AUTOIMMUNE CONNECTIVE TISSUE DISEASES

ERASMUS SYNDROME ASSOCIATED WITH SEVERE LIVER INJURY

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Erasmus syndrome associates systemic scleroderma (ScS) with exposure to silica. It is common among miners and masons. We report a case of Erasmus syndrome with severe liver injury seen in Antananarivo Madagascar.

A 48-year-old man,smoking with a history of Raynaud's syndrome, dysphagia was hospitalized for NYHA III dyspnea evolving since 3 months. He worked for 28 years as a mason. Clinical examination included extensive diffuse cutaneous sclerosis of the face, sclerodactyly, frank mucocutaneous icterus, crackling rales of the two pulmonary bases. The biological examinations revealed an inflammatory syndrome, a cholestatic syndrome, moderate hepatic cytolysis. The serologies of hepatitis B and C were negative. Electrophoresis of serum proteins showed polyclonal hypergammaglobulinemia and hypoalbuminemia. The antinuclear antibody was positive. Radiography and pulmonary CT showed diffuse interstitial syndrome. The cardiac echodoppler revealed a PAH with a PAPS at 47 mmHG. FOGD found a HTP with esophageal varices stage III and erosive gastritis. The diagnosis of Erasmus syndrome associated with pulmonary fibrosis and severe hepatic involvement was evoked. The evolution was quickly fatal.

ScS during Erasmus syndrome is identical to ScS "idiopathic" clinically and paraclinically. Pulmonary involvement is common, while liver damage remains rare. The peculiarity of our case lies in the occurrence of severe hepatic involvement with hepatic cirrhosis complicated by oesophageal varices rupture. Indeed, hepatic involvement during an SSc can be asymptomatic and revealed by complications. A professional and unprofessional toxic investigation must be done in front of a Scs.

Keywords: Erasmus syndrome, man, liver injury, Madagascar