Liver resection for the treatment of a congenital intrahepatic portosystemic venous shunt

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ABSTRACT: Intrahepatic portosystemic shunts (IPSS) are rare congenital anomalies arising from disordered portal vein embryogenesis. It has been described in both children and adults and may be asymptomatic or be associated with a variety of neurophysiological and pulmonary complications. When recognized, early intervention to occlude the shunt will reverse the associated complications. Literature review reports of surgical and radiological occlusion of the shunt, but due to its rarity, a standard therapeutic protocol has not been established. A case of a 38-year-old woman with abdominal pain and low grade encephalopathy, diagnosed with an IPSS and treated by right hepatectomy was reported.

(***Hepatobiliary Pancreat Dis Int 2016;15:329-333***)

KEY WORDS: intrahepatic portosystemic shunt; liver resection

Introduction

Congenital intrahepatic portosystemic shunt (IPSS) is a rare abnormal communication between the intrahepatic portal and hepatic veins.[1] Four types have been identified by Park et al based on the morphology of the shunts, which, by definition, have to be at least 1 mm in diameter.[2] Those include a single vessel communication, which can be either between a main branch of the portal vein and inferior vena cava (IVC) (type 1), peripheral location in one segment (type 2), or through an aneurysm (type 3), and multiple small communications distributed diffusely in both lobes (type 4).[2, 3]

IPSS may occur as a result of liver pathology, trauma, intervention, or rarely as a congenital anomaly as described in this report. The latter develops as a result of abnormal portal vein embryogenesis.[4, 5] Patients diagnosed in early life with congenital IPSS commonly present with secondary complications including neurocognitive dysfunction, developmental delay, hypoxia and focal liver lesions (e.g. adenoma, focal nodular hyperplasia). Of note, it has been reported incidentally on imaging in adults presenting with low grade encephalopathy, pulmonary hypertension, hypoglycemia, chronic abdominal pain, and complex neurological disorders, which develop as a consequence of porto-systemic shunting and elevated ammonia levels.[4-9]

CT, MRI and ultrasound can be used to confirm the diagnosis plus measurement of arterial ammonia levels in asymptomatic patients. Conservative management has been proposed previously for those who are able to tolerate their elevated ammonia levels with a stable shunt; however, intellectual and psychosocial function may be improved by early intervention. For patients who do not respond to medical treatment (dietary control, non-absorbable antibiotics), and develop complications, such as encephalopathy or who have an increasing shunt size, definitive treatment should be considered.[8, 10] Reported interventions include image guided endovascular occlusion of the shunt by interventional radiologists, surgical ligation and excision either by formal liver resection or even transplantation. Various endovascular methods have been reported using a wide range of embolization materials.[8, 10]

A patient with low grade intermittent encephalopathy and chronic abdominal pain, with a large congenital IPSS, was treated with right hepatectomy. The case of this patient and review of the literature are presented.

Case report

A 38-year-old woman presented to her local hospital with a history of right upper quadrant pain of short

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doi: 10.1016/S1499-3872(16)60067-X
Published online January 19, 2016.

duration and a visible superficial vein along the midline axillary line of the abdominal wall. There was no history of abdominal trauma. Subsequent CT imaging revealed a large IPSS in segment 8 of the liver with a communication between the right anterior branch of the portal and the middle hepatic veins, measuring 24 mm in diameter (Fig. 1). The liver parenchyma was unremarkable.

There was no medical history other than iron deficiency anemia. Liver biochemistry and arterial ammonia level were within normal limits. No treatment was given and the patient was reviewed one year later for continuing abdominal pain and persistence of the dilated vein of the abdominal wall. It was suspected that an aberrant vessel related to her shunt was communicating with this superficial vein. She underwent laparoscopy, which failed to identify any abnormal venous communication with the abdominal wall. No dilated veins were seen within the abdominal cavity or on the parietal peritoneum. The possibility of embolization of the shunt was discounted by the radiologists due to the large size and the high risk of inadvertent migration of embolic agents (coils, glue etc.) to the pulmonary artery. Of note, the dilated vein on the abdominal wall disappeared after laparoscopy, but her symptoms were unchanged.

