

Biliary cast syndrome in portal hypertensive biliopathy: direct cholangioscopic findings and endoscopic therapy with metal stent

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Abbreviations

CT	computed tomography
HCV	Hepatitis C virus
EHPVO	extrahepatic portal vein obstruction
ERCP	endoscopic retrograde cholangiopancreatography
HCC	hepatocellular carcinoma.

A 61-year-old man with asthma, diabetes mellitus, cirrhosis secondary to chronic hepatitis C virus (HCV) infection, and hepatocellular carcinoma (HCC), was admitted to our hospital because of progressive abdominal pain and jaundice of 7 days duration. On clinical examination, the patient had abdominal distention, tenderness in the right upper quadrant, and jaundice. His hemoglobin was 8.7 mg/dL; hematocrit, 25%; white blood cells, $1.7 \times 10^9/L$; international normalized ratio (INR), 1.4; total bilirubin, 15 mg/dL; direct bilirubin, 7.6 mg/dL; indirect bilirubin, 7.4 mg/dL; and alkaline phosphatase, 65 IU/L. The remaining laboratory parameters were within reference ranges.

A computed tomography scan (CT) revealed cirrhosis, HCC, portal vein thrombosis and cholelithiasis with intrahepatic filling defects and ductal dilation (Fig. 1). During endoscopic retrograde cholangiopancreatography (ERCP) marked portal hypertensive gastropathy and duodenopathy was evident. ERCP showed irregular filling defects within the entire common bile duct (Fig. 2). During standard cannulation the wire would not advance into the bile duct because of the presence of the large impacted filling defect. Therefore, a biliary precut-fistulotomy was performed with subsequent extraction of a worm-shaped structure (“biliary cast”) (Fig. 3). Upon removal of the biliary cast, hemobilia was observed, which was successfully treated by inserting a fully covered self-expanding metal stent (fcSEMS; Wallstent 10 mm diameter \times 80 mm

length, Boston Scientific, Massachusetts, United States) (Fig. 4). The length of the stent was chosen based on the diffuse characteristics of the portal biliopathy.

Two-days later, a direct cholangioscopy (DC) using Spyglass (Boston Scientific, Massachusetts, United States) for biliary cleansing was performed. During the DC, the inner mucosal lining of the bile duct was observed, demonstrating a typical snake-skin mucosal pattern consistent with portal hypertensive biliopathy (Fig. 5, Video 1). The histology showed a biliary cast (Fig. 3c). The clinical condition stabilized and the patient was discharged home one-week later. The stent was removed uneventfully four-weeks later. At the four-month follow-up, he did not have any further bleeding.

Discussion

Extrahepatic portal vein obstruction (EHPVO) is a common cause of portal hypertension and occurs in up to 40% of all patients with portal hypertension, but it is not confined to those patients [1]. Similarly, EHPVO can occur in liver cirrhosis. A rare manifestation of portal hypertension is hemobilia, which is believed to be caused by rupture of abnormally dilated veins in and around the bile duct wall that results from elevated portal venous pressure [1]. In our patient, the bleeding likely originated from the diffuse mucosal changes of biliopathy leading to a large blood clot, which mixed with bile, cells, and debris, and was trans-

Video

The video demonstrates the typical snake-skin pattern of portal hypertensive biliopathy.

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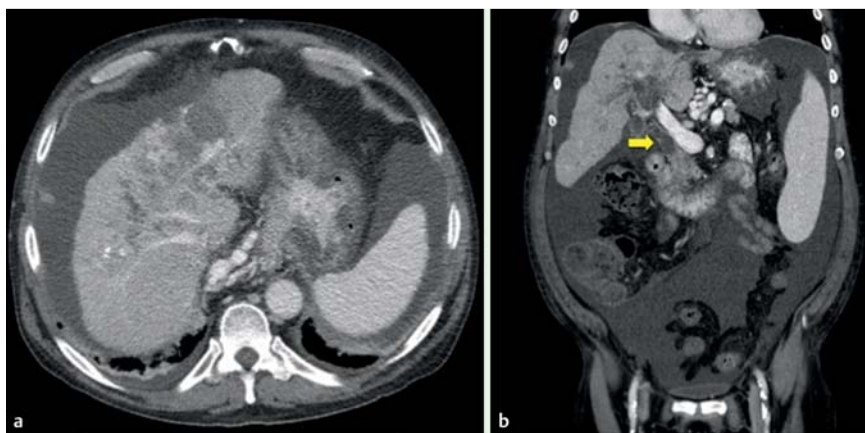


Fig. 1 CT scan. **a** The CT scan revealed cirrhosis, HCC, portal vein thrombosis and choledocholithiasis with intrahepatic filling defects and ductal dilatation. **b** Sagittal CT showing the distal bile duct filled with a biliary cast.

