Case Report

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Whole clinical process in a patient with portal hypertensive biliopathy: a case report

Yimin Ma[1], Rencheng Cai[1], Duanming Zhuang[1], Yuehua Tang[1], Youhong Cao[1], Xiaoping Wang[1] and Zhenguo Qiao[2]

Abstract

Portal hypertensive biliopathy is characterized by abnormalities in the biliary tract secondary to portal hypertension, especially extrahepatic portal vein obstruction. Most patients are asymptomatic; only about 20% have clinical symptoms. We herein report a case of portal hypertensive biliopathy caused by cavernous transformation of the portal vein with the development of recurrent cholangitis with common bile duct stones and stricture. This patient underwent endoscopic retrograde cholangiopancreatography, a surgical operation, and a transvenous intrahepatic portosystemic shunt procedure during the whole clinical process. Finally, we found the recurrent plastic stent insertion at endoscopic retrograde cholangiopancreatography was the best option for him at present. In addition, we also discussed the diagnosis and management of this disease.

Keywords

Portal hypertensive biliopathy, endoscopic retrograde cholangiopancreatography, treatment, transvenous intrahepatic portosystemic shunt, stent, case report

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[1]Department of Gastroenterology, Gaochun People's Hospital of Nanjing, Gaochun Branch of Nanjing Drum Tower Hospital, Nanjing, China
[2]Department of Gastroenterology, Suzhou Ninth People's Hospital (Affiliated Wujiang Hospital of Nantong University), Suzhou, China

Corresponding author:
Zhenguo Qiao, Department of Gastroenterology, Suzhou Ninth People's Hospital (Affiliated Wujiang Hospital of Nantong University), 2666 Ludang Road, Suzhou, Jiangsu 215200, China.
Emails: qzg66666666@163.com or qiaozhenguo@ntu.edu.cn

Introduction

Portal hypertensive biliopathy (PHB) is a complication of portal hypertension that often develops secondary to extrahepatic portal vein obstruction (EHPVO).[1] Only about 10% to 15% of patients are symptomatic; more than 80% of patients are asymptomatic but have cholangiographic features.[2] The pathogenesis of PHB is unclear, but one hypothesis is that PHB is caused by ischemic stenosis of the common bile duct (CBD).[3] Another hypothesis is that the high external pressure of the bile duct is caused by a network of dilated choledochal veins or cavernous transformation of the portal vein.[4] Abdominal pain, jaundice, and cholangitis are the main clinical symptoms of PHB.[5] Bleeding of esophageal varices with resultant severe hemobilia is relatively rare.[6] Treatment options for PHB are limited. The patient described in this case report underwent nearly all known treatment methods.

Case report

A 55-year-old man presented with abdominal pain, fever, and a 2-year history of jaundice. He had no history of viral hepatitis, immune system disease, or hematological disease. At the first admission, his serum bilirubin concentration was 177.4 μmol/L (reference range, 1.7–21 μmol/L), direct bilirubin concentration was 150 μmol/L, glutamyl transpeptidase concentration was 1429 U/L (reference range, 10–60 U/L), and serum alkaline phosphatase concentration was 296 U/L (reference range, 45–125 U/L). All other liver function parameters were normal. No obvious esophagogastric varices were found by gastroscopy. A computed tomography (CT) scan showed cavernous transformation of the portal vein and extensive intrahepatic and extrahepatic varices; part of them surrounded the CBD. The lower section of the CBD was compressed, deformed, and narrow, and the upstream section showed segmental dilatation (Figure 1). Magnetic resonance cholangiopancreatography (MRCP) showed that part of the intestine overlapped with the relatively narrow and smooth CBD (Figure 1). Endoscopic retrograde cholangiopancreatography (ERCP) demonstrated a 3-cm-long region of stenosis of the lower segment of the CBD and significant dilation of the upper segment. The stenosis of the bile duct corresponded to the site of the cavernous transformation of the porta hepatis identified on CT and MRCP. ERCP revealed several filling defects in the CBD, and a plastic stent was placed. No obvious tumor cells were found in the stenotic lower segment of the CBD.

Two weeks later, the patient was admitted to Jiangsu Provincial People's Hospital for further treatment. Choledocholithotomy, T-tube drainage, and cholecystectomy were planned for 26 December 2017. During the surgery, however, we found that the cavernous transformation had resulted in circuitous vein balls above the CBD. The choledocholithotomy was stopped, and only the cholecystectomy was performed.

