Case Report

A case of portal biliopathy in a young patient with portal cavernoma secondary to neonatal umbilical vein catheterization✩

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ABSTRACT

Portal biliopathy (PB) refers to biliary obstruction caused by cavernous transformation of the portal vein (CTPV). CTPV occurs most frequently in patients with liver cirrhosis or malignancy. Less common causes include congenital malformations and neonatal umbilical vein cannulation. We present a case of portal biliopathy in a 28-year-old man with CTPV secondary to umbilical vein catheterization in neonatal age. The case illustrates portal biliopathy as a late complication of neonatal invasive procedures and highlights the importance of a multimodality imaging approach to achieve a prompt diagnosis.

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Introduction

Portal biliopathy (PB) refers to biliary obstruction caused by vascular compression and ischemic damage to the biliary tree in patients with cavernous transformation of the portal vein (CTPV).

Clinical presentation is heterogeneous. While most of the patients remain asymptomatic, others may develop jaundice, choledocholithiasis, cholangitis or hemobilia, even many years after the diagnosis of CTPV.

Case report

A 28-year-old male was admitted to our Emergency Department with nausea, vomiting and epigastric pain. No fever or change in bowel habits was noted.

His past medical history revealed a preterm 26-weeks, iatrogenic portal vein thrombosis and CTPV caused by umbilical vein catheterization due to severe neonatal asphyxia associated with hyaline membrane disease.

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Abdominal US showed cavernomatous transformation of the portal vein and dilatation of the common bile duct (CBD), with a maximum diameter of 21 mm (Fig. 1).

US also revealed an overstretched gallbladder with normal walls and an infundibular gallstone of 2.5 cm.

Severe splenomegaly with multiple hyperechoic spots in the splenic parenchyma was also noted.

A contrast-enhanced CT scan of the abdomen was performed to clarify US findings.

CT confirmed cavernous transformation of the portal vein with multiple venous collaterals at the porta hepatis, in the splenic lodge and in periumbilical region. The ectatic veins caused compression of the pre-papillary CBD and a proximal dilatation of intra- and extrahepatic bile ducts (CBD transverse diameter 20 mm) (Fig. 2).

A non-contrast magnetic resonance cholangiopancreatography (MRCP) (Fig. 3) was performed for a better evaluation of the biliary tree.

MRCP confirmed multiple ectatic veins at the porta hepatis causing compression of the CBD and upstream dilatation of the intrahepatic and extrahepatic bile ducts.

The main pancreatic duct was normal.

MR also demonstrated multiple tiny hypointense foci in the splenic parenchyma indicative of siderotic nodules (Gammagandy bodies).

Endosonography (EUS) ruled out choledocholithiasis and depicted a normal Vater’s papilla.

Numerous venous collaterals compressing the CBD were confirmed.

CTPV was complicated by bleeding esophageal and gastric varices (for which he received endoscopic sclerotherapy) and severe splenomegaly (treated with multiple splenic embolizations 13 years before).

Laboratory tests revealed increased total, direct and indirect bilirubin, respectively 3 mg/dl (normal range 0.3-1.2), 0.86 mg/dl (normal range <0.4) and 1.86 mg/dl (normal range 0.1-0.8) and pancytopenia.

Fig. 1 – Liver US depicting severe dilatation of the common bile duct (21 mm, white arrow).

Fig. 2 – Axial (A) and coronal (B) contrast-enhanced portal phase CT scan of the abdomen, demonstrating cavernomatous transformation of the portal vein with multiple varicose venous collaterals at the hepatic hilum causing compression on the common bile duct and a proximal dilatation of the biliary tree (white arrowheads in a and b). Notice the severe splenomegaly (asterisks in A and B).
The patient was treated with pharmacological therapy and was discharged after 3 days in good clinical conditions. At 1-month follow-up, blood tests were normal.

Discussion

Cavernous transformation of the portal vein (CTPV) is caused by chronic portal vein obstruction which leads to the formation of periportal venous collaterals. It was first reported in 1869 by Balfour and Stewart and is also referred to as “portal cavernoma” due to the sponge-like appearance of the portal vein [1–3].

Portal vein thrombosis (PVT) and cavernoma formation are rare complications following neonatal liver abscess and sepsis, prior umbilical cannulation, and infection [4–9].

After umbilical cannulation, CPTV may remain asymptomatic for years before giving any clinical presentation. It may present with gastroesophageal variceal bleeding, splenomegaly, and thrombocytopenia.

Portal biliopathy (PB) refers to cholangiographic abnormalities in patients with portal cavernoma occurring as a consequence of pressure on bile ducts by tortuous paracaval, epiploic, and choledochal collaterals. These collaterals enlarge in an attempt to decompress the venous blockage caused by PVT [10]. Bile duct ischemia may occur due to prolonged wall compression or insufficient blood supply; fibrotic cavernoma may cause encasement of bile ducts. Although biliary symptoms appear to be uncommon in patients with portal cavernoma, almost all of them show abnormalities at endoscopic retrograde cholangiography.

PB has been reported in 70%-100% of patients with extrahepatic obstruction of the portal vein (EHOPV). PB is less common in cirrhotic portal hypertension, probably because in cirrhosis the obstruction of the venous circulation occurs at the level of the hepatic sinusoid, causing collaterals to form distant from the extrahepatic bile ducts [11].

In patients with EHFVO, 5%-38% develop symptomatic PB, presenting with cholestasis, jaundice, gallstones, cholangitis. PB is a progressive condition that develops late in the course of portal hypertension and may progress to secondary biliary cirrhosis [12].

Blood tests, color Doppler US, MRCP and ERCP are the most useful diagnostic tools for the evaluation of portal biliopathy. Treatment strategies are directed to symptomatic patients only. Asymptomatic patients do not need any treatment, especially if liver function tests are normal. Pharmacological therapy is the first-line treatment. The placement of a biliary stent with balloon dilatation is recommended in patients who don’t benefit from drugs.

In the presence of symptomatic biliary obstruction not susceptible to endoscopic therapy, a portosystemic shunt is indicated. Biliodigestive shunts are contraindicated without previous decompression of the portal vein because of the high risk of bleeding [10,11,13–15].

Conclusions

The case illustrates portal biliopathy as a late complication of neonatal invasive procedures.

As long-standing portal biliopathy can lead to serious complications such as cholangitis, choledocholithiasis and secondary biliary cirrhosis, radiologists must be aware of this condition to provide an early-stage diagnosis and treatment.

Ethical approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Patient consent

Informed consent was obtained from all individual participants included in the study.

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