

CASE REPORT

Bile duct stricture due to caused by portal biliopathy: Treatment with one-stage portal-systemic shunt and biliary bypass

Melroy A. D'Souza · Devendra Desai · Anand Joshi · Philip Abraham · Sudeep R. Shah

Abstract Portal biliopathy is a rare complication of extrahepatic portal vein obstruction. Jaundice occurs in symptomatic patients with fibrotic strictures. Short-term improvement in such patients can be achieved with endoscopic retrograde cholangio-pancreatography with balloon dilatation and stent placement. Surgery in these patients is traditionally two staged. We report the results of a one-stage procedure combining non-selective portal-systemic shunt surgery with biliary bypass, performed successfully on a 24-year-old man with a tight biliary stricture resulting from portal biliopathy. At 18-month follow up, the patient shows he is doing well, with normal liver function tests.

Keywords Bile ducts · Magnetic resonance cholangiopancreatography · Gall bladder calculi

Introduction

Portal biliopathy is the term used to describe abnormalities of the extrahepatic and intrahepatic bile ducts in patients with portal hypertension.¹ It is more common in patients with extrahepatic portal vein obstruction (EHPVO) compared with other causes of portal hypertension.^{1,2} We report the case of a 24-year-old man with EHPVO with symptomatic bile duct obstruction caused by portal biliopathy, who was managed by a single-stage procedure with portal-systemic shunt and biliary bypass.

Case report

A 24-year-old man presented with upper abdominal pain. He was afebrile, and physical examination revealed jaundice, hepatosplenomegaly and dilated abdominal wall veins. Laboratory tests showed direct hyperbilirubinemia (9 mg/dL) with raised alkaline phosphatase levels (540 U/L). Ultrasonography showed gall bladder calculi, dilated intrahepatic biliary radicles (IHBR), a dilated common hepatic duct, splenomegaly and a thrombosed portal vein with cavernoma formation, and large extrahepatic collaterals. These findings were confirmed on contrast-enhanced CT scan of the abdomen (Fig. 1). Endoscopy revealed grade I esophageal varices.

Magnetic resonance cholangiopancreatography (MRCP) showed similar findings and, in addition, common bile duct (CBD) stricture (Fig. 2). In view of the biliary obstruction and jaundice, endoscopic retrograde cholangio-pancreatography (ERCP) was performed, which revealed a tight mid-CBD stricture with upstream duct dilation. The stricture was dilated and a 10-French plastic stent inserted into the CBD. The patient was discharged with a plan for surgery.

However, the patient presented 8 weeks later with cholangitis caused by a stent blockage. A repeat ERCP demonstrated a persisting tight CBD stricture, which necessitated a stent exchange.

In view of the recurrent biliary obstruction and jaundice, definitive surgical intervention was planned. Surgery would entail a portal-systemic shunt to decompress the collaterals and then a biliary bypass to tackle the tight fibrotic stricture of the bile duct. It was planned that if the shunt procedure was uneventful, a biliary bypass would be performed during the same surgery, if feasible, thus avoiding the need for two laparotomies. At surgery, in addition to the gallstones and large spleen, multiple periportal, pericholedochal, retroperitoneal and splenic hilar collaterals were seen. A splenectomy was performed, followed by an end-to-side proximal spleno-renal shunt. A cholecystectomy was then performed, followed by a choledochotomy above the fibrotic mid-CBD stricture. The bleeding encountered was effectively controlled by a Pringle maneuver and also due

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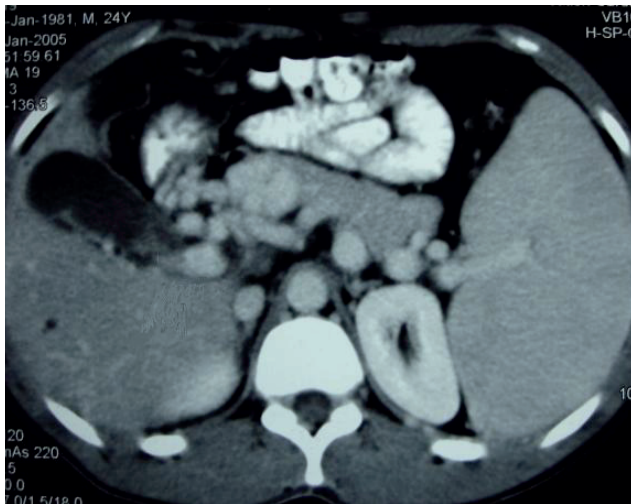


