Recurrent bilateral pleural effusions without ascites as an initial presentation of Budd-Chiari syndrome

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Budd-Chiari syndrome (BCS) typically presents with abdominal pain, ascites, and hepatomegaly [1-2]. Bilateral pleural effusion without ascites is a rare presentation of BCS [3]. A 51-year-old female presented with right-sided abdominal pain with recurrent bilateral pleural effusions for 3 months. She was referred for recurrent dyspnea that required repeated thoracentesis. Pleural fluid analysis revealed a transudative process. Her physical exam showed decreased breath sounds in bilateral lung fields and right-sided abdominal tenderness. There was no bulging flanks or shifting dullness. Laboratory tests included: hemoglobin 17.2 g/dL, hematocrit 52%, erythropoietin 7.2 mIU/mL (2.5-18.5), mean corpuscular volume 81 fL, aspartate transaminase 116 U/L, alanine transaminase 71 U/L, alkaline phosphatase 190 U/L and bilirubin of 2 mg/ dL. An echocardiogram showed preserved ejection fraction. CT scan of the chest and abdomen revealed bilateral pleural effusions (Fig. 1A) and hepatomegaly with caudate lobe enlargement (Fig. 1B).

Abdominal ultrasound with Doppler demonstrated dampened hepatic venous waveforms. Hepatic venogram demonstrated central hepatic vein branches that were patent but with very slow flow. The peripheral hepatic vein branches were irregular suggestive of BCS (Fig. 2). A transjugular intrahepatic portosystemic shunt was deployed which resulted in improvement of the hepatic portal venous gradient from 18 to 3 mmHg. The liver biopsy demonstrated centrilobular congestion and fibrosis consistent with BCS. Polycythemia vera was diagnosed as the predisposing etiology for the patient's BCS. She was positive for JAK2 mutation. She underwent therapeutic phlebotomy and long-term anticoagulation. There was no recurrence of pleural effusions after 2 months on follow up.



Figure 1 (A) CT chest with bilateral pleural effusions. (B) CT abdomen showing hepatomegaly and caudate lobe enlargement



Figure 2 Angiography showing spider collaterals

Conflict of Interest: None

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