DESCRIPTION

A 60-year-old man from Qom was first admitted to the lung clinic complaining his preexisting dyspnea had progressively worsened during the past month. He also complained of high fever, weakness, and dysphagia during the past month. Chest radiography and high-resolution computed tomography showed no lung involvement except pulmonary hypertension and function class 2 dyspnea. Because he also had skin thickening of the fingers, cold fingertips, and red spots on the face appearing 2 years before, he was referred to the rheumatology clinic.

The patient had been diagnosed with celiac disease (CD) and hepatic cirrhosis 10 years before. Back then, he had complaints of severe weight loss, decreased appetite, hemoptysis, and anemia, and his prothrombin time and international normalized ratio were abnormal. Lower abdominal and pelvic ultrasound showed a small liver with coarse and heterogeneous echotexture, signifying cirrhotic changes. Port vein thrombosis was also visible alongside a noticeable ascitic fluid in the abdominal and pelvic cavity. Endoscopy with duodenal biopsy revealed gastrointestinal bleeding.

A liver examination showed negative virological markers and autoimmune hepatitis–associated antibodies. A liver biopsy then yielded no specific findings, and he was given supportive treatment with a diagnosis of cryptogenic cirrhosis. The duodenum tissue specimen confirmed CD (modified Marsh grade 3a[1]), and with an antigliadin antibody titer of 120 (reference range, 0–25), the patient was started on a gluten-free diet. The diet was effective; he remained asymptomatic for 10 years before the recent referral. His current medication was propranolol 10 mg twice daily.

On physical examination, dryness of the eyes and oral mucosa were evident. He had macular changes on his face (Fig. A and B) and his fingers (Fig. C). Telangiectasia and palpable stiffness were also present on the face. Jugular venous pressure was approximately 12 cm high. Crackles were heard in the base of the lungs, and dullness was noted in both auscultation and percussion. The patient's hands had increased stiffness and thickness. Transverse slots of the dorsal surface of the fingers had disappeared. Raynaud phenomenon and lower limb edema (+1) were evident.

A chest radiograph revealed mild pleural effusion in the lungs' bases. High-resolution computed tomography showed glass opacity with mild pleural effusion, especially in the lower lobe. Spirometry showed a significant decrease in diffusing capacity of lung for carbon monoxide. Echocardiography revealed pulmonary artery pressure of 55 mm Hg in favor of hypertensive pulmonary disease.

FIGURE. Macular changes evident in patient's face (A, B) and hands (C).

A sign scleroderma diagnosis was followed by positive anti–centromere Ab and antinuclear antibodies. He was then started on prednisolone (2 mg twice daily), bosentan (2 mg twice daily), and sildenafil (100 mg 4 times daily). Soon after 2 weeks, his symptoms showed marked improvement (pulmonary artery pressure = 35 mmHg). The patient implied he has been pleased with the treatment progress and had major improvements.

Our reported case provides some evidence regarding the link between CD and scleroderma, which has not been extensively investigated. Having a previously diagnosed immunological disease such as CD can point the physicians toward the probable development of other diseases of its kind, facilitating the diagnosis and patient education about the symptoms that might appear later on.

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