

Purpuric Variant of Keratosis Lichenoides Chronica

Variante purpúrica de queratosis liquenoide crónica

To the Editor:

Keratosis lichenoides chronica or Nekam disease is a rare dermatosis of unknown etiology. It is characterized by linear and reticulate lesions of lichenoid appearance, scaly plaques, and keratotic papules on the trunk and limbs. In addition to these 3 characteristic features, there may also be an eruption on seborrheic areas of the face. This clinical variability, the numerous confusing names given since Kaposi's first description in 1895,¹ and the nonspecific histological findings ("lichenoid pattern") have contributed to the underdiagnosis of this disease.

We report a case of keratosis lichenoides chronica in which we observed a marked purpuric component both clinically and histologically, in addition to the usual findings described.

The patient was a 68-year-old man on treatment with omeprazole for hiatus hernia. He consulted for a 2-year history of pruritic skin eruption, for which he had received no previous treatment. The lesions had first appeared on both sides of the chest and had progressively spread over the entire trunk and extensor surfaces of the limbs. A few isolated papules had resolved spontaneously with no residual pigmentation.

Physical examination revealed purpuric, lichenoid papules that coalesced into scaly, retiform plaques, located on the extensor surfaces of the lower legs and thighs, trunk, elbows, and on the backs of both hands (Figure 1).

Follicular keratotic papules with a violaceous erythematous color were also observed on the knees, thighs, and elbows (Figure 2).

There were scaly purpuric macules in the interdigital spaces of both hands, and several nail plates were dystrophic (thickened and yellowish). The head and neck region, palms, soles, and mucosas were unaffected.

The complete blood count showed mild leukopenia ($3 \times 1000/\text{mL}$) and the patient was referred to the hematology department, where hematological disease was ruled out at the time. Biochemical tests only revealed hypercholesterolemia (270 mg/dL). The other test results (coagulation studies, thyrotropin, vitamin B12, folic acid, hepatitis B and C serology, erythrocyte sedimentation rate, rheumatoid factor, and antinuclear antibodies) were normal or negative.

Biopsies of 2 keratotic papules (left knee and thigh) revealed hyperkeratosis with focal parakeratosis. The superficial dermis presented a band-like inflammatory infiltrate formed of lymphocytes, histiocytes, and few plasma cells, with spongiotic degeneration of the basal layer and some colloid bodies. There were extravasated red blood cells at the dermal-epidermal junction and variable amounts of hemosiderin in the cytoplasm of the macrophages (Figure 3).



Figure 1 Scaly erythematous plaques with a reticulate pattern are observed on the leg, together with keratotic papules.



Figure 2 Image of the purpuric component of the lesions.

Acitretin 25 mg/d was prescribed for 3 months, resulting in a slight improvement of the hyperkeratotic lesions but with no change in the scaly plaques or the purpuric component; the treatment was therefore withdrawn. The patient refused UV-A therapy, and the lesions have remained stable during subsequent follow-up.

Although keratosis lichenoides chronica was first described by Kaposi in 1895 as "lichen ruber acuminatus verrucosus et reticularis", the term now used was coined by Margolis in 1972. The varied nomenclature used has progressively added more confusion to the diagnosis of this disease, in itself clinically heterogeneous.¹ Keratosis lichenoides chronica is a rare dermatosis, considered by some authors as a variant of lichen ruber planus, due to its histological lichenoid features.²

Although scaly erythematous plaques, lichenoid papules, and follicular plugs are characteristic findings, other types

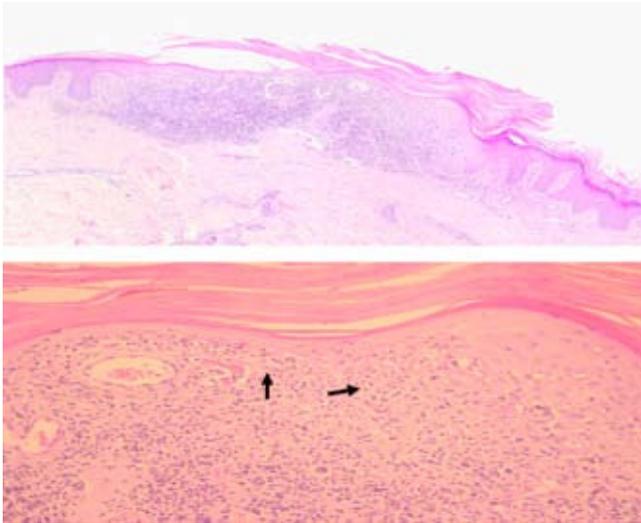


Figure 3 Hyperkeratosis and lichenoid infiltrate in the dermis. Extravasated red blood cells are marked by arrows. (Hematoxylin-eosin $\times 50$, $\times 100$).

of lesions associated with the disease have also been reported, such as a seborrheic eruption on the face, nail dystrophy, palmoplantar hyperkeratosis, alopecia, eye involvement, and even infiltration of the epiglottis.³

The differential diagnosis usually includes lichen planus, pityriasis lichenoides et varioliformis acuta, mycosis fungoides, and connective tissue diseases such as lupus erythematosus. Due to its atypical clinical presentation, however, the main differential diagnosis in our case was pigmented purpuric lichenoid dermatitis (Gougerot-Blum syndrome). This disease is clinically identified by purpuric macules and orange-brown pigmentation together with lichenoid papules, mainly located on the legs. Histologically, pigmented purpuric lichenoid dermatitis and keratosis lichenoides chronica share a chronic lichenoid pattern, though extravasated red blood cells in the dermis had not been described in the latter to date. Biopsy in our patient showed no sign of lymphocytic vasculitis of the vessels in the papillary dermis, as is typical of pigmented purpuric lichenoid dermatitis (this term is not accepted by all authors as there is no fibrin deposition in the vessel wall). However, because this finding is not constant, the absence of vascular damage does not constitute a definitive differentiating feature of the two dermatoses. This differential diagnosis was therefore based on clinical findings such as the extensive distribution of the skin lesions, initially on the trunk, the reticulate pattern of the scaly erythematous plaques, follicular hyperkeratotic papules, and the absence of purpuric macules and hyperpigmentation. These findings led to the diagnosis of keratosis lichenoides chronica, with the peculiarity of a clinical and histological purpuric component.

A pediatric variant of keratosis lichenoides chronica has been described as apparently showing characteristic findings such as erythematous-purpuric macules on the

face (generally as the initial lesions), frequent alopecia, and a lesser degree of involvement of the mucosas, nails, and eyes, in addition to an autosomal recessive pattern of inheritance reported in some pediatric cases but not in adults.⁴

The literature also describes a vascular variant of keratosis lichenoides chronica in which, in addition to the usual findings, there are associated reticulate telangiectasias.⁵ However, red blood cell extravasation has not been reported as a notable finding in the biopsies of these 2 variants.

Treatment of keratosis lichenoides chronica is generally disappointing, as in our case. Topical calcipotriol, oral acitretin, and psoralen UV-A (alone or in combination) are the most commonly used therapeutic modalities. More recently, other treatments have occasionally been used, such as narrowband UV-B, photodynamic therapy,⁶ and efalizumab.⁷ Spontaneous remission of the lesions is possible, though very uncommon.

In conclusion, we report a case of keratosis lichenoides chronica with an atypical presentation due to the marked clinical purpuric component together with extravasated red blood cells and hemosiderin-laden macrophages in the upper dermis. To our knowledge, no such findings have been described as part of this dermatosis to date.

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