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Letters to the Editor

Extrahepatic shunt. Unusual cause of hepatic encephalopathy

Key words: Portosystemic shunt. Encephalopathy.

Dear Editor,

The most common cause of hepatic encephalopathy (HE) is chronic liver disease.

Although there are rare extrahepatic diseases that may present with hepatic encephalopathy as a clinical manifestation, we report on a case of a patient with HE secondary to portosystemic shunt.

Case report

A 76 year-old woman with a history of diabetes mellitus type 2 and surgery for endometrioma and cholecystectomy. The patient was admitted with symptoms of low level of consciousness without neurological deficit. She reported similar symptoms with spontaneous resolution. The patient denied other symptoms of hepatic decompensation. Blood test showed high ammonium levels with other analytical parameters within normal (complete liver analysis and urine toxic). A CT and brain MRI, EEG and CSF study were performed and all test results were normal. Abdominal doppler ultrasound described: normal liver size with slightly lobed edges (FibroScan® of 4.8 kPa [F0]) and presence of reverse flow in the superior mesenteric vein; the porta vein was permeable. A magnetic resonance angiography confirmed these findings and revealed the presence of a communication between ileocolic vein, distal branch of the superior mesenteric vein (SMV), and right gonadal vein draining the latter to the inferior vena cava (IVC). Portal catheterization was performed obtaining a hepatic venous pressure gradient of 4.5 mmHg. A gastroscopy ruled out the presence of esophageal varices**.** It led to the diagnosis of HE secondary to portosystemic shunt. We proceeded to close the communication by embolization by interventional radiology department. After one year of clinical follow-up, the patient is asymptomatic without other episodes.

Discussion

Portosystemic shunts in non-cirrhotic patients are attributed to embryological origin, portal vein thrombosis, trauma or previous abdominal surgery. There are two types of congenital portosystemic shunts: Intrahepatic and extrahepatic. The first is usually due to communication between branches of the portal vein and the hepatic veins or IVC. Extrahepatic can be type 1 (complete derivation of portal blood to the systemic territory in the absence of intrahepatic portal branches; 1a: Splenic vein and SMV drain separately to the IVC, 1b: Drain forming a common trunk) and type 2 (intrahepatic portal circulation preserved; there is a deviation of portal flow to systemic venous circulation). Extrahepatic type 1 is more common in girls and is asso-

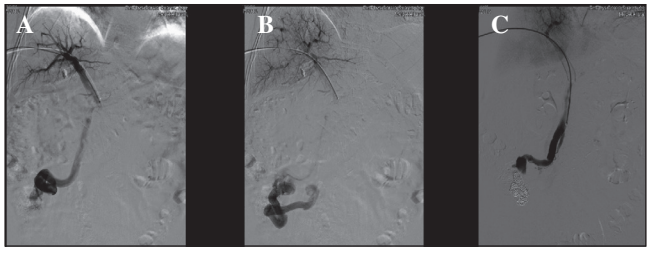

Fig. 1. Portography sequence. A and B. Abnormal communication between SMV (ileocolic vein) and IVC (right gonadal vein). C. Image after closure of the communication.

ciated with multiple malformations (heterotaxia, polysplenia or congenital heart defects). Type 2, without gender partnership, is not usually associated with malformations.

Clinical manifestations are secondary to abnormal development of the liver in which low blood flow can lead to liver ischemia and compensatory increased arterial blood flow may appear as focal nodular hyperplasia or adenoma. Another clinical manifestation due to the presence of shunt is HE especially in older adults, whose brain is more sensitive to hyperammonemia (2). This disease should be suspected in patients with behavioral disturbances and fluctuations in the level of consciousness without evidence of chronic liver disease with elevated blood ammonium. Initially an abdominal Doppler ultrasound will be performed and also a CT angiography or magnetic resonance angiography as a confirmatory test. Symptomatic patients, with liver dysfunction or shunt ratio > 60% even in asymptomatic patients (3) should be treated. In type 1, treatment is liver transplantation, and in type 2, treatment is closure by surgery or embolization.

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