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Case report

Early reversal of portal biliopathy in a young girl with extrahepatic portal venous obstruction

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ABSTRACT

Abnormalities of the walls of the biliary tree with occasional obstruction in patients with mainly extrahepatic portal hypertension leads to an entity defined as portal biliopathy. These patients are generally young, asymptomatic and predominantly from Asian countries. Symptomatic patients with portal biliopathy may present with pain, jaundice and upper gastrointestinal (GI) bleeding. They are difficult to manage and may require both diagnostic and therapeutic endoscopic procedures followed, if necessary, by a lienorenal shunt. After a shunt, the portal biliopathy usually regresses in most patients, but in some, especially those who have undergone endoscopic intervention, the obstruction remains. The reversal of portal biliopathy after portal decompression has been described usually six months after surgery. We report a reversal of portal biliopathy after a splenorenal shunt as early as eight days after the procedure.

1. Introduction

Portal biliopathy is defined as abnormalities (wall indentations or obstruction) in the extrahepatic biliary system including the cystic duct and gall bladder in patients with portal hypertension. This is usually due to cavernomatous transformation of the portal vein, also known as extrahepatic portal venous obstruction (EHPVO).

Portal biliopathy is usually asymptomatic and found on magnetic resonance or endoscopic biliary studies in patients with EHPVO, but it also may present with symptoms of bile duct obstruction. Its treatment is often difficult because the ducts are inaccessible being surrounded by large high-pressure venous collaterals which make a bilioenteric anastomosis difficult. Therefore, the recommended strategy is to perform a portal venous decompression operation via a splenorenal shunt to lower the pressure in the distended veins in the bile duct and relieve the obstruction.[1] However, many of these patients have usually undergone repeated biliary stenting and even after an adequate decompressive procedure, the portal biliopathy may not be reversed because of a permanent bile duct fibrous stricture which is due to repeated infection or ischaemia after stenting.[2] Reports of the successful reversal of biliopathy after portal decompression have been at least 6 months after the procedure.[3]

We report a very early reversal of portal biliopathy after a lienorenal shunt after only 8 days.

2. Case report

A 21-year-old female from Himachal Pradesh presented with episodes of massive haematemesis to the Postgraduate Institute of Medical Education and Research in Chandigarh 15 years ago when she was 6 years old. Investigations carried out then revealed that she had EHPVO, splenomegaly and large bleeding oesophageal varices, and she underwent endoscopic variceal ligation on numerous occasions. She remained well, but for the last 6 weeks, she developed jaundice and left hypochondrial pain and recurrent melaena.

On abdominal examination, it was found that she had jaundice and an enlarged spleen which was approximately 8 cm below her left subcostal margin. Investigations revealed a haemoglobin (Hb) level of 13.7G, total leucocyte count (TLC) of 6000, platelets of 57000 and a raised bilirubin level of 6.3 mg. Upper gastrointestinal (UGI) endoscopy revealed small oesophageal varices. Ultrasound examination showed a contracted gall bladder which contained a 15-mm-sized calculus. The spleen was grossly enlarged and measured 19.7 cm in diameter. There were large collaterals

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around the bile ducts, in the peripancreatic region and splenic hilum (Fig. 1). Magnetic resonance cholangiopancreatography (MRCP) (Figs. 2 and 3) was suggestive of EHPVO, splenomegaly, portal biliopathy with indentations of the common bile duct and cholelithiasis.

3. MRCP (preoperative)

3.1. Management

3.1.1. Surgery

The patient underwent a splenectomy and a lienorenal shunt (Fig. 4) with a partial cholecystectomy and gallstone removal (using Pribram's technique[4]) and made an uneventful recovery. Postoperatively on day 8, her Hb was 14.2, TLC 9350 and platelet 399000 and the bilirubin had fallen to 1.78 mg/dl. Doppler USG showed that the lienorenal shunt was patent with a peak systolic velocity of 30 cm/sec. A repeat MRCP examination (Fig. 5) which was conducted early because she lived far from adequate medical help showed that the changes of portal biliopathy had regressed (Figs. 6 and 7). On follow-up after 5 months, shunt was patent with normal liver function tests (LFT).

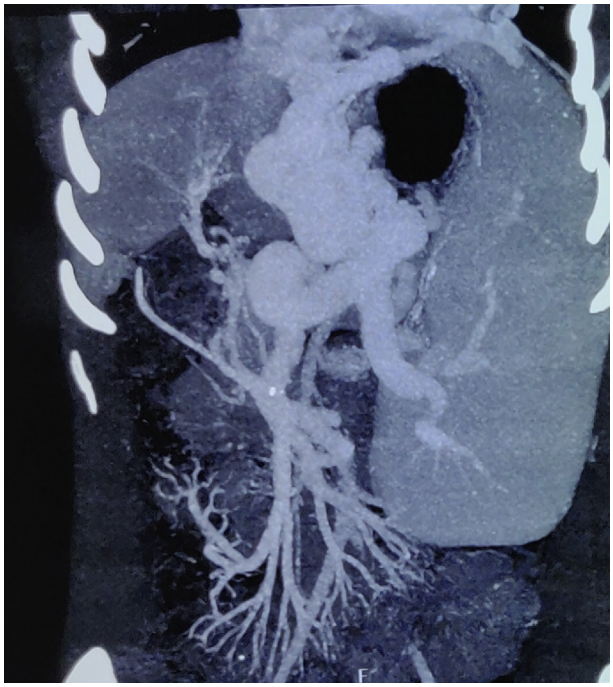

Fig. 1. Contrast enhanced computerised tomography (CECT) showing extensive collateral veins in the abdomen.

