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Keratosis lichenoides chronica: First case reported in Chile

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Abstract

We present a woman with a history of years of evolution of confluent hyperkeratotic papules and plaques with a generalized linear and reticulate pattern. Histopathological characteristics concordant with keratosis lichenoides chronica were finally evidenced after several non-specific biopsies. The cutaneous manifestations, chronicity, histopathology findings, and refractoriness to therapies are typical of this rare dermatosis.

Keywords: keratinization disorders, keratosis lichenoides chronica, Nekam disease

Introduction

Keratosis lichenoides chronica (KLC) or Nekam disease is an uncommon inflammatory disorder of keratinization with a poorly elucidated pathogenesis and few cases reported in the literature. It was described for the first time by Kaposi in 1895. In 1938, Nekam called the disease lichenoid porokeratosis [1], but Margolis formally proposed the term KLC in 1972 [2]. Clinically, the primary lesions are erythematous-violaceous hyperkeratotic papules similar to lichen planus, which, as they increase in number, converge to form warty plaques. Lesions begin on the limbs and then extend to the trunk and the rest of the body in a linear or reticulate pattern [3, 4].

Case Synopsis

A 67-years-old woman, presented with a 30-year history of an eruption characterized by red-to-purple papules and plaques located on the face, trunk, and limbs. The patient denied a family history of skin conditions. She was treated for years as having psoriasis and/or verrucous lichen planus.

On physical examination, erythematous-violaceous papules were found symmetrically distributed in a linear or reticulate pattern, with a warty appearance and a thin and superficial whitish scale. On the face, there were lesions predominantly on the eyebrows, forehead (**Figure 1A)**, cheeks, and ears (**Figure 1B)**, respecting the perioral area. Some erythematous papules were found on the hard palate. On the trunk and limbs, a linear and reticulate distribution of the lesions were seen, particularly on the lower abdominal area and along the upper limbs (Figure.2-A). Multiple desquamative papules were also symmetrically seen on the dorsal and ventral areas of the hands. In the extensor region of the thighs and legs, erythematous to dark purple-blue nodules were found (**Figure 2B**).

On the external area of the right leg, dermoscopy showed a desquamative plaque, with irregular

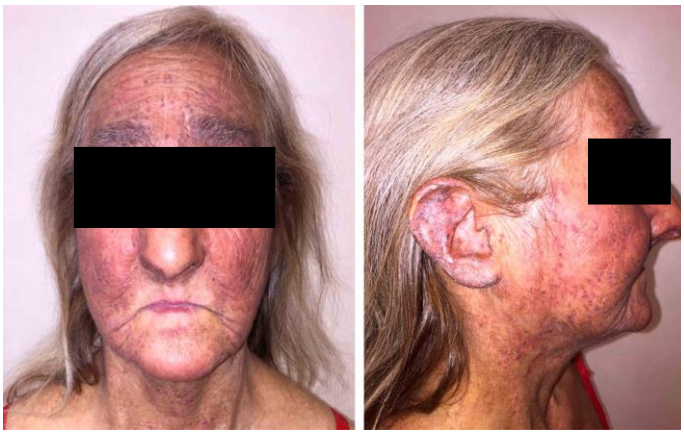

Figure 1. ***A)*** *Hyperkeratotic and erythematous-violaceous papules, located in the face, neck, and chest.* ***B)*** *Confluent hyperkeratotic and erythematous-violaceous papules in the nose, cheeks, and superior part of the neck, associated to erythematous-desquamating papules in both ears.*

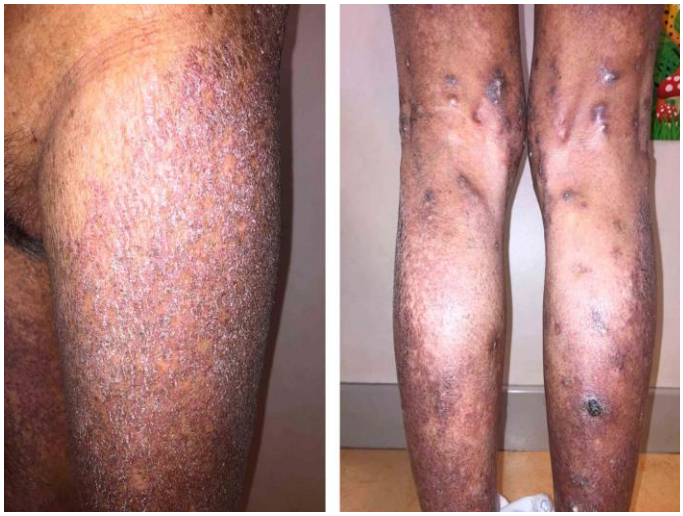

Figure 2. ***A)*** *Converging erythematous-violaceous papules, with linear and reticulate shape in the arms, with a thin scale.* ***B)*** *Erythematous papules in reticulate pattern with desquamative purpuric nodules in the posterior area of lower limbs.*

