Acta Derm Venereol 2002; 82: 128–130

Taylor & Francis
healthsciences

CLINICAL REPORT

Vascular Variant of Keratosis Lichenoides Chronica Associated with Hypothyroidism and Response to Tacalcitol and Acitretin

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Keratosis lichenoides chronica (KLC) is a rare chronic progressive cutaneous disease that is part of the heterogeneous group of lichenoid dermatoses. The typical clinical presentation is characterized by lichenoid hyperkeratotic papules and nodules arranged in a linear and reticular pattern on the trunk and extremities. Our case confirms the existence of a vascular variant of KLC. There is no consensus about its treatment, since it is refractory to many different treatment modalities. We report the effectiveness of acitretin in KLC in combination with tacalcitol. KLC is of unknown aetiology, but is perhaps associated with systemic diseases, most importantly glomerulonephritis and lymphoma. This is the second case associated with hypothyroidism. *Key words: acitretin; hypothyroidism; keratosis lichenoides chronica; tacalcitol.*

(Accepted December 19, 2001)

Acta Derm Venereol 2002; 82: 128–130.

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Keratosis lichenoides chronica (KLC), a rare chronic progressive cutaneous disease that is part of the heterogeneous group of lichenoid dermatoses (1), was described by Kaposi more than a century ago as *'lichen ruber acuminatus verrucosus et reticularis'*. In 1938, Nékam reported a similar case and named it *'porokeratosis striata lichenoides'* (2). Eventually, in 1972, Margolis gave the dermatosis its widely accepted name of KLC. Still, however, KLC remains a source of controversy, probably because of its rarity and the absence of a clear definition. At present it is accepted as a distinct entity, clinically and histologically, at one end of the spectrum of lichen planus (3). The typical clinical presentation of KLC is characterized by lichenoid hyperkeratotic papules and nodules arranged in a linear and reticular pattern on the trunk and extremities. Less frequently, seborrheic-dermatitis-like lesions on the face, palmoplantar keratoderma, nail changes and mucosal lesions have been described (2, 4–7). A rare vascular variant of KLC has also been described (8). KLC is of unknown aetiology, but may be associated with systemic diseases, most importantly glomerulonephritis and lymphoma (2). One previous case was associated with hypothyroidism (9).

There is presently no consensus about the treatment of KLC, since it is refractory to many different treatment modalities. We report here the effectiveness of acitretin in combination with tacalcitol.

CASE REPORT

A 48-year-old woman presented with a pruritic lichenoid papular eruption, the intensity of which had waxed and waned over a period of 20 years. She experienced an improvement in the summertime, especially when she was exposed to the sun in southern Europe. She described having had a seborrheic-dermatosis-like erup-

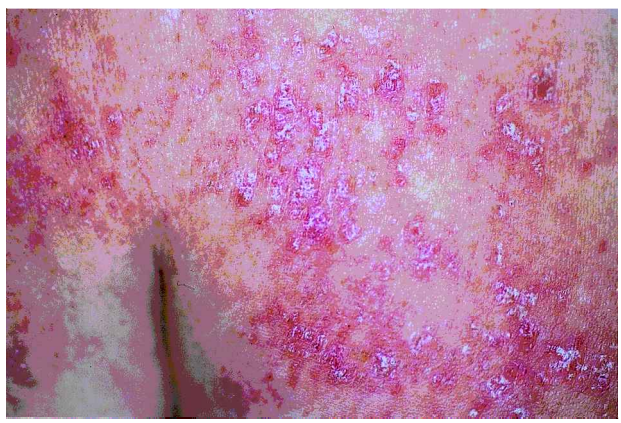

Fig. 1. Pruritic erythematous lichenoid hyperkeratotic papules on the upper back of a patient with keratosis lichenoides chronica.

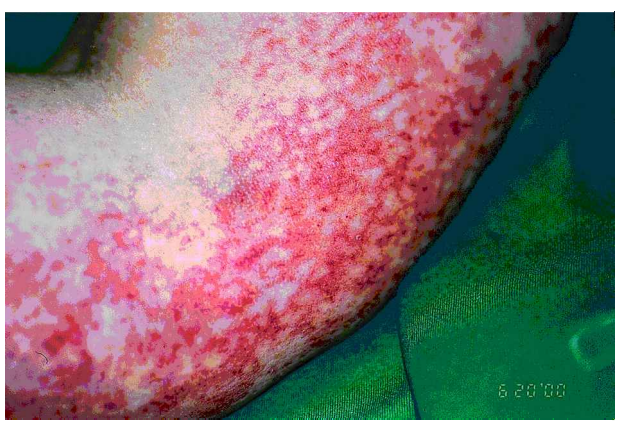

Fig. 2. An extensive reticular network of telangiectasia in combination with hyperkeratotic ridges on the flexor side of the arms in a patient with kerotosis lichenoides chronica.

tion on the face a year previously. Furthermore, she experienced recurrent oral aphthosis. The patient's medical history showed an auto-immune hypothyroidism since the age of 2 years, for which she was given thyroxine. She did not take any other drugs. Family history did not reveal significant diseases.

