Keratosis Lichenoides Chronica: Marked Response to PUVA in Combination with Acitretin

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Accepted February 29, 2005.

Sir,

Keratosis lichenoides chronica (KLC) is a rare, chronic and progressive dermatosis. The disease usually affects adults aged 20–50 years and is very resistant to therapy (1). Although KLC shares some clinical and dermatopathological features with lichen planus, it is considered to be a distinct entity (2). We report a patient with recalcitrant KLC, who had a marked response to PUVA in combination with acitretin.

CASE REPORT

A 52-year-old male patient was admitted to our department with a 5-year history of non-pruritic pink-purple coloured papular lesions on upper and lower extremities. The patient had been treated in the preceding year with the diagnosis of lichen planus by oral prednisolone 1 mg/kg for 2 months, griseofulvin 1 g daily for 1 month and acitretin at a daily dose of 60 mg for 6 months, but no beneficial effect was obtained with these treatments.

The medical history of the patient was unremarkable and no other family member was affected. Dermatologic examination revealed violaceous lichenoid hyperkeratotic papules and plaques arranged in a bilaterally symmetrical linear and reticular pattern on the flexor regions of forearms, popliteal fossae, thighs and dorsal aspects of hands and feet (Fig. 1A). Erythematous scaly eruption was seen on his face. Palmoplantar hyperkeratosis was present and dystrophic changes and subungual hyperkeratoses in the toenails were noted. Examination of the oral mucosa revealed eroded lesions on the hard palate (Fig. 2). Conjunctival hyperaemia was present. General physical examination revealed normal findings. Routine laboratory examinations including erythrocyte sedimentation rate, complete blood count, liver and kidney function tests were within normal limits. Dermatopathologic examination showed hyperkeratoses, focal parakeratoses, acanthosis in the epidermis, necrotic keratinocytes in the basal cell layer and dense band-like lymphocytic infiltration in the upper dermis (Fig. 3). The patient was diagnosed as having KLC according to the typical clinical and dermatopathologic findings. PUVA treatment was initiated three times per week. After 3 months partial improvement was noted and PUVA was combined with acitretin 35 mg/day. Marked improvement was noted after 2 months with clearance of all lichenoid hyperkeratotic lesions (Fig. 1B).

DISCUSSION

KLC was originally described by Kaposi in 1895 as lichen ruber verrucosus et reticularis (3). Margolis et al. coined the term keratosis lichenoides chronica in 1972 (4). As KLC shares many clinical and histological features with lichen planus, some authors have considered KLC to be a variant of lichen planus (5). However, Braun-Falco & Bieber (6) suggested that it was a distinct entity characterized by lichenoid hyperkeratotic papules arranged in a linear pattern, erythematosquamous plaques and seborrheic dermatitis on the face.

KLC is characterized by lichenoid hyperkeratotic pink-purple coloured papules and plaques arranged symmetrically in a linear or reticular pattern on the extremities and trunk. In 75% of cases there is a facial

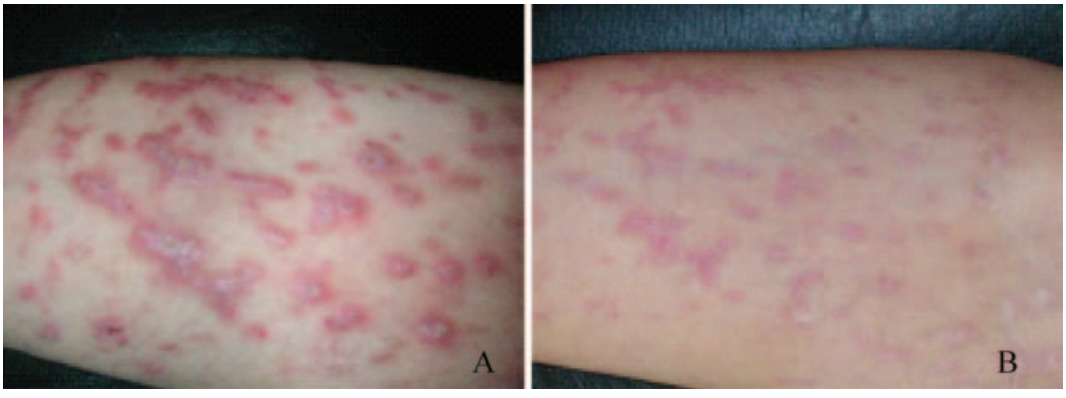

Fig. 1. (A) Close up view of the lesions located on the forearms. (B) Resolution of the lesions after 2 months of PUVA and acitretin combination therapy.

