Generalized cutaneous morphea in a patient with post-hepatitis C cirrhosis

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SUMMARY -

A case of generalized cutaneous morphea in a a 55-year-old female patient known to have suffered 10 years from post-hepatitis C virus cirrhosis is reported. Anti-HCV antibodies were present, whereas screening for HBs antigen and anti-HBc antibodies was negative.

Introduction

Cutaneous morphea is a patchy scleroderma mainly affecting the skin. It is usually idiopathic; an association with hepatitis C virus (HCV) infection has been reported only exceptionally.

Case report

We report the case of a 55-year-old female patient known to have suffered from post-hepatitis C cirrhosis for 10 years. The diagnosis of cirrhosis was established by the following signs: the presence of edematous ascites syndrome, splenomegaly, and hepatomegaly with sharp edges; endoscopic findings of esophageal and gastric varicosities; and a dysmorphic liver revealed by ultrasonography. Biochemical investigations showed an elevation in serum transaminases, a low prothrombin time, and pancytopenia. Anti-HCV antibodies were

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present, whereas screening for HBs antigen and anti-HBc antibodies was negative. Moreover, IgG and IgA cryoglobulins were noted. Other potential causes for cirrhosis were excluded. Transparietal hepatic biopsy was contraindicated because of a possible hemorrhage, and the patient refused a transjugular biopsy.

The patient was referred to the dermatology unit because of generalized sclerotic, atrophic, and dyschromic patches of 6 years' duration. Her history failed to reveal Raynaud's phenomenon, dysphasia, or dyspnea. The patient denied photosensitivity or cutaneous fragility.

Cutaneous examination showed multiple pearly, white, sclerotic patches predominantly on the trunk, where they were confluent. Similar lesions were also expressed on the limbs and the scalp, appearing as scarring alopecia. There were no acrosclerosis, blisters, ulcerations, or milia. A cutaneous biopsy supported the diagnosis of scleroderma. Antinuclear factors were nega-

K E Y W O R D S

generalized morphea, cirrhosis, hepatitis C virus infection, case report tive in indirect immunofluorescence (IIF) on rat liver sections but were weakly positive with Hep2 cells as a substrate. Serologic screening for *Borrelia burgdorferi* was negative. The diagnosis of generalized cutaneous morphea associated with post-hepatitis C cirrhosis was thus confirmed.

Discussion

Chronic infection with HCV is reported in association with several dermatoses, such as lichen planus, cutaneous vasculitis, and porphyria cutanea tarda (PCT) (1–4). Abu-Shakra et al. (1) reported a case of systemic sclerosis associated with an HCV infection and considered it a systemic autoimmune disorder similar to the association of HCV with systemic lupus erythematosus. The association of HCV with cutaneous morphea seems to be rare. To our knowledge, only one article reporting two cases of cutaneous morphea associated with chronic active hepatitis C has been published (3). However, cutaneous morphea has never been reported in

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association with cirrhosis resulting from a HCV infection.

A variant of porphyria cutanea tarda (PCT) expressing scleroderma-like lesions may be associated with HCV (4). Our patient did not show signs of PCT-like photosensitivity, skin fragility, or milia. The role of HCV in the pathogenesis of auto-immune diseases is not completely understood. It is thought that extra-hepatic virus replication, especially within mononucleated cells, may suppress immune tolerance in genetically predisposed individuals (1). Proliferation of lymphocytes and monocytes can be observed in scleroderma. The increase in T helper cell function may lead to the stimulation of B lymphocytes and the synthesis of auto-antibodies and lymphokines that stimulate collagen synthesis (5). Such a reaction might be the trigger factor for scleroderma in patients with HCV infection. The number of reported cases is not large enough to allow conclusions on a possible causal relationship between the two conditions. A more reliable evaluation would be possible if HCV serology were regularly performed in patients with cutaneous morphea.

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