Physical examination showed symmetric clusters of erythematous lichenoid hyperkeratotic papules and several excoriations on the upper back, the shoulders, the lateral aspects of the trunk, and the buttocks (Fig. 1). The flexor side of the arms and forearms showed an extensive reticular network of teleangiectasia bilaterally, with distinct linear hyperkeratotic ridges and papules (Fig. 2). There was no oral aphthosis or genital ulcers. The nails were not affected.

Histology of the lichenoid papules showed a superficial perivascular infiltrate forming a mild band-like infiltrate which consisted of lymphocytes and histiocytes, the latter often loaded with melanin pigment.

Moreover, we noticed multiple eosinophilic colloid bodies in the papillary dermis and an extensive vacuolar alteration of the basal keratinocytes. High in the spinous layer there were multiple necrotic keratinocytes. Furthermore, there was epidermal hyperplasia with several V-shaped spots of hypergranulosis, focal spongiosis and parakeratosis. On direct immunofluorescence the colloid bodies stained with IgM, and a focal immunoreactivity to fibrinogen was seen at the dermal-epidermal junction.

Routine laboratory examinations, including erythrocyte sedimentation rate, complete blood count and liver and kidney function tests were within normal limits. Immunologic investigation showed a positive ANA of 1/80 with fine granular staining pattern, and an elevated IgE (234 U/ml). The thyroid function was well controlled. Hepatitis C serology was negative.

The treatment consisted of tacalcitol once a day and acitretin, which was initiated at a dose of 10 mg a day. She tolerated the acitretin well, so the dose was increased to 30 mg a day and led to an almost complete clearing of the lichenoid eruption, leaving only a slight erythema, within 4 weeks. Also the teleangiectasia appeared less prominent than before, but did not completely vanish on the inner side of her arms. After 4 months, acitretin was tapered back to 10 mg a day as maintenance therapy in association with tacalcitol, with success. During a 9-month follow-up, the improvement was maintained.

DISCUSSION

The great variety of synonyms used for KLC implies that there is no complete consensus about this rare disorder. Braun-Falco et al. (10), who reviewed 37 cases of KLC, confirmed the three hallmarks introduced by Pinol-Aguadé, namely, lichenoid papules, linear hyperkeratotic ridges and erythematosquamous plaques. Our patient experienced large clusters of itchy lichenoid hyperkeratotic papules on the back without a vascular background. On the inner aspects of the arms we noticed a prominent network of teleangiectasia with reticular hyperkeratotic ridges and less prominent papules. We also noticed several other characteristic clinical features of KLC, namely: the history of sun-induced improvement, the seborrheic-dermatitis-like lesions on the face, and the recurrent aphthosis (2). Nail involvement, palmoplantar hyperkeratosis, oral, genital and the eye involvement, are less frequently reported in patients with KLC (4–7). KLC is a polymorphous dermatosis, and there is little uniformity in the diagnostic criteria (2).

As regards the distinct vascular network on the arms we refer to cases reported in the French literature as 'Keratose lichénoide striée', the cases of Wätzig et al. and David et al., who described it as a vascular variant of KLC (8, 11–13). Noteworthy is the striking similarity of the teleangiectatic pattern on the inner side of the arms with the latter report. Interestingly, both cases were associated with severe itching, whereas KLC is normally asymptomatic, suggesting a possible relation between the itch and this subtype of KLC. Microscopic examination of the biopsy specimen showed a lichenoid pattern, although the dense band-like infiltrate, which is so characteristic for this group, was only mild in our case. However, in 13% of cases the typical lichenoid infiltrate was absent in a comparative pathological study of KLC (2). In conclusion, our clinical and histological observations are compatible with KLC.

KLC has a progressive course that may wax and wane chronically, although there are two reports of spontaneous resolution after 7 and 13 years, respectively (14, 15). Unfortunately, it is refractory to various treatment modalities. Several topical ointments, such as potent steroids, tretinoin, anthralin and keratolytics have failed to give significant improvement (2). Recently, calcipotriol twice a day was reported to give good results in two patients with KLC (9, 16). Nevertheless, tacalcitol was proposed because of the once a day application. The largeness of the affected hyperkeratotic area obliged us to add a systemic therapy. An oral retinoid was started, since this drug interferes with the epidermal proliferation and has anti-inflammatory actions (17). Moreover, the other vascular variant of KLC responded well to etretinate, and recently Avermaete et al. reported a case successfully treated with acitretin (8, 18). We therefore started acitretin 10 mg a day and increased the dose to 30 mg a day (0.5 mg/kg/day), since she tolerated it well. This led to an almost complete clearing of the lichenoid eruption after 4 weeks and complete relief from the itching. The teleangiectasia also appeared less prominent than before: we noticed a reduction in the redness and thickness of the cutaneous blood vessels. This was not the case in the other vascular variant of KLC (8). An alternative systemic treatment would have been PUVA, but the

data about its effectiveness are not consistent in the literature (2, 19–21). Systemic corticosteroids, methotrexate, cyclosporine, dapsone, vitamin A, gold, and tetracyclines are mentioned sporadically, but have been ineffective (2).

