Clinical Medicine Insights: Case Reports

CASE REPORT

An Unusual Cause of Biliary Obstruction

Sern Wei Yeoh

Austin Hospital, Liver Transplant Unit, 145 Studley Road, Heidelberg, Victoria, Australia.
Corresponding author email: sernwei.yeoh@austin.org.au

Abstract: Portal biliary ductopathy (PBD) is a condition in which biliary and pancreatic ducts are extrinsically compressed by collateral branches of the portal venous system, which in turn have become dilated and varicosed due to portal hypertension. While the majority of patients with PBD are asymptomatic, a minority can present with symptoms of biliary obstruction and cholangitis with the potential of developing secondary chronic liver disease. This paper reports the case of a 29 year old male presenting with acute cholangitis, in whom PBD was diagnosed radiologically. A brief review of current literature regarding the diagnosis and management of this condition will also be presented.

Keywords: portal hypertension, biliary obstruction, cholangitis, MRCP, biliary stenting

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Case Report

A 29 year old male presented to the Emergency Department with a one week history of painful jaundice, nausea, lethargy, fevers and rigors. He had latent tuberculosis but had otherwise been well and did not consume alcohol. Upon examination he was febrile, icteric and tender in the right upper quadrant with a temperature of 39 degrees Celsius. Liver function tests were abnormal as follows: bilirubin 212 μmol/L (RR 2–14 μmol/L), alkaline phosphatase 242 U/L (RR 40–130 U/L), alanine transaminase 75 U/L (RR 8–40 U/L), gamma glutamyl transferase 295 U/L (RR 0–50 U/L). International Normalised Ratio was 1.4. Full blood examination demonstrated anaemia with a haemoglobin of 119 g/L (RR 130–180 g/L) and thrombocytopenia with platelets 109 × 10^9/L (RR 150–400 × 10^9/L). Electrolytes were normal and serological tests for viral hepatitides, human immunodeficiency virus, schistosomiasis, strongyloides, malaria and fasciola were negative. An abdominal ultrasound showed gross biliary duct dilatation with sludge in the gallbladder. The portal vein was mildly dilated at 1.4 cm diameter but flow within the lumen was anterograde with no sign of portal venous obstruction or thrombosis.

A subsequent contrast-enhanced computed tomography (CT) scan of the abdomen showed dilatation of the portal vein in the region of the bifurcation, with collateral portal varices causing compression of the proximal common bile duct (CBD) (Fig. 1). A magnetic resonance cholangio-pancreatogram (MRCP) confirmed these findings and demonstrated intrahepatic bile duct dilatation with a beaded appearance, especially in the left lobe (Figs. 2 and 3). Splenic vein dilatation was also noted with concomitant splenomegaly, and the sludge previously visualised in the gallbladder was seen to extend into the CBD. A liver biopsy showed non specific changes consistent with biliary obstruction, periportal fibrosis and no cirrhosis.

This patient had cholangitis secondary to biliary tract obstruction. While biliary sludge likely contributed to the obstruction, this patient also exhibited radiological evidence of portal hypertension resulting in varicosed portal venous collaterals compressing adjacent intrahepatic and extrahepatic bile ducts—a rare phenomenon termed 'portal biliary ductopathy' (PBD). The cause of portal hypertension in this patient is believed to be idiopathic portal hypertension. He has undergone multiple placements of biliary stents via endoscopic retrograde cholangio-pancreatography (ERCP) since his initial presentation complicated by recurrent structuring, cholangitis and ERCP-related pancreatitis. Furthermore, as a result of portal hypertension, he has developed oesophageal varices necessitating endoscopic banding and internal haemorrhoids causing intermittent per-rectal bleeding. He is currently planned for insertion of a portosystemic shunt.

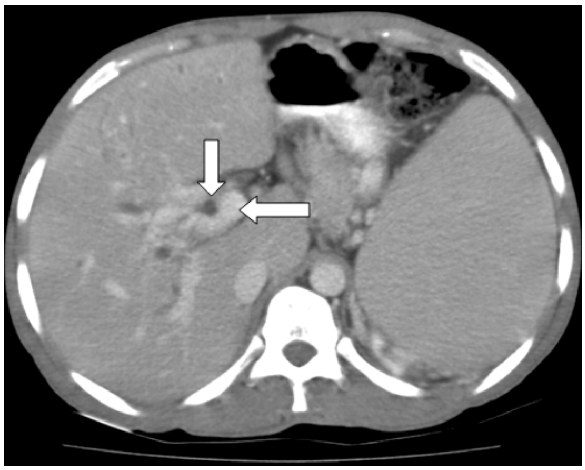

Figure 1. An axial CT image demonstrating the common bile duct.
Notes: Vertical arrow: diameter 3.9 mm, flanked by varicosed portal venous collaterals; Horizontal arrow: diameter 10.1 mm.

