Case Report

Portal Biliopathy


Abstract
A young male with portal venous thrombosis presented with obstructive jaundice, due to common bile duct stricture secondary to portal biliopathy.

INTRODUCTION
Jaundice in portal hypertension is uncommon. We report a case of portal hypertension presenting with obstructive jaundice secondary to portal biliopathy.

CASE REPORT
A 24 years male, diagnosed as portal venous thrombosis in 1993, presented with cholestatic type of jaundice for 2 weeks in April 2004. Patient was deeply icteric with scratch marks; he had no pedal edema. There was a mild hepatomegaly and moderate splenomegaly. Gallbladder was not palpable. There was no ascites. In 2001 he was diagnosed to have gall stones and stones in the common bile duct (CBD). He had endoscopic removal of stones. Patient was asymptomatic until the present episode.

Investigations revealed a normal hemogram and peripheral smear, S. Bilirubin total: 24 mg/dL, direct: 18 mg/dL, ALT 80 IU/L and AST: 72 IU/L, S. Alk phos 470 IU / L. Viral markers for A, B, C, and E were negative. Ultrasound and Doppler confirmed cavernomatous transformation of portal vein and stones within the gallbladder and CBD. Magnetic Resonance Imaging (Fig.), showed multiple stones within the CBD and the gallbladder (Fig. 1). Bile duct stricture was noted (Fig. 2). The portal and splenic veins showed cavernomatous transformation and seen to encircle the CBD. Extensive collateral veins were seen in the retroperitoneum and upper abdomen (Fig. 3). The splenic parenchyma showed Gamma Gandy bodies (Fig. 4). An initial nasobiliary drain followed by double stent deployment was done for the patient. There was a significant fall in the serum bilirubin level (3 mg/dL) at the end of two weeks. He is currently asymptomatic.

DISCUSSION
Portal biliopathy is referable to biliary ductal changes in patients with portal hypertension. It is common in...
portal vein thrombosis and less often in cirrhosis and noncirrhotic portal fibrosis. The changes include stricture and dilatation of both extra-and intra-hepatic bile ducts secondary to varices encircling the CBD and gallbladder wall. When these changes become significant the individual manifests with obstructive jaundice. Choledocholithiasis is a common sequel. Sarin et al observed these changes in 80% of patients with EHPVO. The precise explanation of higher frequency of portal biliopathy in EHPVO is not clear. The biliary stricture is either secondary to ischemia, or due to a prolonged compression of the biliary tree by the portal cavernoma. Dilawari and Chawla had observed biliary changes in all the 20 patients and labeled the radiological abnormalities as pseudosclerosing cholangitis.

Cholangiography is useful in the diagnosis and classifying the type of portal biliopathy. Prominent changes include: irregularity and strictures of CBD and hepatic ducts which may be smooth and tapering, strictures may be single or multiple, short or long. Localized saccular dilatations with filling defects are suggestive of CBD calculi. The findings simulate sclerosing cholangitis. Based on the location and extent of cholangiographic abnormalities, a simple classification of portal biliopathy has been proposed.

Type I: Involvement of extrahepatic bile duct.
Type II: Involvement of intrahepatic bile ducts only.
Type IIIa: Involvement of extrahepatic bile duct and unilateral intrahepatic bile duct (Left or Right)
Type IIIb: Involvement of extrahepatic bile duct and bilateral intrahepatic bile duct.

The clinical features depend on the location and extent of bile duct obstruction. When partial, patients are asymptomatic; when complete, patients present with features of ascending cholangitis. Prolonged obstruction often leads to the development of secondary biliary cirrhosis. Serum alkaline phosphatase level is often elevated.

Ultrasound with Doppler is a useful modality to diagnose EHPVO and portal biliopathy. Color Doppler flow imaging can differentiate the periportal collaterals from the dilated bile ducts. Gall bladder varices are seen as colour-filled round-to-serpiginous channels within the gall bladder wall. MRC has a definite role in portal biliopathy. ERCP has an additional therapeutic role of CBD stone removal and stent placement. The latter is necessary to maintain lumen patency. Dilatation of the stricture segment-using balloon may be necessary in specific situations.

The differential diagnoses include CBD calculi with stricture formation, biliary ascariasis in the tropics and primary sclerosing cholangitis.

In patients with symptomatic biliary obstruction not amenable to endoscopic therapy, a portosystemic shunt is indicated, to decompress the portal system. While regression of ectopic varices has been noted in most, a proportion of patients continue to have bile duct changes. Hepaticojejunostomy may be beneficial in these non-responders.

REFERENCES