The patient presented by Grunwald et al. had had hypothyroidism for years, which was treated with thyroxine (9). Because our patient suffers also from hypothyroidism and is treated with thyroxine, we would like to draw attention to this association. Further cases are needed to evaluate a possible relationship. Concerning possible other associated diseases, two reports of chronic hepatitis related to keratosis lichenoides chronica – Wätzig & Schaarschmidt (13) and Marschalko et al. (22) – ought to be mentioned.

REFERENCES

1. Bleicher PA, Dover JS, Arndt KA. Lichenoid dermatoses and related disorders. II. Lichen nitidus, lichen sclerosus et atrophicus, benign lichenoid hyperkeratoses, lichen aureus, pityriasis lichenoides, and keratosis lichenoides chronica. J Am Acad Dermatol 1990; 4: 671–675.
2. Masouyé I, Saurat JH. Keratosis lichenoides chronica: the centenary of another Kaposi's disease. Dermatology 1995; 191: 188–192.
3. Grunwald MH, Amichai B, Finkelstein E, Kachko L. Keratosis lichenoides chronica: a variant of lichen planus. J Dermatol 1997; 24: 630–634.
4. Schnitzler L, Boutieller G, Bechteoille A, Verret JL. Keratosis lichenoides chronica with mucous membrane involvement and ocular pseudo-pemphigus. Follow-up study over 18 years. Retinoid therapy. Ann Dermatol Vénéréol 1981; 108: 371–379.
5. Thieulent N, Grézard P, Wolf F, Balme B, Perrot H. Guess what! Isolated palmoplantar hyperkeratosis revealing keratosis lichenoides chronica. Eur J Dermatol 1999; 6: 497–499.
6. Wallach D, About V, Kuffer R, Girard JC, Cottenot F. Kératose lichénoïde striée: deux cas avec manifestations orales. Rev Stomatol Chir Maxillofac 1984; 85: 307–310.
7. The nails in keratosis lichenoides chronica: characteristics and response to treatment. Arch Dermatol 1984; 120: 1471–1474.
8. David M, Filhaber A, Rotem A, Katzenelson-Weissman V, Sandbank M. Keratosis lichenoides chronica with prominent teleangiectasia: response to etretinate. J Am Acad Dermatol 1989; 21: 1112–1114.
9. Grunwald MH, Hallel-Halevy D, Amichai B. Keratosis lichenoides chronica: response to topical calcipotriol. J Am Acad Dermatol 1997; 37: 263–264.
10. Braun-Falco O, Bieber T, Heider L. Chronic lichenoid keratosis: disease variant or disease entity? Hautartz 1989; 40: 614–622.
11. Fraitag S, Oberlin P, Bourgault I, De Prost Y, Dubertret L, Roujeau JC, et al. Kératose lichénoïde striée. Ann Dermatol Vénéréol 1989; 116: 900–902.
12. Degos R, Labouche F, Civatte J, Touraine R, Balouet G. Keratosé lichénoïde striée. Ann Dermatol Vénéréol 1974; 101: 391–392.
13. Wätzig V, Schaarschmidt H. Keratosis lichenoides chronica. Z Hautkr 1986; 61: 783–787.
14. Van de Kerkhof PCM. Spontaneous resolution of keratosis lichenoides chronica. Dermatology 1993; 187: 200–204.
15. Chapman RS. Lichen verrucosus and reticularis. Dermatologica 1971; 142: 363–373.
16. Chang S, Jung E, Hong S, Choi J, Sung K, Moon K, et al. Keratosis lichenoides chronica: marked response to calcipotriol ointment. J Dermatol 2000; 27: 123–126.
17. Saurat JH. Retinoids and psoriasis: novel issues in retinoid pharmacology and implications for psoriasis treatment. J Am Acad Dermatol 1999; 41: S2–S6.
18. Avermaete A, Kreuter JA, Stucker M, Von Kobyletzki G, Altmeyer P, Jansen T. Keratosis lichenoides chronica: characteristics and response to acitretin. Br J Dermatol 2001; 144: 422–424.
19. Lang PG, Jr. Keratosis lichenoides chronica: successful treatment with psoralen-ultraviolet-A therapy. Arch Dermatol 1981; 117: 105–108.
20. Lazarova AZ, Rodriguez MA, Diaz MA. Chronic lichenoid keratosis: apropos of a case. Med Cutan Ibero Lat Am 1988, 22: 671–675.
21. Ryatt KS, Greenwood R, Cotterill JA. Keratosis lichenoides chronica. Br J Dermatol 1982; 106: 223–225.
22. Marschalko M, Papp I, Szalay L, Harsing J, Horvath A. Keratosis lichenoides chronica with chronic hepatitis: a coincidence? Acta Derm Venereol 1996; 76: 401–402.