

Unilateral Keratosis Lichenoides Chronica

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Abstract

Observation: Keratosis lichenoides chronica (KLC) or Nekam's disease is a rare, chronic and progressive dermatosis. Around 70 cases have been reported in the literature. It is characterized by violaceous hyperkeratotic papules and plaques that are generally symmetrically arranged in a linear or reticular pattern on the trunk and extremities. The lesions are usually bilateral. KLC is chronic, progressive and medically resistant course.

Herein, we report a 27-year old presenting with thick hyperkeratotic plaques, some of them evolved into annular shapes, on a background of violaceous erythema along lines of Blaschko extending from the dorsum of the left foot to the left inguinal region. A plaque of 5 cm in diameter located on left side of his trunk and another plaque of 4 cm in diameter on the left proximal upper extremity. A diagnosis of Nekam's disease was clinically and histologically features. Oral acitretin treatment was started, with partially improved. We report a patient showing KLC unilateral distribution and covering several areas exclusively on the left side of his body.

Introduction

Keratosis lichenoides chronic (KLC) or Nekam's disease is a rare, chronic and progressive dermatosis. The disease was first described by *Kaposi* in 1895 as lichen ruber verrucosus et reticularis. It was named after *Nekam* who reported a typical case in 1938. The current descriptive term was introduced in 1972, and the disease is generally thought to represent a special form of lichen planus [1]. Around 70 cases have been reported in the literature [2]. The course of KLC is chronic and progressive. The disease is very resistant to treatment. It occurs most commonly in adolescents and young adults, with a slight predominance in males. It is characterized by violaceous hyperkeratotic papules and plaques that are ge-

nerally symmetrically arranged in a linear or reticular pattern on the trunk and extremities. The lesions are usually bilateral. KLC is chronic, progressive and medically resistant course [2,3].

In the case described we present a 27-year old-male patient with unilateral keratosis lichenoides chronica.

Case Report

A 27-year-old male patient was admitted to our clinic with the complaint of scaling on his left leg. He explained that his skin lesions had been present since shortly after birth and had progressed slowly thereafter. No family history of a similar eruption



Figure 1. Multiple keratotic papules on the lower limbs

was found. He was taking no medication, and had not been previously treated. Systemic examination was unremarkable.

Dermatologic examination showed thick hyperkeratotic plaques, some evolving into annular shapes, on a background of violaceous erythema along lines of Blascko extending from the dorsum of the left foot to the left inguinal region, a plaque of 5 cm in diameter on left side of his trunk and another plaque of 4 cm in diameter on the left proximal upper extremity (**Figures 1 and 2**).

Complete blood count, blood biochemistry and urinalysis were within normal limits. Histopathology of biopsy obtained from a plaque showed parakeratosis, basket-weave hyperkeratosis, papillomatosis, follicular plugging, wedge-shaped mild hypergranulosis, irregular acanthosis, mild spongiosis, scarce exocytosis of lymphocytes, necrotic keratinocytes, focal vacuolar degeneration, damage of the basal layer, mild edema, pigment incontinence and numerous melanophages in the upper dermis, an increase in the number of capillary vessels and moderate-to-severe perivascular lymphocytic infiltration in the dermis (**Figure 3**). The patient was diagnosed with Nekam's disease based on clinical and histological features.

He was started on oral acitretin 25 mg/day and an ointment containing mometasone furoate plus salicylic acid 5%. At the end of 2-month of the treatment period hyperkeratosis was partially improved (**Figure 4**).



Figure 2. Erythematous keratotic left proximal upper extremity

Discussion

KLC is a rare dermatosis characterized by violaceous papular and nodular lesions, often arranged in a linear and reticulate pattern on the dorsal hands and feet, extremities, and buttocks. The mucous membranes, genitalia, nails, palms, and soles may also be affected [1]. The elementary lesions can converge to form warty plaques. These keratotic structures are dry, purple to brown, and sometimes telangiectatic. The papules of the trunk are classically thinner. Moreover, in 75% of the cases, an erythematous eruption on the mediobuccal area is found, mimicking seborrheic dermatitis [4]. It commonly affects adults aged 20–50 years. There are no known differences in prevalence between genders or races [2,3]. KLC of pediatric onset is considered extremely rare. Our knowledge, only 14 reported cases have been present in childhood or started during infancy [5,6]. In adults, KLC developed at a mean age of 28.5 years, 9 while KLC of pediatric onset was congenital in 3 patients and started during the first year of life in all other cases except one (2 years of age) [5]. Our patient has these lesions since his birth, and location of his lesions are unilaterally and covering several areas exclusively on the left side of his body.