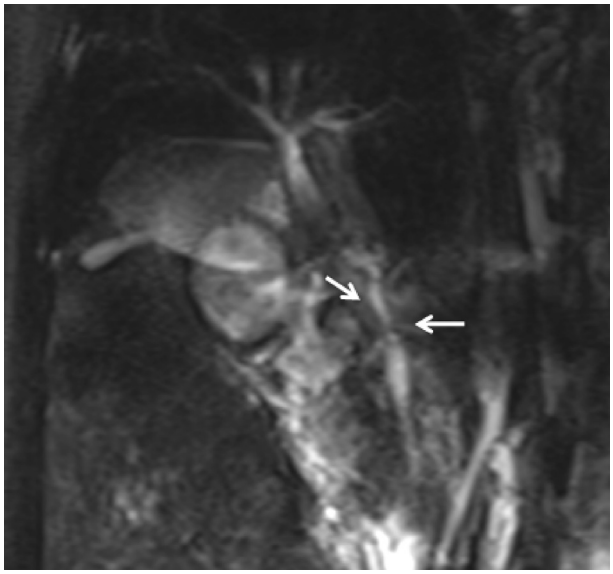

Fig. 2. Magnetic resonance cholangiopancreatography (MRCP) image showing chronic compression of common bile duct (CBD) with marked irregularity of the duct.

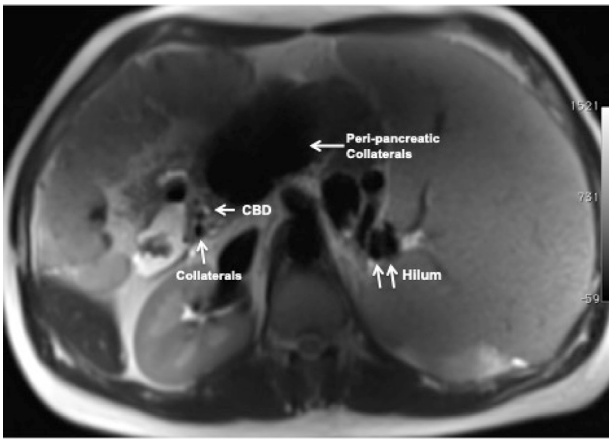

Fig. 3. T2W axial image showing multiple pericholedochal collaterals compressing the common bile duct (CBD). Multiple collaterals in the retropancreatic region and splenic hilum.

4. Discussion

EHPVO is a common cause of major upper GI bleeding among children in developing countries where it constitutes up to 40% of all patients with portal hypertension.[5,6]

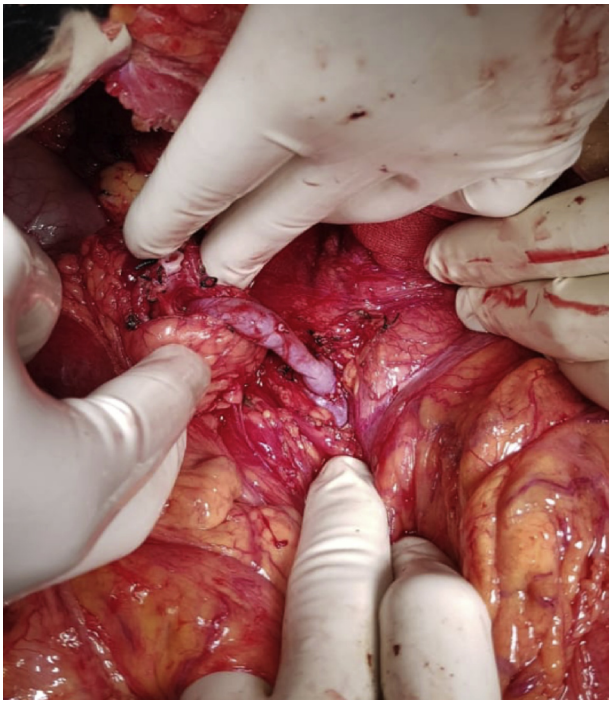

Fig. 4. Proximal splenorenal shunt.

