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Case Report

Transhepatic embolization of a congenital intrahepatic portosystemic shunt for the treatment of hepatic encephalopathy in a noncirrhotic patient using Amplatzer vascular plug device

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ABSTRACT

A 73-year-old male with no history of liver disease was hospitalized for weakness, confusion, ataxia, and new onset hepatic encephalopathy with hyperammonemia. After management with lactulose and rifaximin, his symptoms persisted, and he underwent transjugular liver biopsy. Biopsy showed normal liver, but a portosystemic shunt was incidentally identified on postbiopsy venogram. The patient underwent occlusion of the shunt with two Amplatzer vascular plugs and four Nester coils. Following embolization, the patient's symptoms resolved completely. Our case reports one of the oldest adults to present with symptoms from a congenital portosystemic shunt. Congenital portosystemic shunts can be considered in patients with new onset hepatic encephalopathy in the absence of underlying liver disease. Prognosis after embolization of congenital portosystemic shunt is great, and embolization may result in full reversal of symptoms.

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Case report

A 73-year-old man with no history of liver disease presented to our hospital in July 2015 with weakness, confusion, asterixis, and ataxia. His additional medical history was significant for congestive heart failure, hypertension, and atrial fibrillation. He had been recently seen by an outpatient physician for similar symptoms and had a workup that included a normal vitamin B12, folate, antinuclear antibody, human immunodeficiency virus, rheumatoid factor, cortisol, and thyroid-stimulating hormone. Laboratory studies during his admission showed an elevated serum ammonia (117 umol/L), hyperbilirubinemia (2.4mg/dL), and elevated INR (3.7). He was hospitalized and seen by gastroenterology and neurology. He had no history of trauma in the liver. After an extensive workup, his encephalopathy was poorly explained and thought to be due to hepatic congestion caused by congestive heart failure and cirrhosis after a remote history of alcohol

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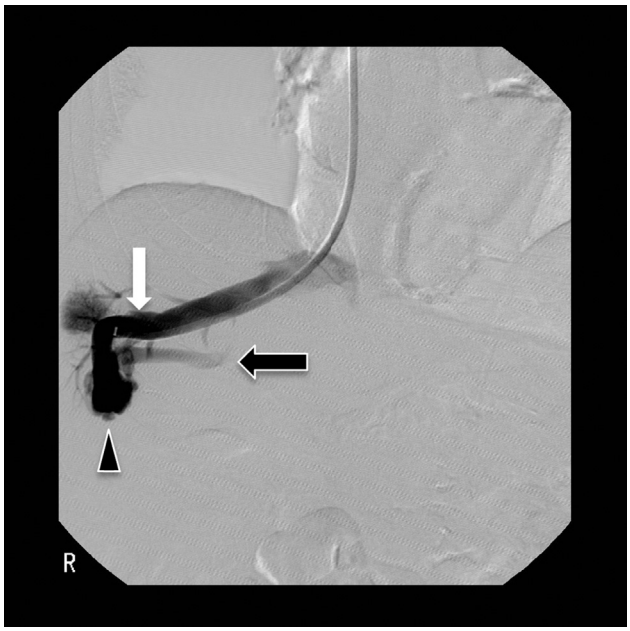

Fig. 1 – Transjugular hepatic venogram demonstrates a dilated area (arrowhead) of right hepatic vein (white arrow) communicating with the right portal vein (black arrow.)

abuse 40 years prior. He was started on lactulose and rifaximin and noted some improvement of his symptoms. The patient continued to have increasing difficulty ambulating, slurred speech and generalized weakness and was followed by a hepatologist following discharge from the hospital. His laboratories and physical examination findings were not consistent with cirrhosis, so it was suggested that he undergoes a transjugular liver biopsy with hepatic venous pressure measurement to further characterize his liver disease.

A transjugular liver biopsy was done in November 2015, and during the procedure, corrected wedge pressures were found to be 5–6 mm Hg, biopsy samples were taken, and a venous abnormality was noted on postbiopsy hepatic venogram (Fig. 1). The lesion was characterized as a dilated area of hepatic vein in the right lobe and communicating with the right portal vein, and further imaging was recommended.

Pathology report of the biopsy showed normal liver with minimal nonspecific portal chronic inflammation and no evidence for congestive hepatopathy. Follow-up CT of the abdomen showed hepatic segment V/VI portosystemic shunt with a likely vascular malformation measuring 2.6 cm (Fig. 2).

