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IMAGE | LIVER

Pseudo Symmer's Pipe-Stem Fibrosis in Idiopathic Noncirrhotic Portal Hypertension Associated with POEMS Syndrome

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CASE REPORT

A 44-year-old man without history of travel was referred to our clinic for evaluation of refractory ascites for 1 year, including low serum-to-ascites albumin gradient and low protein intractable to diuretic. He reported no drug intake beyond treatment for polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy (immunoglobulin M), and skin changes, together known as POEMS syndrome. The POEMS treatment history initially included bevacizumab 5 mg/kg once weekly, which was stopped after the first week due to worsening ascites and breathlessness. He then had been initiated on thalidomide 50 mg/d, incremental to 50 mg weekly to a maximum of 200 mg/d, with dexamethasone 20 mg/m^2 on days 1–4, increased monthly for 4 months. This treatment was discontinued due to worsening neuropathy, after which he was switched to lenalidomide 25 mg/d for days 1–21, cyclophosphamide 300 mg/m^2 each day on days 1–5, and dexamethasone 20 mg/m^2 each day on days 1–5 each month.

Clinical examination revealed scleroderma-like skin thickening, muscle wasting, and grade 3 ascites with splenomegaly. Blood tests revealed hemoglobin 9.8 g/L, total leukocyte count 3,800/L, platelet count 80,000/L, total bilirubin 0.8 g/dL, alanine aminotransferase 22 U/L, aspartate aminotransferase 18 U/L, alkaline phosphatase 121 U/L, blood urea nitrogen 28 mg/dL, serum creatinine 1.2 mg/dL, and serum sodium 132 mmol/L. Urine routine showed 1+ albumin, and 24-hour urine for protein revealed microalbuminuria. Workup for chronic infections, autoimmune diseases, and prothrombotic and myeloproliferative disorders, including antiphospholipid syndrome, were noncontributory. Ultrasound of the abdomen revealed the

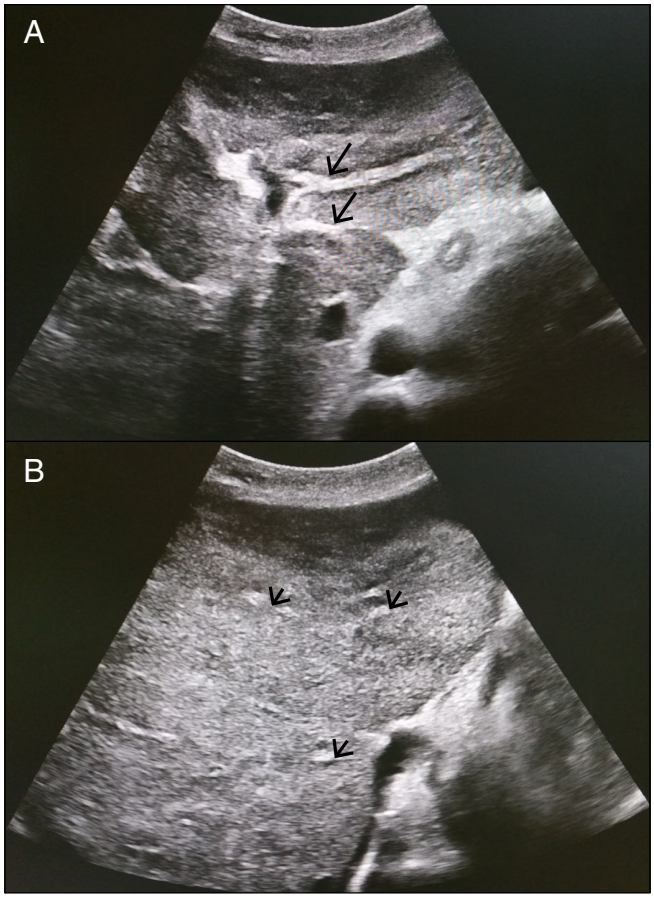

Figure 1. Ultrasound of the abdomen showing (A) the irregular surface of the liver and hyperechoic thickened walls of the main portal vein (arrows), and (B) thickening of the portal vein branches and venules (arrows), giving a Symmer's pipe-stem pattern of periportal fibrosis.