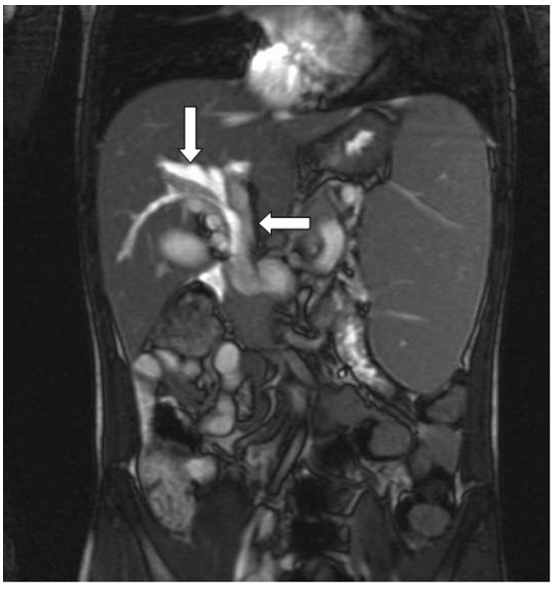

Figure 2. A coronal MRCP image demonstrating the dilated intrahepatic biliary tree.
Notes: Vertical arrow: diameter of duct indicated 10 mm, with varicosed portal collaterals surrounding the common hepatic duct; Horizontal arrow: diameter 7.3 mm.

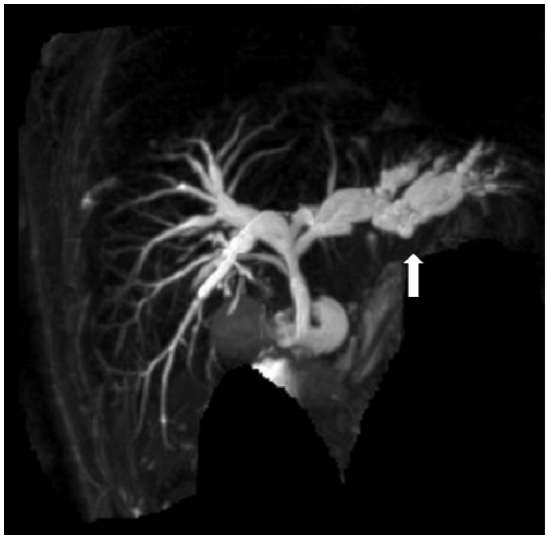

Figure 3. An MRCP image demonstrating extensive dilatation and structuring of the left intrahepatic biliary tree.
Notes: Vertical arrow: diameter of duct indicated 15 mm, in preference to the right.

Discussion

The hepatic biliary system comprises the left hepatic and right hepatic ducts which drain segments 2–4 of the left hemiliver and segments 5–8 of the right hemiliver respectively. These ducts exit the liver at the hilum and abut the left and right branches of the portal vein respectively, before joining at a confluence along the right branch of the portal vein to become the common hepatic duct. This in turn runs along the anterior border of the portal vein before exiting into the duodenum. As such, the biliary system is closely apposed to the portal venous system for much of its course.[1]

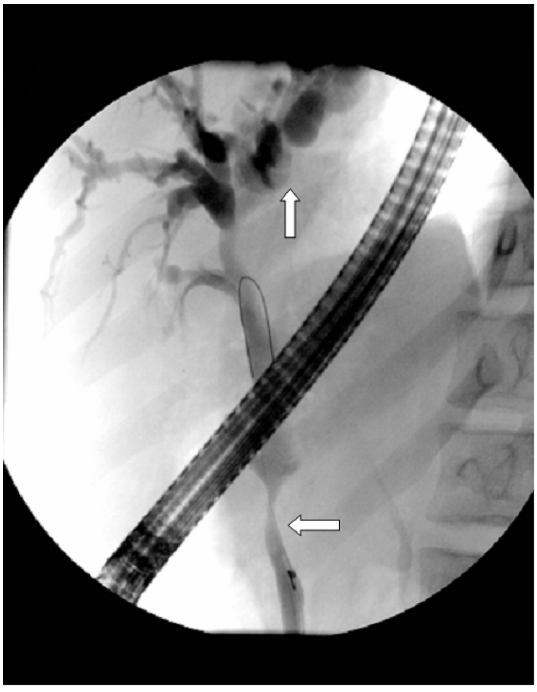

Figure 4. A fluoroscopy image taken during ERCP demonstrating smooth proximal bile duct compression.
Notes: Horizontal arrow: diameter at area indicated 2.8 mm, and secondary distal bile duct dilatation; Vertical arrow: diameter at area indicated 15.4 mm.