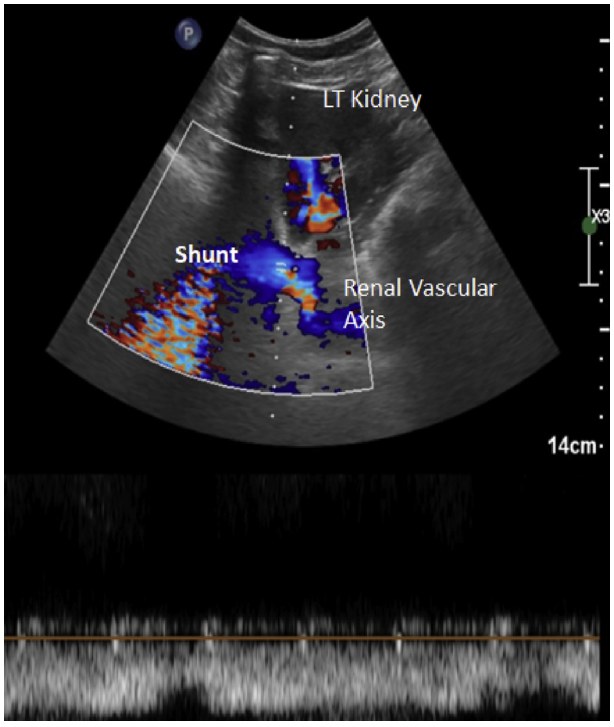

Fig. 5. Doppler ultrasound upper abdomen.

Some patients with EHPVO also develop biliary obstruction due to distended veins in the bile duct walls, a condition known as portal biliopathy. Most of these patients initially undergo repeated endoscopic biliary stenting procedures. However, the stents often become blocked in about 3 months and need repeated changes. Their presence may also be associated with recurrent cholangitis and the development of permanent biliary strictures due to infection or ischaemia.[2]

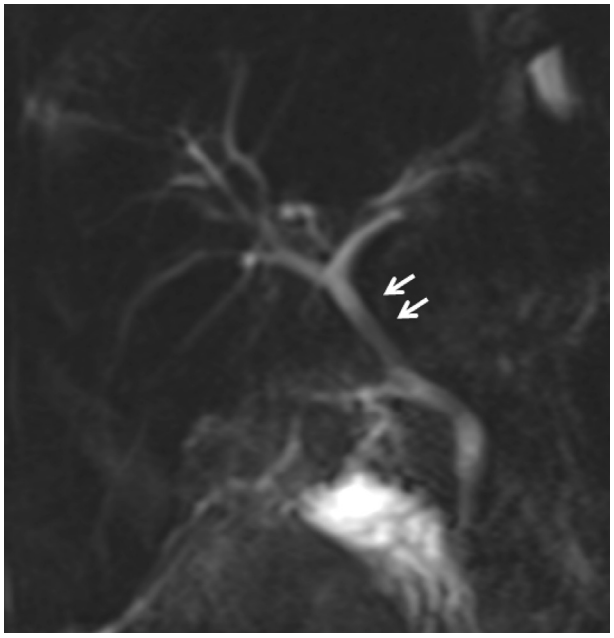

Fig. 6. Postoperative magnetic resonance cholangiopancreatography (MRCP): regression of the indentations of the common bile duct.

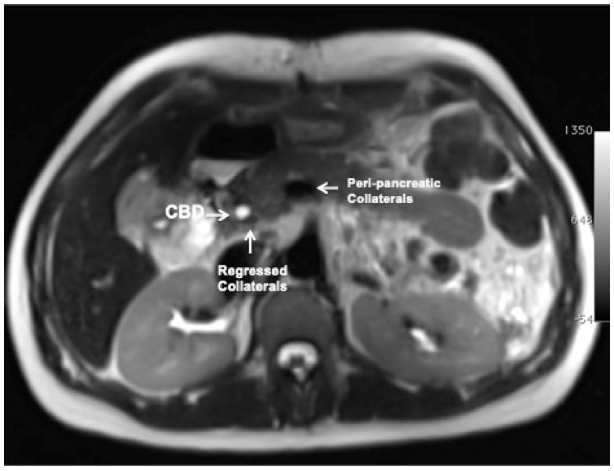

Fig. 7. T2W axial image: regression of the pericholedochal and the peripancreatic collaterals.

The surgical options include portosystemic shunts whose patency rates, if performed by experts, is reported to be from 84 to 96% even after anastomosing veins of down to 4 mm in diameter. If the shunt is patent at the end of the operation, it rarely becomes blocked later.

We first described the reversal of the changes of portal biliopathy after a decompressive procedure,[7] and we have also described that in some patients, the obstruction is not reversed even after a shunt which is patent.[2] We attributed this to the development of permanent strictures which resulted after repeated stenting. The treatment of portal biliopathy is, we surmise, likely to be more effective in a patient who has not undergone previous stent insertion, and this is, to our knowledge, the first report of the early and successful reversal of the changes only 8 days after a successful decompressive procedure.

Conflict of interest

None.

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