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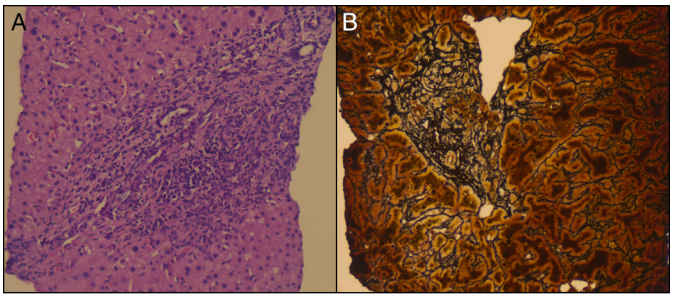

Figure 2. Liver biopsy showing (A) mild portal fibrosis, dilated portal veins, plasma-rich portal triaditis, and mild hepatocellular cholestasis, and (B) increased reticulin in the absence of cirrhosis.

irregular surface of the liver with splenomegaly and hyperechoic thickened walls of the main portal veins, its branches, and venules, giving a pipe-stem (Symmer's) pattern of periportal fibrosis, indicative of hepatic schistosomiasis (Figure 1). Endoscopy showed grade 2 esophageal varices. Serology for *Schistosoma* species was negative; repeated stool and urine examinations did not reveal eggs of the *Schistosoma* species. Hepatic venous pressure gradient was 6 mm Hg. Transjugular liver biopsy showed mild portal fibrosis, dilated portal veins, plasma-rich portal triaditis, mild hepatocellular cholestasis, and increased reticulin in the absence of cirrhosis, features of autoimmune hepatitis, eggshells, granuloma, or viral inclusions (Figure 2). Serology for autoimmune hepatitis including total immunoglobulin G were non-contributory. A diagnosis of idiopathic noncirrhotic portal hypertension (INCPH) associated with POEMS syndrome was made. Because one year of medical therapy for POEMS syndrome did not improve ascites, and given the additional diagnosis of INCPH, the patient was advised to undergo transjugular intrahepatic portosystemic shunt (TIPS) for control of ascites.

INCPH is a rare disease characterized by portal hypertension (PHT), and 50% of cases are associated with immunological and prothrombotic disorders, chronic infections, and exposure to medications or toxins with ascites.[1] In the presence of long-standing and refractory ascites and absence of cirrhosis, it is important to consider other causes of PHT. INCPH in POEMS syndrome has been described only 4 times in the literature.[2] Another rare liver association described with POEMS syndrome is hepatic vein thrombosis.[3] Ascites in POEMS syndrome typically demonstrate a low serum-ascites albumin gradient and low protein, and they usually respond to medical therapy.[4,5] Histology of the liver can show hypoplastic or dilated portal tracts, fibrous thickening of the portal vein wall, paraportal shunting vessels, mixed portal inflammation, sinusoidal dilatation, congestion, and pericellular fibrosis.[6] The Indian subcontinent is a low-risk region for human schistosomiasis, and PHT presentation is not described. The absence of snails of *Bulinus* species and the resistance of common snails to miracidia of human schistosomes are implicated in the relative absence of human schistosomiasis in India.[7] Pipe-stem fibrosis is classically described on imaging and gross morphology in hepatosplenic schistosomiasis. Liver histology usually reveals intense granulomatous reaction near fibrotic portal tracts with or without eggs and eggshell remnants. The association of INCPH with POEMS syndrome is very rare, and presentation of PHT with pseudo pipe-stem fibrosis is novel. TIPS improves ascites in patients with INCPH.[8]

DISCLOSURES

Author contributions: CA Philips wrote the manuscript and is the article guarantor. P. Shenoy provided the clinical images. R. Paramaguru revised the manuscript and provided the histopathology images. P. Mahadevan and P. Augustine edited the manuscript.

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