Balfour and Stewart[2] first described in 1869 'cavernous transformation' of the portal vein whereby collateral venous routes became, due to underlying portal hypertension, recanalised and varicosed. Hunt[3] in 1965 was the first to describe a case of adjacent bile duct compression caused by these varices. This phenomenon has had varied nomenclature including 'pseudosclerosing cholangitis', 'portal biliopathy' and 'portal hypertensive biliopathy'. Bayraktar, in a 2011 review, proposes the term 'portal biliary ductopathy' to more accurately describe the morphological changes arising from this condition.[4] Histological changes including neogenesis, ischaemic necrosis of duct walls and development of periportal fibrotic connective tissue have been purported to consolidate and worsen bile duct compression.[5–6]

The underlying portal hypertension in PBD is usually secondary to portal vein thrombosis which in up to 50% of patients is idiopathic.[7] Identifiable causes of thrombosis include omphalitis or intra-abdominal sepsis in children; and tumour invasion, chronic pancreatitis or myeloproliferative disorders in adults.[7] PBD can also occur, as in this case report, due to idiopathic portal hypertension,[8] otherwise known as noncirrhotic portal fibrosis.[9] A detailed review of this condition is beyond the scope of this report; however it is defined as "a clinical disorder of unknown aetiology characterized by splenomegaly, anaemia, and portal hypertension in the absence of cirrhosis, blood disease, parasites in the hepatobiliary system, and occlusion of the hepatic and portal veins".[8] Histological features of this condition are heterogenous but include sclerotic and fibrotic periportal changes. Schistosomiasis is another significant cause of portal hypertension worldwide which should be excluded in patients with PBD.

The aforementioned aetiologies tend to cause extrahepatic cavernous transformation of the portal vein and thus compression of extrahepatic biliary ducts, which in turn causes retrograde dilatation of the intrahepatic biliary system. However radiological

case series have demonstrated that varices course into the liver parenchyma and, where they abut intrahepatic bile ducts, can cause intermittent duct compression.[10] The left hepatic duct is more frequently involved than the right for unclear reasons and the combination of intrahepatic duct dilatation and intermittent compression causes a beaded appearance on contrast-enhanced imaging. Both of the phenomena are illustrated in this case report. Portal hypertension due to intrahepatic cirrhosis rarely causes PBD and if so tends to cause purely intrahepatic ductal compression.[11]

The majority of patients with radiological evidence of PBD are asymptomatic and may have normal liver function tests. Jaundice, pain and infective cholangitis occur in 5%–40% of patients.[12] Accumulation of bile duct sludge or stones within constricted ducts can precipitate and worsen obstruction, as in our patient.[13] Prolonged biliary obstruction may lead to secondary biliary cirrhosis and, in the long term, derangement in liver function.[14]

Magnetic resonance cholangio-pancreatography (MRCP) is the recommended first-line investigation in diagnosing PBD as it is non-invasive, demonstrates both intrahepatic and extrahepatic biliary changes and can concurrently evaluate the biliary and portal systems.[15] However endoscopic ultrasonography with colour Doppler may be used complementarily to survey structures around the bile duct, delineating peri-ductal varices and ruling out concurrent pathology such as tumours.[16]

In patients with symptomatic PBD, therapeutic insertion of stents or biliary dilatation via ERCP may be considered. These procedures risk causing haemobilia by damaging varices that have herniated into the bile duct lumen.[16] Despite this risk, endoscopic procedures remain a common, first line strategy to manage biliary obstruction.[17] Failure of endoscopic therapy and/or recurrence of stricturing may necessitate surgical decompression of the portal hypertensive varices through creation of a porto-systemic shunt. Should this not relieve biliary obstruction (due to established ischaemic or fibrotic duct changes), biliary bypass surgery via hepatico-jejunostomy may be indicated.[18] It is critical that portal venous decompression occurs before biliary bypass surgery due to the high risk of intraoperative bleeding from hypertensive portal vessels- indeed, dilated veins are difficult to distinguish intraoperatively from bile ducts.[19] A definitive procedure for PBD is liver transplantation, which should be reserved for patients who develop secondary biliary cirrhosis.[20]

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Author Contributions

Wrote the first draft of the manuscript: SWY. Contributed to the writing of the manuscript: SWY. Agree with manuscript results and conclusions: SWY. Jointly developed the structure and arguments for the paper: SWY. Made critical revisions and approved final version: SWY. All authors reviewed and approved of the final manuscript.

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Conflicts of Interest

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