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EDUCATION AND IMAGING

# Hepatology: Portal vein cavernoma imitating cholangiocarcinoma in a patient with erythropoietic protoporphyria

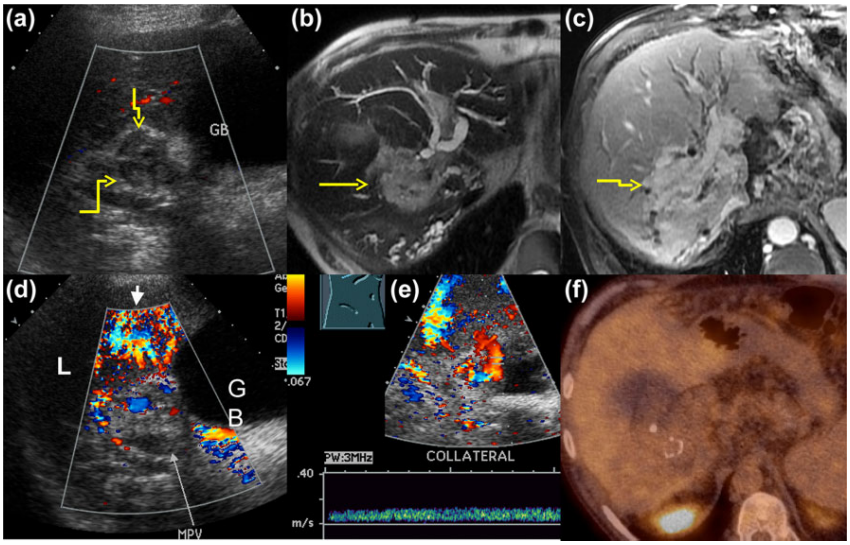


**Figure 1** A mass (arrows) in the porta hepatis near the gallbladder (GB) was found on ultrasound (a) and MRI (b and c), showing a high T2-weighted signal (b) and a marked diffuse enhancement on post contrast T1-weighted images (c). Color Doppler imaging showed thrombus within the main portal vein (long arrow) and a highly vascular lesion (short arrow) in the porta hepatis extending toward the right hepatic lobe (d). Spectral Doppler of the collateral vessels in the porta hepatis demonstrates portal venous-type waveforms (e). The mass was not FDG-avid (f).

A 56-year-old man with a medical history of erythropoietic protoporphyria (EPP) and portal hypertension of unknown etiology manifested by ascites, portosystemic encephalopathy (PSE), and a history of esophageal variceal bleed presented with chronic vague right-sided abdominal pain. He underwent abdominal ultrasound that showed a large, irregular mass within the porta hepatis extending into the right hepatic lobe (Fig. 1a). Subsequent magnetic resonance (MRI) cholangiopancreatography/magnetic resonance imaging of the abdomen with contrast demonstrated a 6.8-cm mass within the porta hepatis extending into the right hepatic lobe with delayed enhancement and intrahepatic biliary dilatation favoring cholangiocarcinoma (Fig. 1b and c). Alpha fetoprotein and Ca 19-9 levels were normal. He underwent percutaneous liver biopsy of the mass, which showed portal fibrosis with bile ductular proliferation, but no evidence of protoporphyrin, cirrhosis, nor malignancy. There was concern for inadequate biopsy as no tumor was found. The abdominal ultrasound was repeated with spectral Doppler for better delineation between the lesional and hepatic vasculature that showed thrombosis of the main, right, and left portal veins associated with multiple collateral vessels including gallbladder wall varices (Fig. 1d). Doppler waveforms were consistent with collateral vessels from cavernous transformation due to portal vein thrombus (Fig. 1e); therefore, the patient could confidently be diagnosed with a cavernoma rather than malignancy even though the biopsy was initially thought to be inconclusive. The mass was also not $_{18}$Fluoro 2-Deoxy-D-Glucose (FDG) avid on positron emission tomography–computed tomography imaging (Fig. 1f), unlike most large cholangiocarcinomas.

Portal vein cavernomas are rare phenomena resulting from portal vein thrombosis and can be mistaken for malignancy. A cavernoma is a tumor-like mass composed of a venous meshwork bypassing a thrombosed portal vein. Spectral Doppler evaluation is particularly useful in distinguishing cavernoma from cholangiocarcinoma as it can differentiate portal venous blood flow from arterial flow seen in tumor neovascularity. Multiple and large collaterals can form a "pseudotumor" at the porta hepatis with features of portal venous blood flow including continuous flow, usually with minimal cardiac or respiratory variation, and low mean and peak velocities (16–40 cm/s). Doppler waveforms in cholangiocarcinoma, however, would reflect arterial blood flow. In summary, we present a case of EPP with non-cirrhotic portal hypertension due to portal vein thrombosis of unknown etiology, with formation of a large, mass-like cavernoma. The mass, originally diagnosed as cholangiocarcinoma, was later found to be a cavernoma using Doppler ultrasound. As a result, repeat biopsy of the mass was avoided, which could have been associated with an increased risk of bleeding.

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