**Case Report**

**Bile duct wall calcification: A rare entity in portal cavernoma cholangiopathy**

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**ABSTRACT**

Cavernomatous transformation of the portal vein, seen in extrahepatic portal venous obstruction (EHPVO), can cause impingement or ischemic insult on bile ducts manifesting as “portal cavernoma cholangiopathy” (PCC). Bile duct wall calcification in portal biliopathy is a rare occurrence and has not been reported in the literature to the best of our knowledge. We report a 59-year-old male, a known case of EHPVO, who had undergone laparoscopic cholecystectomy, splenectomy, and splenorenal shunt in the past. The patient had now presented to us in view of recurrent episodes of cholangitis for which a biliointestinal bypass was planned. Intraoperatively, dilated and densely thickened bile ducts with multiple pericholedochal collaterals were noted. Incision of common hepatic duct and left hepatic duct showed completely calcified ductal wall with no visible healthy mucosa. Calcifications were removed partially from the bile duct walls near choledochotomy site. With the anticipation of futile benefit from biliointestinal bypass, Roux-en-Y HJ was abandoned. Hepaticoduodenostomy was done to prevent bile leak from choledochotomy site.

**KEY WORDS:** Bile duct calcification, EHPVO, portal cavernoma cholangiopathy

**Introduction**

In patients with extrahepatic portal vein obstruction (EHPVO), cavernomatous transformation of the portal vein can cause impingement or ischemic insult on bile ducts manifesting as “portal cavernoma cholangiopathy” (PCC).

Calcification of portal vein and mesenteric vein are known lesions reported to be associated with EHPVO. Bile duct wall calcification in portal biliopathy is a rare occurrence and has not been reported in the literature to the best of our knowledge.

**Case Summary**

A 59-year-old male presented with symptoms of jaundice with recurrent right upper abdominal pain and fever. Patient is a known case of EHPVO diagnosed at the age of 24 years. He underwent laparoscopic cholecystectomy and multiple endoscopic cholangiography stent exchanges for recurrent cholangitis at the age of 43 years. Two years later, patient underwent splenectomy and proximal splenorenal shunt. He was asymptomatic for next 10 years. Now he presented with recurrent episodes of cholangitis probably due to stent blockade.

On examination, he had jaundice. Laboratory studies revealed white blood cells 11,420 cells/mm³, hemoglobin 11.7 g/dL, platelets 253,000/mL, aspartate aminotransferase (AST) 73 IU/L, alanine aminotransferase (ALT) 26 IU/L, total bilirubin 2.2 mg/dL, alkaline phosphatase 332 U/L, and international normalized ratio 1.19. Abdominal imaging showed contrast-enhanced computed tomography (CECT) showed bilateral moderate-to-severe intrahepatic biliary radical dilation (IHBRD) with multiple large dense calculi, predominantly in the right lobe of the liver. Atrophy of the right lobe of the liver with hypertrophy of left and caudate lobes was pronounced.

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Cavernomatous transformation of the portal vein was noted. Grossly dilated common bile duct with circumferential wall calcification and stones were identified [Figure 1]. Endoscopy disclosed scarring in the lower esophagus due to the previous endoscopic variceal ligation (EVL) with no evidence of any varices. Mild portal hypertensive gastropathy changes were noted.

In view of recurrent episodes of cholangitis requiring multiple stent exchanges, patient was initially planned for Roux-en-Y hepaticojejunostomy (HJ). Intraoperatively, multiple perihilar and pericholedochal collaterals with dilated and densely thickened extrahepatic bile ducts were noted. Choledochotomy of common and left hepatic duct showed completely calcified ductal wall with no visible healthy mucosa [Figure 2]. No active bile output was noted from intrahepatic ducts. A plastic stent was noted in situ. Calcifications were removed partially from the bile duct walls near choledochotomy site. With the anticipation of futile benefit from biliointestinal bypass, Roux-en-Y HJ was abandoned. Hepaticoduodenostomy was done to prevent bile leak from choledochotomy site and to provide further endoscopic access to biliary system for calculi extraction. Postoperatively, patient had an uneventful course except for mild cholangitis episodes that were managed conservatively. In the follow-up, it was planned to assess response to biliary decompression and to perform endoscopic biliary calculus retrieval following CECT triphasic imaging at 3 months. After stone retrieval, patient will be kept under three monthly follow-up and managed accordingly.

**Discussion**

PCC is defined as abnormalities in the extrahepatic biliary system including the cystic duct and gallbladder with or without abnormalities in the first and second generation biliary ducts in a patient with portal cavernoma. PCC is found to be seen in the form of biliary changes in nearly 81%–100% patients of EHPVO. However, only 5%–38% of them are symptomatic. Patients with PCC develop symptoms after longer duration of EHPVO and at a higher age like the patient in the present report. The pathogenesis of PCC has been debated. Dilated venous collaterals causing extrinsic compression over the biliary ductal system constitutes the reversible component of PCC. Most of the biliary changes are expected to be resolved following successful shunt surgery. However, not all biliary findings are reversed following shunt surgery. This nonreversibility of biliary changes can possibly be explained by the concept of fixed component of PCC. The fixed component of PCC occurs most likely due to ischemic changes in the bile duct or by presence of fibrous tumor-like cavernoma causing persistent compression on the bile ducts. In the present report, the persistence of symptomatic PCC even after shunt surgery will most probably be explained by the fixed component of PCC. In the present patient, hepaticoduodenostomy was performed to aid in the endoscopic access for further biliary stone retrieval in addition to biliary drainage.

Bhatia reviewed several studies for cholangiography findings in PCC and proposed nomenclature for the findings as extrinsic impressions, shallow impressions, irregular ductal contour, stenosis, upstream dilations, filling defects, bile duct angulation, and ectasia. The finding of circumferential biliary duct wall calcification in PCC is a rare entity highlighted in the present report. According to stages in the natural history of PCC by Kumar and Saraswat, patient in the present report can be staged under complicated stage with advanced cholangiopathy features. Bile duct wall calcification is expected to be due to dystrophic calcifications in the bile duct wall as a consequence of ischemic vascular changes in PCC.

Presence of bile duct wall calcification is associated with the prolonged course and advanced stage of portal cavernomatous cholangiopathy. Prospectively assessing patients further throughout the course of PCC will provide more insight into this finding of bile duct calcification, thus contributing insights into the pathogenesis of PCC.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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**Figure 1:** Computed tomography image showing circumferential bile duct calcification

**Figure 2:** Intraoperative picture showing choledochotomy site (black arrow) with calcified wall of common hepatic duct
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Conflicts of interest
There are no conflicts of interest.

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