Abstract

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A Case of Porto Sinusoidal Vascular Disorder of The Liver In A Patient Suffering From Systemic Sclerosis

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Background

A 55 year-old man was admitted to emergency department for abdominal distension, peripheral edema and dyspnea. He suffered from aplastic anemia and systemic sclerosis (Ssc), an autoimmune disease of connective characterized by vasculopathy and skin and internal organs fibrosis treated with methotrexate and folate. Alcohol consumption was occasional. Three months before he suffered from SARS-CoV2 pneumonia. He showed worsening of an already known anemia (Hb 8.4 g/d), thrombocytopenia (96000/mcL), hypoalbuminemia (2.9 g/dL), presence of anti citrullin and anti-centromeric antibodies Screening for HBV and HCV: negative. Echocardiography was normal. Abdominal ultrasound showed ascites, increased portal vein diameter (20mm) and flow velocity (29.4 cm/sec), splenomegaly (22 cm). CT-scan evidenced bilateral pneumonia and pleural effusion, ascites, splenomegaly, liver steatosis. Paracentesis showed serum-ascites albumin gradient>1.1 g/dL, suggesting portal hypertension related ascites, spontaneous bacterial peritonitis excluded. Endoscopy evidenced moderate severe congested gastropathy, colic mucosa congestion, no gastro-esophageal varices. Liver stiffness by fibroscan was 10.7 KPa. Diuretics and antibiotics were administrated with resolution of acute condition . The patient was referred to a tertiary care center for further investigation. Hemodynamic study revealed normal atrial and inferior vena cava pressure and a hepatic venous pressure gradient (HVPG) of 7 mmHg (wedge-hepatic venous pressure: 16 mmHg, free hepatic venous pressure: 9 mmHg) suggesting pre-sinusoidal more than intra-sinusoidal component of portal hypertension (PH). Transjugular liver biopsy evidenced focal areas of hepato-portal sclerosis and nodular regenerative hyperplasia configuring a vascular porto-sinusoidal disorder (PSVD).

PSVD describes a group of liver vascular disorders characterized by lesions encompassing portal venules and sinusoids irrespective of PH and with no histological evidence of cirrhosis. The injury and occlusion of the intrahepatic portal microvasculature could increase resistance at pre-sinusoidal level. This condition has been defined for a long time "Idiopathic non cirrhotic portal hypertension'(INCH): this term had several limitations excluding patients with no PH, as may occur in early stages, despite similar findings on biopsy. Biopsy findings are fundamental: nodular regenerative hyperplasia, obliterative portal vein stenosis, incomplete septal fibrosis/cirrhosis. Liver stiffness exceeding 20 Kpa exclude PSVD. Patients present with signs and symptoms of PH as varices, hypersplenism, thrombocytopenia, ascites; some are asymptomatic. Treatment consists on managing PH-related complications, anticoagulation should be considered. Prognosis is generally better than that of cirrhosis but advanced lines of therapies (e.g. transjugualr intra-hepatic porto-systemic shunt, liver transplantation) may be necessary.

Multiple studies have reported an association between PSVD and autoimmune disorders probably linked to hyper activation of intrasinusoidal T lymphocytes.

Some cases of PSVD have been described in patients with SSc even if liver involvement in SSc is atypical. Cells in endothelial to mesenchymal transition in the vessels of SSc patients could cause endothelial dysfunction. The obliterative portal damage of sclerosis could increase resistance leading to PH, condition worsened by autoimmune disease prothrombotic state. More studies are needed to highlight pathogenesis behind the association of autoimmune diseases and PSVD. Standardized criteria are necessary. The hub-and-spoke model of care is the best way to warrant a complete diagnostic and therapeutic approach for patients with portal hypertension of unknown etiology.