Fig. 1 CT venography showing pericholedochal collaterals

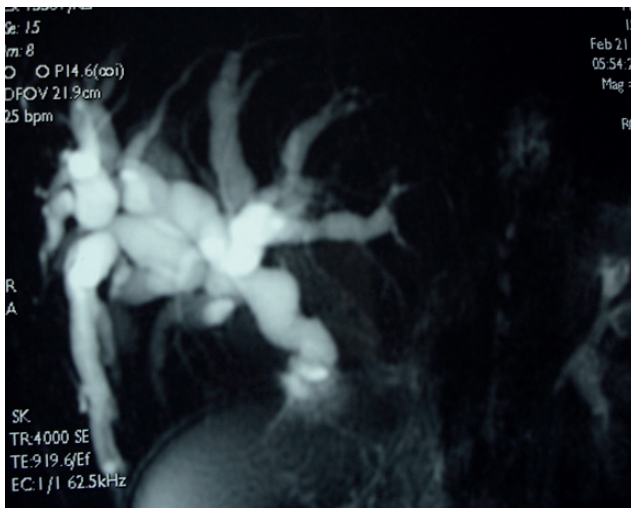


Fig. 2 MRCP showing biliary stricture

to the immediate decompression of the varices produced by the shunt. These varices were then overrun. The Pringle maneuver was in place for 15 min during the choledochotomy to control bleeding from the varices. It completely stopped the bleeding from the pericholedochal collaterals. Finally, a Roux-en-Y hepaticoducho-jejunostomy was performed with 5-0 polydioxanone sutures. The operation time was 4 hours, the blood loss was 1.7 liters, and 4 units of packed blood were transfused. The patient's postoperative recovery was uneventful and liver function tests were normal over a follow up of 18 months. At last follow up, direct bilirubin was 0.8 mg/dL and alkaline phosphatase was 79 U/L.

Discussion

The veins draining the extrahepatic bile ducts are arranged in two systems. The epi-choledochal plexus (Saint) lies on the surface of the bile duct, and the para-choledochal plexus (Petren) runs parallel to it. After a portal vein thrombosis,

new collaterals develop to bypass the obstruction, resulting in the formation of a portal cavernoma. This process also leads to dilation of the venous plexi around the bile duct. The dilated pericholedochal varices compress the bile duct, producing areas of focal narrowing and dilatation.³ Gallstones, bile duct stones and cholangitis may develop due to the obstruction. The inflammatory process gives rise to fibrosis, which leads to stricture formation and eventually to secondary biliary cirrhosis.

The biliary obstruction manifests as jaundice, abdominal pain and sometimes cholangitis.¹ The majority of patients with portal biliopathy are, however, asymptomatic in spite of having elevated alkaline phosphatase levels and showing characteristic changes on ERCP and MRCP.⁴ ERCP findings of portal biliopathy include bile duct strictures and irregularity, dilatation, displacement, and angulations.⁵

Only symptomatic patients with portal biliopathy require treatment. Patients with obstructive jaundice caused by bile duct strictures, in the absence of CBD stones, are recommended to have ERCP with balloon dilatation of the stricture and placement of a biliary stent;⁴ this, however, is not a definitive treatment. Total portal-systemic shunting usually relieves the biliary obstruction. Vibert *et al.* showed a reduction in biliary obstructive symptoms within 3 months of portal-systemic shunt surgery in 70% of patients with portal biliopathy.⁶ However 50% of these patients subsequently required a biliary bypass.⁶ Bile duct obstruction may persist because of fibrosis in the pericholedochal areas and stricture formation. These patients require a biliary bypass, which is rendered to be safer with a portasystemic shunt, as attempts at bypass without a shunt procedure have resulted in complications and even death.¹

In the present case, the tight stricture required a balloon dilatation and stent change. A two-stage repair is usually recommended in such cases, with a shunt surgery followed by biliary bypass.¹ We have described a single-stage procedure. In our patient, the portal-systemic shunt not only decompressed the varices but also served to prevent bowel edema. The hepaticoducho-jejunostomy could then be safely and effectively performed using the Pringle maneuver to control any bleeding. This technical modification, i.e. of performing the shunt and then the biliary bypass with the Pringle maneuver for controlling portal inflow, can be applied in patients with portal biliopathy in whom the biliary obstruction is caused both by compression by pericholedochal collaterals and biliary stricture. This avoids a second surgery.

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 IMAGE

Pedunculated extra hepatic biliary cystadenoma mimicking as a stone

An eighteen year old female presented with right upper quadrant pain and jaundice of six months duration. Physical examination revealed icterus and hepatomegaly. Bilirubin and alkaline phosphatase were elevated suggesting cholestasis. Ultrasound abdomen showed multiple gall stones with dilated intra-hepatic biliary radicals (IHBR) due to a mixed echogenic lesion in the common hepatic duct (CHD) suspicious of stone. Magnetic resonance cholangiopancreatogram showed dilated IHBR and hyper intense lesion within the CHD. With a provisional diagnosis of CHD stone, ERCP was done with therapeutic intention which revealed a well defined filling defect in the CHD suggestive of a calculus (Fig. 1). Attempts to remove the stone failed and she was taken up for open cholecystectomy and bile duct exploration. At surgery CHD was filled with a solid soft tissue lesion. Choledochotomy was done and a 5 cm oblong soft tissue lesion popped out of the choledochotomy wound. It was attached to the anterior wall of the CHD by a narrow base. The lesion was completely removed and a completion choledochoscopy was

done to confirm the clearance. Pathologist reported it as a mucinous cystadenoma

Cystadenomas of the biliary tract are rare entities that have predilection to affect middle age females.^{1,2} Hepatobiliary cystadenoma could be mistaken for hydatid cyst or choledochal cyst or simple liver cyst. Multiseptated cystic appearance is the classical radiological appearance of hepatobiliary cystadenoma.³ The case presented here is interesting as it was misdiagnosed as a stone. Misinterpretation of the lesion as a stone was due to the well encapsulated and pedunculated nature of the lesion that occupied the common hepatic duct with extension into the left ductal system. Pedunculated biliary cystadenoma has not been reported in the literature.

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Fig. 1 ERCP- an oblong filling defect in the common hepatic duct resembling a stone

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