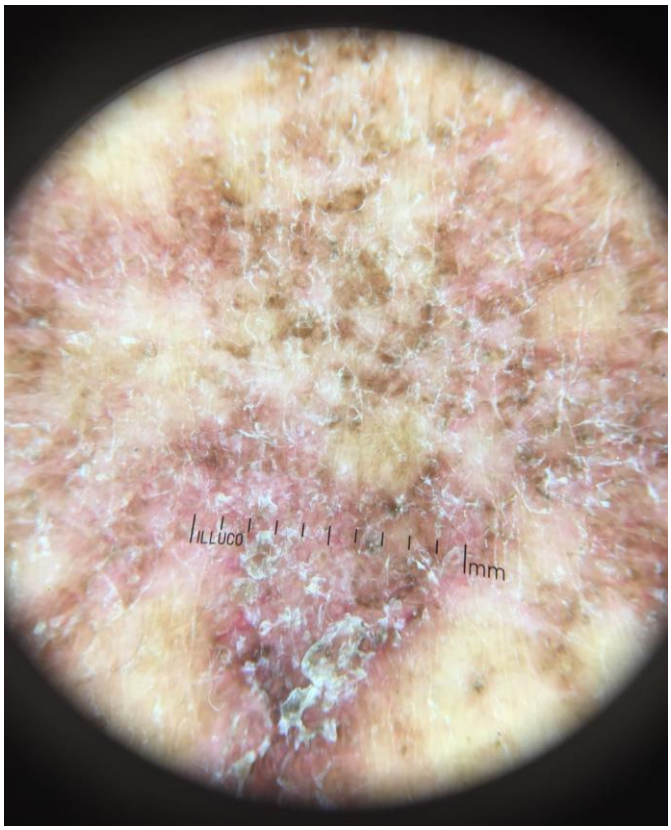

Figure 3. *Dermoscopy of an erythematous-desquamating plaque, in reticulate pattern. Hyperpigmentation and fine whitish scales are seen in an erythematous base.*

edges, arranged in a hyperpigmented reticular pattern with superficial fine white scales on an erythematous-pink base (**Figure 3**). Routine laboratory tests, serological tests for human immunodeficiency virus type 1, hepatitis viruses, and nontreponemal tests for syphilis, were within normal limits.

Initially, histopathology revealed suggestive findings of psoriasiform dermatitis; five years later, the biopsy pointed to hypertrophic lichen planus. Therefore, the patient received treatment with oral methotrexate associated with phototherapy for years without apparent improvement. Recently, a biopsy was obtained from the left leg. The epidermis showed irregular acanthosis, hyperkeratosis, and focal parakeratosis. In addition, vacuolar variation of the stratum basale of the epidermis with telangiectasias and abundant inflammatory infiltrate, including lymphocytes and plasma cells was demonstrated (**Figure 4**). Direct and indirect immunofluorescence was negative.

The diagnosis of KLC was based on clinical findings, multiple histopathological studies, and the chronic course of the disease associated with poor response to therapies. The differential diagnosis included psoriasis, lichen planus, and lupus erythematosus, which were excluded based on the last histopathology and immunofluorescence tests.

Case Discussion

Keratosis lichenoides chronica usually appears in adults between the third and the fifth decade of life. It is considered as an acquired and usually persistent disease [3]. However, there are reports in children, some with possible family association and with an autosomal recessive pattern [5].

Recently, it was discovered that a mutation in the inflammasome sensor protein, NLR family, was the genetic cause in a family with KLC. Nucleotide-binding domain and leucine-rich repeat containing family pyrin domain containing 1 (NLRP1) is the most prominent inflammasome determinant in human skin, and all pathogenic *NLRP1* mutations are gain-of-function alleles that predispose to inflammasome activation. In this context, we now consider that autoinflammatory mechanisms play an important role in the pathogenesis of KLC, at least in familial KLC [6].

