Keratosis Lichenoides Chronica with Chronic Hepatitis: A Coincidence?

Sir,

Keratosis lichenoides chronica is a rare disease, characterised by lichenoid keratotic papular lesions arranged in a linear or reticulated pattern on the extremities or on the trunk. Whether this disease is a variant of lichen planus is still questionable. Many authors suggest that the disease is a distinct entity. We report a case of keratosis lichenoides chronica associated with chronic hepatitis of unknown origin.

CASE REPORT

In 1989 a 41-year-old man presented with a 2-year history of violaceous, moderately itchy, scaling lesions on the trunk and extremities, which later progressed to the face, back of the hands, scrotum and penis.

The lesions had increased over the last 6 years, with some fluctuation. Later most of the back and almost the whole of his arms and legs were involved. Intraoral erosions were observed. In the nose were signs of rhinitis sicca, and he had bilateral keratoconjunctivitis. He did not have a history of chronic alcohol abuse or hepatotoxic drug intake.

In 1995 examination revealed symmetric eruptions on the trunk, extensor surfaces of the limbs, on the back of the hands, on the soles and feet, on the buttock, on the scrotal area, on the glans penis and on the face.

The eruption consisted of bluish red firm, lichenoid papules; they were discrete or grouped, and on the back they showed a reticulated pattern (Fig. 1), on the limbs a linear pattern. On the face there was an erythematous, reddish, scaly, telangiectatic eruption resembling lupus erythematosus. The lateral parts of the eyebrows were thinned.

Individual lesions had coalesced into large parakeratotic psoriasiform plaques on the dorsa of the hands and on the buttocks.

On the lateral borders of the feet and on the soles, there were deep papules with keratotic plugs.

White patches were present on the mucous membrane of the mouth. On the glans penis reticulated lichenoid papules were observed.

Histology showed a lichenoid type of tissue reaction, with epidermal thickening, acanthosis, hypergranulosis and hyperkeratosis, presence of colloid bodies and basal layer liquefaction degeneration. Occasional foci of parakeratosis were seen. There was a band-like infiltrate of mononuclear cells.

Direct immunoflurescent examination with anti-human IgG, IgM, and C3 was negative. The following laboratory studies showed normal values: white blood cell count, erythrocyte sedimentation rate, haemoglobin, haematocrit, differential cell count, urine analysis, serum transaminases, alkaline phosphatase, serum total bilirubin, serum creatinine, serum urea nitrogen, glucose, lactic dehydrogenase, serum sodium chloride, potassium, RPR test for syphilis, rheumatoid factor, total haemolytic complement activity, serum protein electrophoresis, T-lymphocyte subsets, serum IgA, IgM content. Gammaglutamil transferase was slightly elevated during the 6-year observation period. Antinuclear antibody was slightly positive (in 1/80) and serum IgG elevated (21.9 g/l) (normal value: 7.0–15 g/l). Hepatitis B and C serology was negative. Roentgenologic examination of the chest was negative. Abdominal and pelvic ultrasound and scintigraph revealed moderate hepatosplenomegaly. Liver biopsy showed fatty degeneration of the liver, with chronic aspecific hepatitis with portal fibrosis. Gastroscopy revealed atrophic gastritis. Treatment with topical and oral corticosteroids, chloroquine, etretinate and cyclophosphamide was ineffective. PUVA treatment caused marked inflammation of the skin symptoms, so it was stopped.

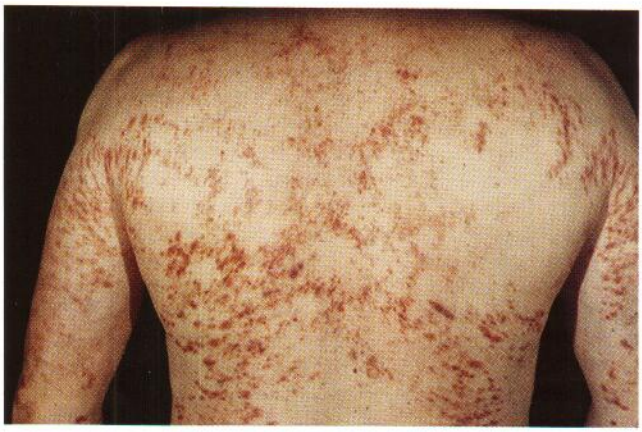

Fig. 1. Lichenoid papules on the back in reticulated distribution.

DISCUSSION

In 1972 Margolis et al. presented a 36-year-old patient with a chronic disease characterised by lichenoid keratotic skin symptoms and chronic course under the name of keratosis lichenoides chronica (1). In 1976 Kint et al. (2) and Petrozzi (3) presented similar patients and they recognized that the disease was identical with that described by Kaposi in 1885 under the name of lichen ruber acuminatus verrucosus et reticularis (4).

The disease was reported under various terms: porokeratosis striata (5), keratose lichenoide striee (6), lichenoid trikeratosis (7), suggesting that the pathogenesis and the aetiology are not clear. The case that we report here has the clinical and histological features described by most authors: the linear and reticulated distribution of the lichenoid papules, the facial eruption resembling lupus erythematosus, the horny keratotic papules on the soles. Other major features were the involvement of the mucous membranes which was prominent in our case: keratoconjunctivitis, involvement of the mouth and the genital mucous membrane, rhinitis sicca and atrophic gastritis were observed. The question arises if the rhinitis sicca and the atrophic gastritis are part of the disease spectrum. There are no previous reports on such manifestations, to the best of our knowledge.

The lichenoid characteristics were shown histologically in our patient. Immunofluorescence was negative, as in the majority of the cases.

The aetiology and the pathogenesis of the disease are not clear. There have been reports on its association with systemic diseases: it was reported in association with toxoplasmosis (8), chronic lymphoid leukemia (9), with multiple sclerosis (10) and with joint involvement (7).

In our patient chronic hepatitis was diagnosed histologically. Whether this is a coincidence or not is difficult to determine.

The question is of interest because an increased prevalence of chronic liver diseases—including primary biliary cirrhosis, chronic active hepatitis or cirrhosis of unknown cause—has been reported in patients with lichen planus, though the association of the two diseases was questioned by others. Recently association of lichen planus and hepatitis C infection was reported (11).

A common immunologic pathway is suggested: T-cell mediated cytotoxicity might cause the same pathological process in the liver and in the skin. Keratosis lichenoides chronica is very similar to lichen planus both clinically and histologically; some authors consider it to be the same process (12, 13). A similar pathomechanism might play a role in the skin symptoms of keratosis lichenoides chronica associated with chronic hepatitis. In our case hepatotoxic drugs or alcohol intake was not justifiable in the patient history, and alcoholic hyaline deposition was not seen in histology. Hepatitis B and C virus serology was negative. Laboratory data and histology had no clues for metabolic or hepatobiliary liver diseases, and the patient had no signs of chronic hepatovenous disease. Steatohepatitis was present, with focal periportal accumulation of lymphocytes and fibrosis. The cause of the chronic hepatitis was unclear.

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Márta Marschalkó[1], Ildikó Papp[1], László Szalay[2], Judit Hársing, and Attila Horváth[1]

[1]Department of Dermatology, and [2]II. Department of Medicine, Semmelweis Medical School, Mária u.41, H-1045 Budapest, Hungary.