DOI: 10.1080/00015550510038269

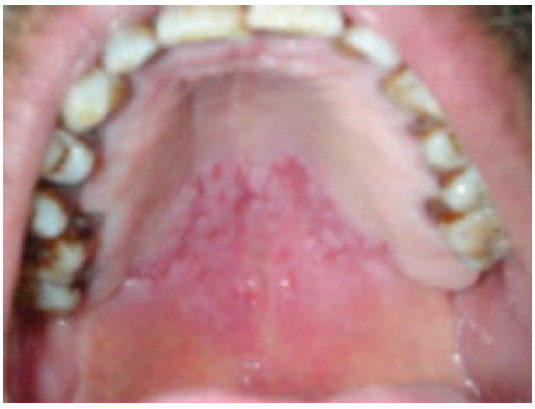

Fig. 2. Eroded lesions on hard palate.

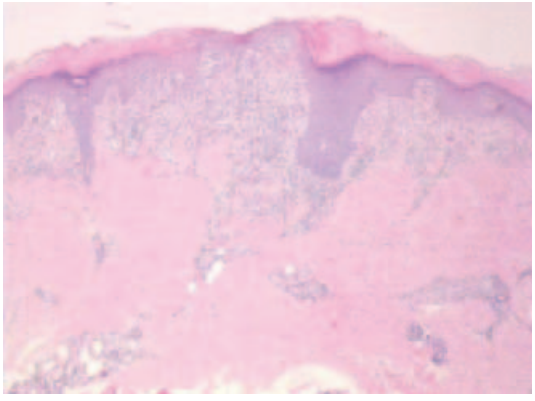

Fig. 3. Hyperkeratosis, focal parakeratoses, acanthosis in the epidermis, necrotic keratinocytes in the basal cell layer and dense band-like lymphocyte infiltration in the upper dermis (H&E, ×40).

eruption resembling seborrhoeic dermatitis and rosacea. Nail dystrophy can be found in 30% of cases and palmoplantar hyperkeratosis in 40% (7). The eruption is usually asymptomatic. It can be associated with oral ulcers, ocular involvement presenting as blepharitis, conjunctivitis, anterior uveitis, iridocyclitis and genital involvement including keratotic papules on the scrotum and penis, chronic balanitis and phimosis (8).

Marked hyperkeratosis, focal parakeratosis, acanthosis and keratotic plug formation may be seen histopathologically in the epidermis. In some cases variable degrees of spongiosis may be observed. There is a dense band-like lymphohistiocytic infiltration in the upper dermis and focal degeneration in the basal layer (7).

Clinically, pink-purple coloured papular lesions of KLC may be misdiagnosed as lichen planus and the linear pattern of the lesions can be confused with Koebner phenomenon. While the lesions in Koebner phenomenon have an asymmetrical linear pattern, linear lesions in KLC are symmetrically arranged. Moreover, the presence of seborrhoeic dermatitis-like eruption on the face, blepharoconjunctivitis, absence of pruritus and resistance to therapies including oral steroids are useful features that help in the differentiation of these two entities (8).

Some reports have noted an association of KLC with a variety of diseases such as hepatitis, chronic lymphoid leukaemia and nephropathy, but it has not been possible to establish a link between KLC and these disorders (9). In our patient no associated pathology was found.

The course of KLC is chronic and progressive. The disease is very resistant to therapy and generally has a duration of more than 10–15 years (8). Spontaneous resolution has been reported in only two cases, after 7 and 13 years (10, 11). Although topical treatments are ineffective in this disorder, in a few cases beneficial effect of calcipotriol was noted (12). Systemic steroids, antimalarial agents, sulphones, methotrexate and cyclosporine are generally ineffective (8). In some cases, oral retinoids, PUVA or the combination of PUVA and retinoids have induced marked improvement of the lesions, but have proved ineffective in others (9). In our case, while acitretin and PUVA had not been efficient when used as a monotherapy, the combination of these treatments provided optimal treatment response.

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