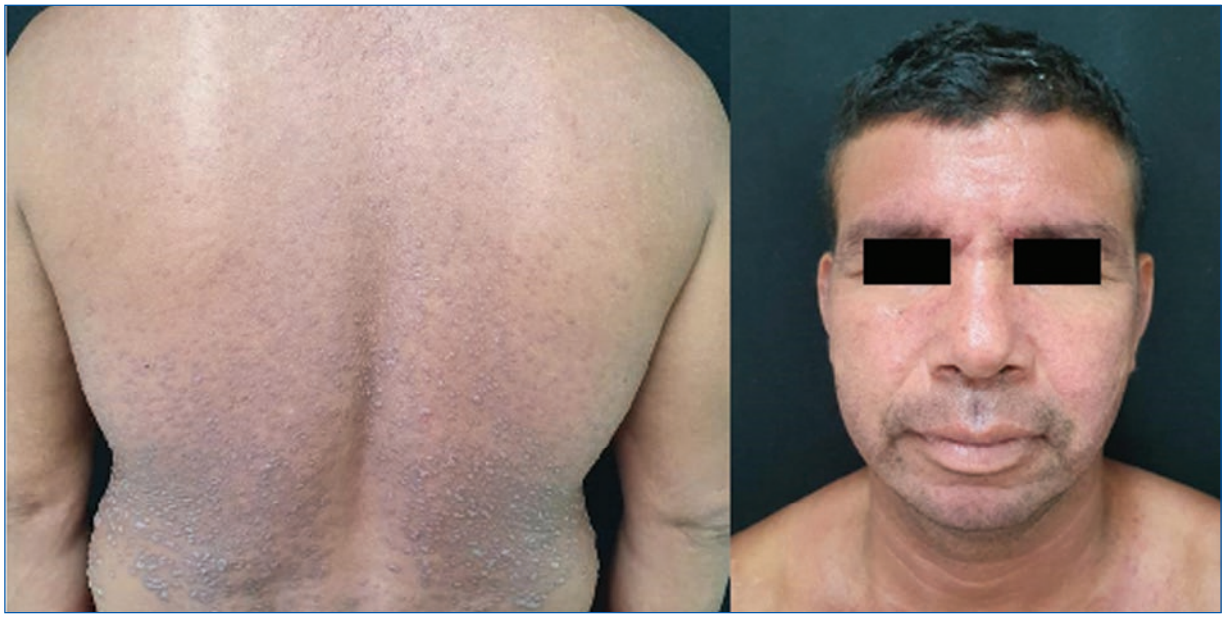

Figure 6. Improvement of trunk and face erythema and reduced lesion thickness after 3 months of continuous use of acitretin.

similar lesions were diagnosed with "lichen ruber acuminatum (verrucosus et reticularis)"[1]. In 1938, Nekam observed acrosyngeal hyperkeratosis in the case published by Kaposi, which caused the disease to be called lichenoid streaky porokeratosis, despite the absence of a horny lamella[2]. Since then, it has been reported under different nomenclatures, among them striated porokeratosis, verrucous and reticular lichen, lichenoid triceratosis and strieé lichenoid keratosis[3]. The widely used term KLC was introduced by Margolis et al. in 1972[2].

KLC is an acquired, chronic, progressive mucocutaneous dermatosis that occurs most commonly in adolescents and young adults, with a slight male predominance (1.35:1)[4,5]. It usually appears between the third and fifth decade of life, with a mean age of 35.6 years at diagnosis[6]. However, there are reports in children, some with a possible genetic association with an autosomal recessive pattern[7]. The mean time between onset and diagnosis was 9 years, with only 30% of patients being correctly diagnosed within 2 years of onset, 20% within 5 years, and 50% after more than 5 years[6].

The exact etiopathology of this disease remains unknown and no significant association with underlying diseases has been proven, but some factors such as trauma, autoimmunity, infections, drugs, infrared radiation, inflammatory diseases, and hematological malignancies have been reported concomitantly[6,8]. Some authors suggest an immunological basis in the epidermis, probably autoimmune[4]. Clinically, KLC is characterized by hyperkeratotic erythematous-violaceous papules and plaques in a linear or reticular pattern, arranged symmetrically mainly on the trunk and extremities[5,9]. Pruritus is a variable finding, being present in less than 20% of patients[10]. A mid-facial erythematous eruption, seborrheic dermatitis-simile, or rosacea-simile, as observed in our patient, may be present in 75% of cases[5,9]. Palmoplantar keratoderma may be evident in approximately 40% of patients[11].

Nail, oral, and genital involvement is frequently seen in adults, whereas alopecia and pruritus are frequent in children[8]. Nail dystrophy may be noted in 30% of cases[12], the most frequent changes include yellowish chromonychia, lamina thickening, and hyperkeratosis of the nail bed[5,8,9]. Oral manifestations, present in 50% of cases, include ulcers and recurrent aphthous lesions. As for genital lesions, keratotic papules may occur on the scrotum and penis, in addition to chronic balanitis and phimosis. Ocular involvement encompasses blepharitis, keratoconjunctivitis, anterior uveitis, and iridocyclitis, as the most common, sometimes leading to visual impairment[13].

Epidermal changes include hyperkeratosis, focal parakeratosis, especial/y in the follicular openings, irregular acanthosis with areas of atrophy, and corneal stoppers. Vacuolar degeneration of the basal layer is a frequent finding. A chronic inflammatory infiltrate usually consisting of lymphocytes, histiocytes, plasma cells, a few eosinophils, and numerous Civatte bodies (necrotic keratinocytes) is observed in the upper dermis. The lichenoid infiltrate is often centered around an infundibulum or acrosyringe[1,3,6,10,14].

Although there is no definitive laboratory test for the diagnosis of this entity, clinical and histopathological clues are sufficient to make the diagnosis[9].

For a long time, KLC was considered a variant of LP that evolved by antigen mimicry followed by epitope dissemination. Today, many authors consider it to be a distinct entity[2].

Other conditions may resemble KLC and should be differentiated, among them chronic cutaneous lupus erythematosus, pityriasis lichenoides, pityriasis rubra pilaris, psoriasis, porokeratosis and mycosis fungoides, including paraceratosis variegata and folliculocentric mycosis fungoides[10].

KLC is a therapeutic challenge, showing resistance to a variety of treatments including topical and systemic corticosteroids, antimalarials, dapsone, tetracyclines, methotrexate, and cyclosporine[1,4,6,9,15]. Varied results have been observed in psoralene with ultraviolet A radiation (PUVA), systemic retinoids, either etretinate alone or combined with PUVA or with narrow-band ultraviolet B radiation (NB-UVB), as well as topical calcipotriol and NB-UVB monotherapy. NB-UVB has proven to be more effective in treating children than adults[1]. Oral retinoids, isotretinoin and especially acitretin, given at a dose of 0.3-0.6 mg/Kg/day, have been shown to be the most effective option, with a rapid onset of action, in approximately 1-2 months. Flattening of the lesions is observed as early as the first month, with significant improvement expected in 4-6 months after starting treatment[6,8], similarly to we observed in our patient. A review of 30 patients diagnosed with KLC and treated with oral retinoids showed partial response in 20% and complete response in 36.6% of patients[6].

KLC is a difficult disease to diagnose, considering its clinical and histological similarity with other dermatoses and also its rarity or possible underdiagnosis, since the findings can be mistaken with other pathologies.

Although it has a benign course, lesions cause significant psychosocial damage, and effective therapeutic strategies should be sought despite the lack of consensus in the literature.

Funding

None.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that no patient data appear in this article.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

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