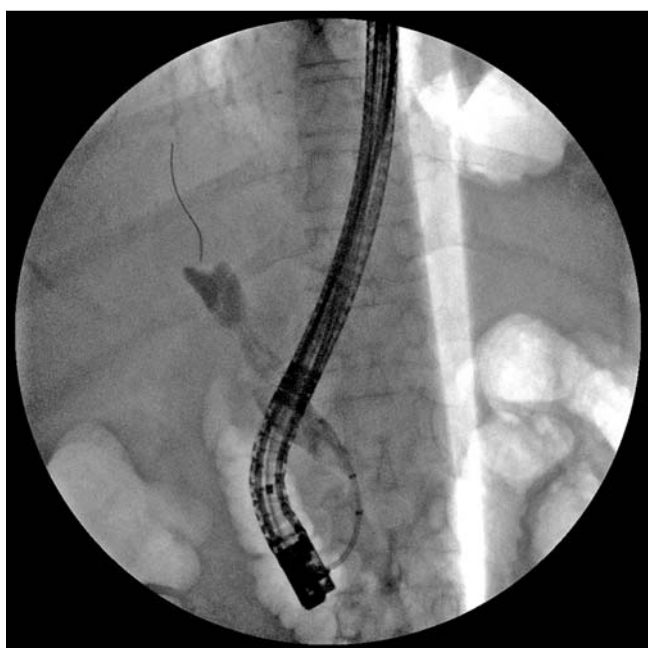


Fig. 2 Cholangiography showed irregular filling defects within the entire common bile duct.

formed into a biliary cast. Because our patient also had HCC, it is possible that tumor invasion into the bile duct led to hemorrhage. However, the rapid response to tamponade with fcSEMS supports the portal biliopathy as the etiologic factor of the hemobilia. Furthermore, hemobilia due to tumor invasion or variceal rupture usually results in a decrease in hemoglobin, which did not occur in our patient. We suspect that the bleeding was slow but steady because it occurred in portal hypertensive gastropathy. The retained blood clots acted as a lithogenic stimulus, leading to the biliary cast [2,3]. Biliary cast syndrome is an unusual condition that has been reported in liver transplantation, post-cholecystectomy, antiphospholipid antibody syndrome, gallbladder hypocontractility, parenteral nutrition, and biliary infection [2–5]. Diagnosis of biliary cast syndrome is established primarily by ERCP or percutaneous transhepatic cholangiography. The endoscopic therapeutic options include sphincterotomy with balloon or basket extraction, and mechanical and electrohydraulic lithotripsy [2–5]. Other modalities include endoscopic nasobiliary drainage, percutaneous transhepatic drainage, and surgery [2–5].

In summary, this case report adds to the literature for three important reasons. First, we provided evidence of the existence of mucosal biliopathy by showing the DC view of this disorder. Second, we showed that this condition can lead to formation of a biliary cast and, finally, the concept of treating bleeding biliopathy with fcSEMS was demonstrated.



Fig. 3 **a** Endoscopic extraction of the biliary cast. **b** Image of the extracted worm-shaped biliary cast. **c** Histology showed a coagulum with a biliary cast.

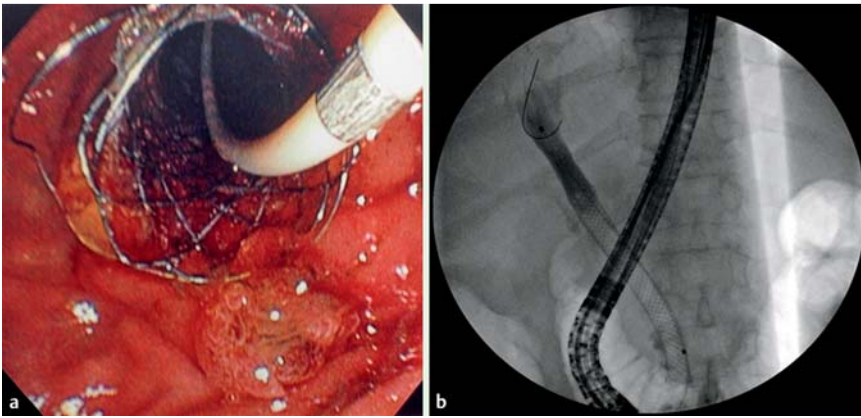


Fig. 4 Upon removal of the cast hemobilia was observed, which was successfully treated by inserting a fully covered self-expanding metal stent.



Fig. 5 Endoscopic appearance of portal hypertensive biliopathy.

Competing interests: None

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