

Generalized Lichen Planus Associated with Primary Biliar Cirrhosis which Resolved after Liver Transplantation

Sir,

Lichen planus (LP) is a relatively common inflammatory dermatosis with characteristic lesions affecting the skin, nails and mucous membranes. The association of lichen planus with primary biliar cirrhosis (PBC) is now well recognized. We report a case of generalized LP associated with PBC, which cleared after liver transplantation.

CASE REPORT

A 61-year-old woman developed in 1987 generalized pruritus, jaundice, fatigue and weight loss. Liver function tests showed evidence of cholestasis: total bilirubin 9.6 mg/dl, direct bilirubin 7.6 mg/dl, SGOT/AST 48 IU/l, SGPT/ALT 47 IU/l, alkaline phosphase 1289 IU/l. Hepatitis serology to virus A, B and C was negative. Anti-mitochondrial antibodies were negative. Antinuclear antibodies were positive at a titre of 1:1600 and anti-DNA antibodies were negative. Liver biopsy was consistent with PBC. Cholestyramine and rifampin were prescribed for the symptomatic relief of her pruritus, with partial improvement. No other specific therapy was given. She did not receive d-penicillamine (d-PCN).

In July 1991 she developed itchy, flat-topped violaceous papules, scattered all over the body, that converged into great plaques. Oral and genital mucosae were clear. A biopsy specimen of cutaneous lesions was typical of LP. The patient was treated with emollients, topical steroids, UVB and UVA light, with partial relief of her pruritus but without clearance of the lesions.

In March 1992 a liver transplantation was performed. Immunosuppressive therapy with CsA at 3 mg/kg/day and prednisone at 15 mg/kg/day was started. There was a great clinical improvement almost immediately after the liver transplantation, with complete relief of her pruritus and gradual flattening of the lesions until the complete resolution of LP in 4 weeks. Prednisone was tapered until withdrawal in October 1992. At this time a new skin biopsy was made, showing only postinflammatory hyperpigmentation. The patient is at present in remission with CsA at 3 mg/kg/day.

DISCUSSION

The association of LP and PBC is probably more than coincidental and may be due to the fact that both conditions are based on an alteration of a cell-mediated immune response and are associated with a variety of other autoimmune disorders (1). The tendency in patients with PBC to develop LP may be exacerbated by d-PCN (2, 3). The presence of LP in patients with PBC supports the hypothesis that these conditions share a common pathogenetic mechanism with graft-versus-host disease (GVHD) (4).

Powell et al. (3) reported 24 patients with LP and PBC from a double-blind, randomized control trial of d-PCN in 268 patients with PBC at the Mayo Clinic. Two patients presented generalized LP, both of them related to d-PCN. When d-PCN was discontinued, the lesions of LP gradually resolved over several months. Topical steroids were the only therapy required for LP in this study.

Our patient presented generalized LP associated with PBC unrelated to d-PCN and unresponsive to usual therapies. To the best of our knowledge, this is the second case of generalized LP with PBC unrelated to d-PCN that has been reported (1). In our patient, the lesions cleared after liver transplantation and immunosuppressive therapy with CsA. The role of liver transplantation in the resolution of LP remains unknown.

CsA given orally has been shown to produce an excellent response in the treatment of cutaneous LP (5, 6). In our opinion, low-dose CsA therapy maintained for a long time may play an important role in the remission of LP in our patient. However, further studies would be necessary to clarify the different roles of liver transplantation and/or CsA therapy in the resolution of chronic, generalized LP associated with PBC unrelated to d-PCN.

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