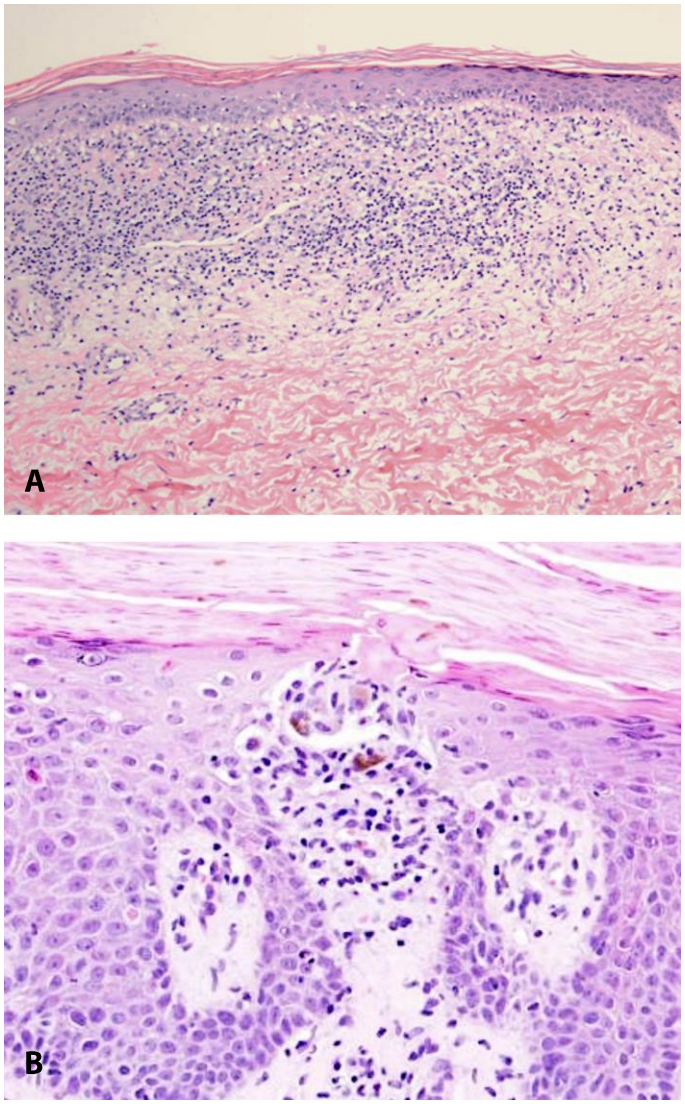

Figure 4. ***A)*** *Areas of focal hyperparakeratosis and orthokeratosis, moderate epitelial hiperplasia and vacuolar degeneration of the stratum basale, with pigmentary incontinence, presence of melanophages, and isolated superficial perivascular lymphocytic inflammatory infiltrate, H&E, 4×.* ***B)*** *Epidermis with hyperkeratosis and hypergranulosis, acanthosis and a large vacuolar degeneration of the stratum basale with dermo-epidermal excision, presence of lymphocytic inflammatory infiltrate and focal hemorrhage. H&E, 40×.*

Facial involvement, simulating rosacea or seborrheic dermatitis has been described in 75% of the cases. Mucosal involvement is frequent; recurrent aphthous ulcerations, whitish papules, soft palate infiltration, epiglottis, and ocular involvement have been reported [4]. Nail dystrophies occur in 30% of patients [7].

Histopathologically, KLC is characterized by vacuolar degeneration of keratinocytes in the dermoepidermal junction, numerous necrotic keratinocytes in the superficial and infundibular epidermis, chronic inflammatory infiltrate in the papillary dermis consisting of lymphocytes, histiocytes, and plasma cells, and vascular dilatation. The granular layer may be more or less prominent, depending on the presence of acanthosis or hyperparakeratosis. Direct immunofluorescence is typically negative [8].

Topical agents are usually ineffective and systemic agents such as corticosteroids, dapsone, methotrexate, retinoids, and phototherapy can be alternatives for treatment [9, 10]. There are reports of KLC successfully treated with monotherapy NB-UVB and efalizumab [11-13].

Conclusion

Nekam disease is a rare dermatosis. When a patient presents with a group of characteristics consistent with lichen planus, seborrhoeic dermatitis, and aphthous ulcers with poor response to treatment, KLC should be considered in the differential diagnosis.

Potential conflicts of interest

The authors declare no conflicts of interests.

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