Over the next 2 years, the patient had further episodes of right upper quadrant pain, weight loss, alteration in bowel habit, and persistent low iron levels despite supplementation. She was extensively investigated for her abdominal pain in 3 other hospitals and no other cause was identified. Subsequently, she represented to our unit with intermittent low grade hepatic encephalopathy, and a raised arterial ammonia level (61 μmol/L). MRI of the brain was unremarkable. Electroencephalogram confirmed the metabolic nature of the encephalopathy and a decision was made to undertake right hepatectomy.

She underwent formal right hepatectomy and during surgery a persistent ductus venosus was also identified and ligated. The liver otherwise appeared normal and there was no portal hypertension. The portal vein pressure rose from 7 to 9 mmHg post ductus venosus ligation and right hepatectomy. The patient made an uncomplicated recovery and was discharged from hospital 6 days after operation.

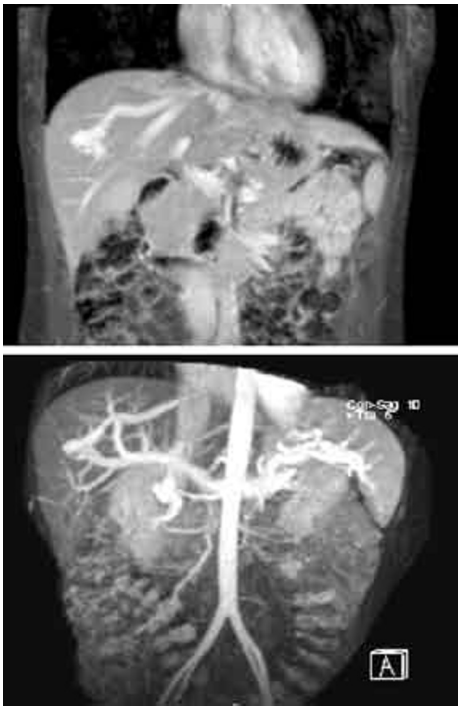

Fig. 1. CT venogram, coronal view. Presence of a vascular anomaly within the anterior aspect of the right liver, this is characterized by a portosystemic shunt from the anterior division of the right portal vein into the middle hepatic vein via the segment 8. The size of the nidus of the anomaly measures up to 24 mm.

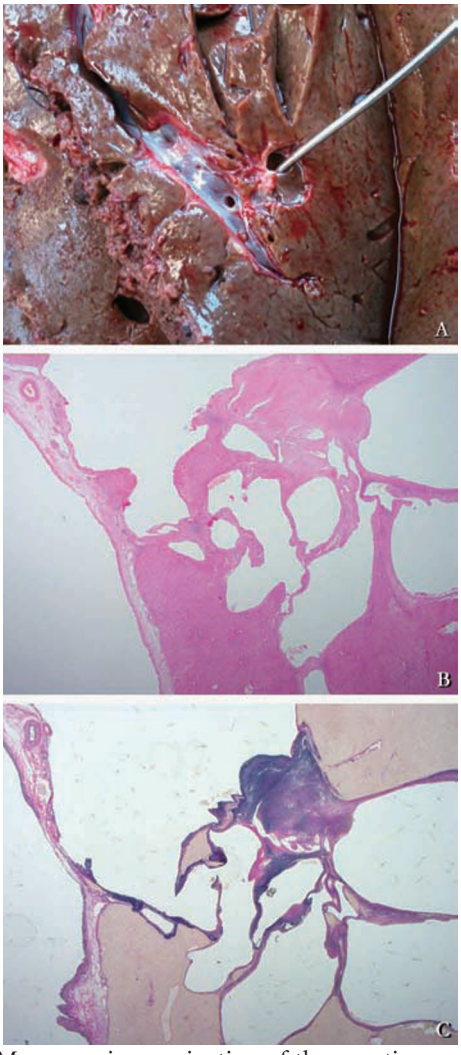

Fig. 2. A: Macroscopic examination of the resection specimen revealed an abnormal juxtaposition of a large hepatic vein to a large intrahepatic portal vein branch. The latter is demonstrated by the probe, which connects to the portal vein branch; **B&C**: Low magnification view of clusters of dilated vessels showing a nodal point of sclera-hyaline tissue (2B, HE) rich in elastic fibres (2C, Elastic Van Gieson) (Original magnification ×20).