As KLC similar clinical and histological characteristics with lichen planus, some authors have considered KLC to be a variant of lichen planus. However, other some authors suggested that it was a distinct entity characterized by lichenoid hyperkeratotic papules arranged

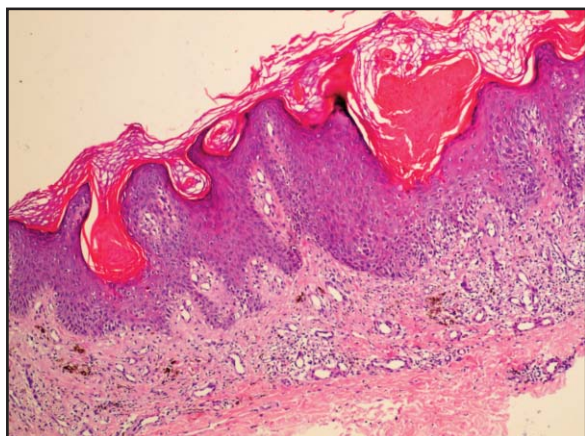


Figure 3 . Section showing focal parakeratosis and follicular plugging in the hyperkeratotic cornified layer; acanthosis, vacuolar changes and necrotic keratinocytes in the epidermis; lichenoid inflammatory infiltrate with melanophages in the the dermo-epidermal junction and dilated capillary vessels in the dermis (H & E stain, x10)

in a linear pattern, erythematous squamous plaques and seborrheic dermatitis on the face, the absence of intense pruritus and the resistance to treatment of KLC [2,7]. Our patient hasn't got intense pruritus.

In 50 percent of cases adult onset KLC, the disorder involves the oral or genital mucous membranes, presenting as ulcerations, infiltration or inflammation. The ocular area and nails may be involved [1,5]. In the patients of pediatric onset of KLC haven't got oral, genital, eye and nail lesions [5]. Our patient hasn't got oral, genital, nail and eye involvement too.

Histological examination of KLC reveals lichenoid dermatitis with hyperkeratosis, focal parakeratosis, numerous necrotic keratinocytes, irregular acanthosis and corneal plugs, a band-like inflammatory subepidermal infiltrate and colloid bodies. The granular layer may increase or decrease. There may be vacuolar alteration of the basal layer along the dermoepidermal junction [2,8]. The histopathologic findings of our patient are compatible with KLC.

The differential diagnosis of the disease includes lichen planus, folliculotropic lichenoid eruption, pityriasis rubra pilaris, mycosis fungoides and drug eruptions [2,3]. The characteristic linear arrangement of the lesions, facial involvement, typical oral lesions, absence of Wickham's striae, the long-term evo-



Figure 4 . The appearance of lesions after treatment

lution, and ineffectiveness of systemic corticosteroids are useful features that help in the differentiation of KLC and lichen planus [3,5]. A diagnosis of KLC was made, based upon typical clinical and histopathological findings.

Therapy with keratolytic agents, tar, glucocorticoids, antimalarial agents, sulfones, gold, superficial x-ray therapy, and cyclosporine is usually ineffective [1,8]. Successful treatment has been reported with oral PUVA, bath-PUVA, oral retinoids (acitretin or isotretinoin) and topical calcipotriol [7,8,9]. Our patients received oral acitretin 25 mg/day and an ointment containing mometasone furoate plus salicylic acid 5%. His lesions cured partially.

References

1. Douri T, Shawaf AZ. Keratosis lichenoides chronica: report of a new case with partial response to PUVA therapy. *Dermatol Online J* 2005; 11: 28. PMID: 16150236
2. Martins LC, Horne M, Moreira Júnior DN, Follador I, Almeida VR. Keratosis lichenoides chronica--case report. *An Bras Dermatol* 2011; 86: 148-151. PMID: 22068797
3. Adışen E, Erdem O, Celepçi S, Gürer MA. Easy to diagnose, difficult to treat: keratosis lichenoides chro-

- nica. Clin Exp Dermatol 2010; 35: 47-50. PMID: 19196302
4. Ghislain PD, Beir AD, Creusy C, Modiano P. Keratosis lichenoides chronica: report of a new case, with success of PUVA therapy. Dermatology Online J 2001; 7: 4. PMID: 11328625
 5. Ruiz-Maldonado R, Duran-McKinster C, Orozco-Covarrubias L, Saez-de-Ocariz M, Palacios-Lopez C. Keratosis lichenoides chronica in pediatric patients: a different disease? J Am Acad Dermatol 2007; 56: S1-5. PMID: 17224382
 6. Redondo P, Solano T. Keratosis lichenoides chronica in childhood. Clin Exp Dermatol 2002; 27: 283-285. PMID: 12139671
 7. Demirci E, Boyvat A, Arica IE, Kocyigit P, Ozdemir E, Heper AO. Keratosis lichenoides chronica: marked response to PUVA in combination with acitretin. Acta Derm Venereol 2005; 85: 552-553. PMID: 16396816
 8. Kunte C, Kerschenlohr K, Röcken M, Schirren C. Keratosis lichenoides chronica: treatment with bath-PUVA. Acta Derm Venereol 2007; 87: 182-183. PMID: 17340035
 9. Chang SE, Jung EC, Hong SM, Choi JH, Sung KJ, Moon KC, Koh JK. Keratosis lichenoides chronica: marked response to calcipotriol ointment. J Dermatol 2000; 27: 123-126. PMID: 10721662