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Liver nodules after portal systemic shunt surgery for extrahepatic portal vein obstruction in children

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Abstract

Background: Liver nodules have been reported after portal systemic shunt surgery (PSSS) in animal experiments or in humans with liver cirrhosis. The aim of our study was to assess the incidence of liver nodules after surgery for extrahepatic portal vein obstruction (EHPVO) in children without associated liver disease.

Methods: We retrospectively reviewed the charts of 45 children who had surgery from 1979 to 2005 for EHPVO in our institution, consisting of 38 PSSS and 7 portal reperfusion procedures (PRPs). We assessed the presence of liver nodules on ultrasonography.

Results: Of 45 patients, 7 (15%) had liver nodules during a median of 80 months of follow-up. All the nodules occurred after PSSS. Five nodules were subjected to biopsy; we found 2 liver cell adenomas and 3 focal nodular hyperplasias.

Conclusions: In this study, liver nodules occurred in 18% of cases after PSSS for EHPVO in children and not after PRP. As many children have undergone PSSS throughout the world, the presence of liver nodules should be considered during the follow-up of those patients.

Extrahepatic portal vein obstruction (EHPVO) is a major cause of portal hypertension in children [1]. Of EHPVOs in children, 65% are of unknown origin; the others are caused by neonatal umbilical vein catheterization, abdominal sepsis, or thrombotic coagulopathy [2]. In EHPVO, the liver parenchyma is supposed to be normal [2]. A frequent consequence of EHPVO is portal hypertension, which carries a high risk of gastrointestinal bleeding [3]. Prevention of gastrointestinal bleeding may be ensured by surgical procedures, which may be either portal systemic shunt

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surgery (PSSS) or one of the portal reperfusion procedures (PRPs) derived from the original mesenteric to left portal vein bypass.

Portal systemic shunt surgery for EHPVO provides a success rate of 87% to 98% on portal hypertension [2,4]. A midterm risk of pulmonary arterial venous shunting or pulmonary artery hypertension has been reported to be as low as 1% after PSSS for EHPVO, making this surgical procedure efficient and safe, provided that there is careful cardiac follow-up [4-6]. More recently, the PRP techniques have been promoted because they restore a physiologic portal circulation.

Liver regenerative lesions were reported after PSSS in animal experiments or in humans with intrahepatic portal hypertension [7-9]. Liver cell adenoma and focal nodular hyperplasia were also reported in patients with congenital portal systemic shunts [10-12]. As liver cell adenoma can develop malignant transformation or bleeding [13,14] and as many children have undergone PSSS throughout the world before the era of PRP, the presence and the natural history of liver nodules should be considered in the follow-up of those patients who underwent PSSS for EHPVO during childhood. The aim of our study was to assess the incidence of liver nodules after PSSS or PRP for EHPVO and to analyze their clinical, imaging, and pathologic characteristics.

1. Patients and methods

1.1. Design of the study

We analyzed the records of 69 children who had surgery for EHPVO at our institution between January 1979 and December 2005.

1.2. Eligibility criteria

Extrahepatic portal vein obstruction was defined by (1) portal vein obstruction or portal cavernoma on ultrasound scan (US), (2) esophageal varices on gastroscopy, (3) no other parenchyma liver disease, defined by the absence of hepatocellular lesions on liver US or biopsy, negative hepatitis B virus and hepatitis C virus serology, and no history of tumor before surgery. Patients with biliary tract dilatation on US owing to the compression by the cavernoma were included only if the operative biopsy showed absence of liver parenchyma lesions.

1.3. Exclusion criteria

Patients with shunt thrombosis and unsuccessful redo surgery were excluded from the study. Patients with less than 1-year follow-up after initial surgery were excluded from the study.

1.4. Patient characteristics

Sixty-nine patients met the eligibility criteria, 27 girls and 42 boys, aged from 0 to 186 months at the diagnosis (mean, 53 months). The origin of EHPVO was an umbilical catheter in 20 cases, thrombotic coagulopathy in 4 cases, and idiopathic in 45 cases.

1.5. Surgery

Surgery for EHPVO was scheduled when there was gastrointestinal bleeding or grade III esophageal varices on fibroscopy. The median time between the diagnosis and surgery was 15 months (0-136 months). The median age at surgery was 7 years (10 months to 11years). Portal systemic shunt surgery was performed in 58 cases, mesenteric or portal caval shunt in 42 cases, and splenic or mesenteric renal shunt in 16 cases. Since 1996, when the left portal branch was patent and communicated with the right portal branch, we performed 11 mesenteric to left portal venous bypasses or equivalent PRP. Fifty patients had a surgical liver biopsy during the shunt procedure. Three patients with slight or moderate intrahepatic biliary dilatation were included in the analysis as they had a normal liver biopsy result. One patient had asymptomatic gallbladder lithiasis with a normal biopsy result. He underwent cholecystectomy and was included in the analysis.

1.6. Follow-up

Follow-up was defined as the time from surgery to the last visit. Nineteen patients were lost for follow-up after 1 year and were subsequently excluded from the analysis—1 with PRP and 18 with PSSS.

1.7. Shunt patency

Short-term shunt patency was assessed by a Doppler US before patient discharge. Long-term shunt patency was assessed by a clinical examination, an esogastrofibroscopy, and a liver Doppler US 6 months after surgery and then every year thereafter.

Eight patients had an early shunt thrombosis—3 patients initially treated by a PRP and 5 by PSSS. One patient with PRP and thrombosis underwent early successful conversion to PSSS and was included in the group PSSS. The 2 other patients with PRP remained thrombosed at last follow-up and were excluded. Of 5 patients with PSSS, 3 remained thrombosed at last follow-up and were excluded (Fig. 1).

1.8. Judgment criteria

During the follow-up, nodule assessment was made by a yearly US. We detailed clinical data, imaging, and when available, pathologic patterns of the nodules. Imaging of the nodules was in favor of liver cell adenoma if the nodule was