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. Intracutaneous reactions were observed. In the nose were signs of rhinitis sicca, and he had bilateral keratoconjunctivitis. He had 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. I), on the limbs a linear pattern. On the face there was an eroded, 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 immunofluorescent 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, lactate 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. Gamma-globulin transferrin 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.

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 Kim et al. (2) and Petrozzi (3) presented similar patients and they recognized that the disease was identical with that described by Kaposis in 1885 under the name of lichen ruber acuminatus verrucosus et reticularis (4).

The disease was reported under various terms: porokeratosis striata (5), keratose lichenoid striae (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).

Fig. 1. Lichenoid papules on the back in reticulated distribution.
Lobular Panniculitis Associated with Venous Congestion in Right-sided Heart Failure

Str.
Nodular panniculitis is a descriptive term which refers to the presence of inflammatory nodules in the subcutaneous fat. Traditionally a discrimination is made between septal, lobular and mixed panniculitis according to the pattern of inflammation, i.e. whether it occurs in the interlobular septa, in the lobule proper or both (1, 2). Septal panniculitis is usually associated with erythema nodosum, while the lobular type has various causes. Some of the different main forms have distinct histological features and/or are associated with typical laboratory findings, aiding the diagnostic evaluation (2, 3), but in about 50% of the cases the aetiology remains unknown (idiopathic panniculitis).

We report here a patient with venous congestion secondary to right-sided heart failure associated with primary pulmonary hypertension, who developed nodular panniculitis in the subcutaneous fat of the abdominal wall. A temporal relationship was noted between the development of severe heart failure and the occurrence of tender subcutaneous nodules. These nodules disappeared promptly after successful treatment of the pulmonary hypertension and heart failure.

CASE REPORT
The patient was a 24-year-old woman, slightly overweight, but previously healthy, except for a history of mild allergic asthma. In May 1991 she noticed for the first time an unusual exertional dyspnoea during a skiing holiday. No further symptoms were experienced until the spring of 1992, when exertional dyspnoea recurred. During the following months, an upper abdominal pain and discomfort were added to her symptoms. She was admitted to a local hospital in the beginning of July 1992 after three episodes of fainting. The tentative diagnosis was acalculous cholecystitis and orthostatic hypotension. During the following 2 months she started to gain weight and to develop oedema of the lower extremities. At the same time, she noticed tender nodules in the subcutaneous fat of the abdominal wall. In September a diagnosis of pulmonary hypertension with secondary right heart failure was made, explaining the episodes of syncope, dyspnoea, abdominal pain and oedema. She was then admitted to our hospital, and on admission was found to have right-sided heart failure of NYHA class IV, and generalised oedema corresponding to a weight gain of approximately 15 kg. Ultrasonographic examination of the abdomen revealed ascites and marked portal hypertension. Tender nodules on the anterior abdominal wall were observed. Punch biopsies of these nodules revealed a picture of panniculitis of the mixed type, but with a predominantly lobular pattern, in the subcutaneous fat (Fig. 1a). Histologically there was quite extensive fat necrosis with early lipophagia. The most striking features were an abundant extravasation of red blood cells and the presence of fibrinoid thrombi in capillary vessels (Fig. 1b). Only a sparse inflammatory response, with mostly macrophages and a few lymphocytes, was observed. There was no sign of vasculitis or of a granulomatous reaction.

The further clinical investigation of the patient included pulmonary vessel radiology with pressure recordings, and laboratory tests for coagulopathy, serum amylose, alpha-1-antitrypsin and other acute phase reactants, antibodies against nuclei, DNA and neutrophil cyto-