Given the patient's history of normal biopsy and hepatic vein pressures, lack of trauma, and absence of malignancy, the malformation was thought to be congenital. At the age of 73 years, our case reports one of the oldest patients to present with symptoms from a congenital intrahepatic portosystemic shunt.

The case was evaluated by the interventional radiology department and was scheduled for a percutaneous transhepatic embolization of the intrahepatic portosystemic shunt. Ultrasound guidance was used to access the right portal vein, and a 6-Fr MAK-NV introducer system (Merit Medical, South Jordan, UT) was used to place a 6-Fr × 25 cm sheath (Terumo, Elkton, MD). During the procedure, contrast injection portal

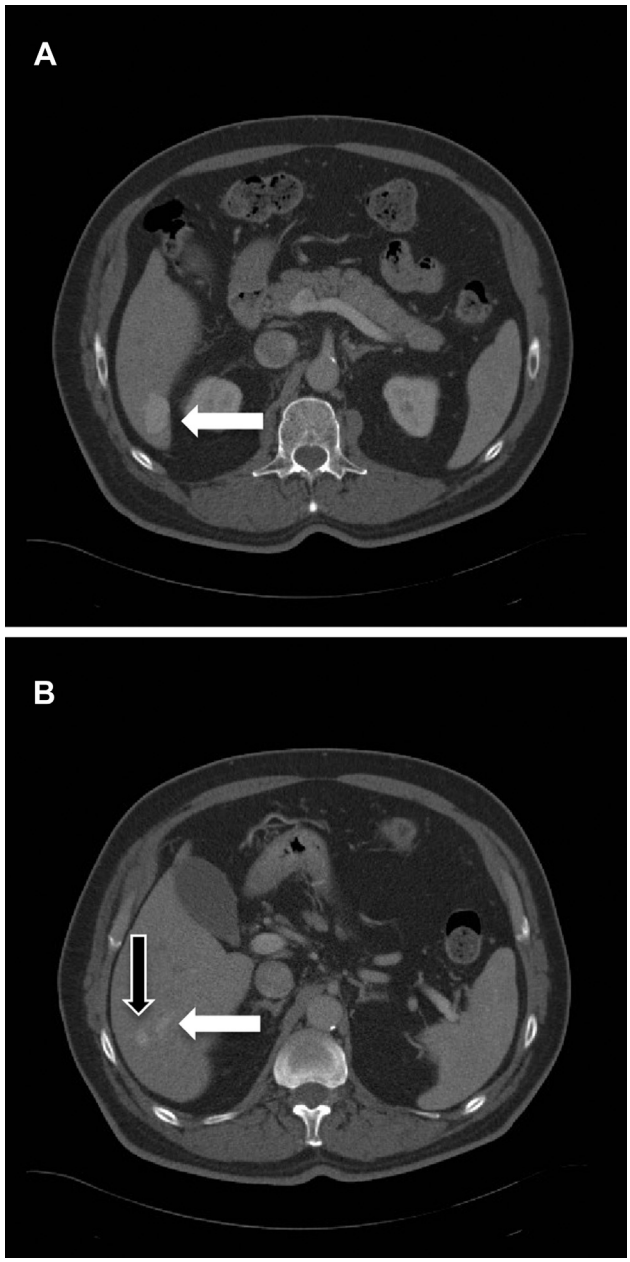

Fig. 2 – (A) Venous phase axial CT scan demonstrates an aneurysmal connection (arrow) between the right posterior portal and right hepatic veins in the segment VI. (B) Superior slice of the CT scan depicts the right posterior portal vein (white arrow) and the right hepatic vein (black arrow) converging into the aneurysmal connection.

venogram using a 5-Fr Omni-flush catheter (Angiodynamics, Queensbury, NY) demonstrated direct fistula from the distal right posterior portal vein branch to the right hepatic vein (Fig. 3).

Given the risk of coil migration due to high-flow fistula from the portal vein to the hepatic vein, an Amplatzer vascular plug was used. An 8-mm Amplatzer 4 vascular plug (AGA Medical Corporation, Plymouth, MN) was advanced through the 5-Fr Simmons-1 catheter (Angiodanymics, Queensbury, NY) into the one of the two feeding portal vein

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