The patient still had recurrent chills and fever after the operation. In June 2018, he developed jaundice again and was transferred to Nanjing Drum Tower Hospital. During this hospital stay, viral hepatitis, immune system diseases, and hematological diseases were ruled out by detection of autoantibodies, anticardiolipin antibodies, prothrombin III, CD55/CD59, homocysteine, viral hepatitis indicators, and other relevant indicators. Additionally, no *JAK2* V617F mutation was detected. Because of the patient's high surgery-related risk, transvenous intrahepatic portosystemic shunt (TIPS) was considered the best treatment option. TIPS was performed on 4 July 2018. During the operation, four attempted

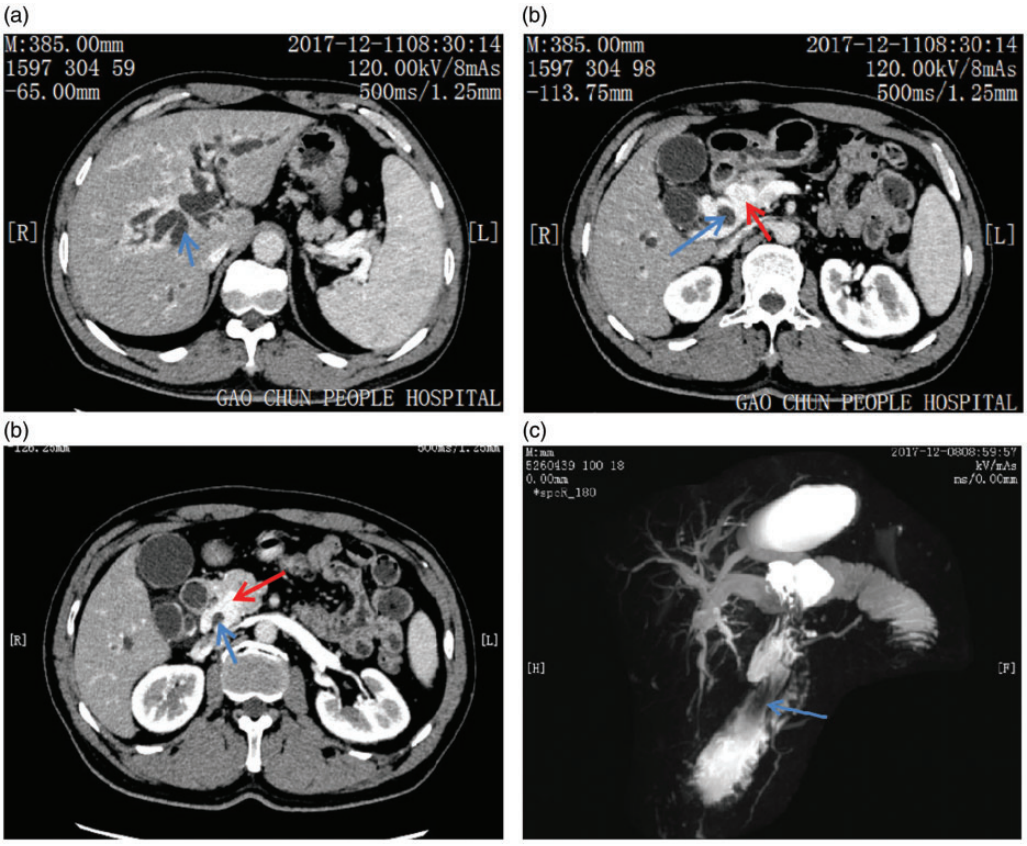

Figure 1. Computed tomography and magnetic resonance cholangiopancreatography images at the patient's first admission. Computed tomography showed significant dilation of (a, b) the intrahepatic bile duct and common bile duct (blue arrows) and (b, c) the collateral pathway of the portal vein (red arrows). (b, c) Additionally, the lower section of the common bile duct was compressed, deformed, and narrow with segmental upstream dilatation (blue arrows). (d) Magnetic resonance cholangiopancreatography showed that part of the intestine overlapped with the relatively narrow and smooth common bile duct (blue arrow).

jugular vein needle punctures failed to enter the main portal vein, and the operation was finally abandoned.

The patient subsequently underwent three ERCP procedures because of symptom recurrence. CT and magnetic resonance imaging + MRCP were counterchecked prior to the most recent ERCP (Figure 2 (a)–(c)), and the stent was replaced during the procedure (Figure 2(d)). At discharge, the patient's serum bilirubin and alkaline phosphatase concentrations were normal and his chills and fever had disappeared.

Ethical approval

This research complies with all relevant national regulations and institutional policies and was performed in accordance with the tenets of the Helsinki Declaration. The study was approved by the authors' institutional review board or equivalent committee.

Informed consent

The patient provided written informed consent for publication of the data in this study.

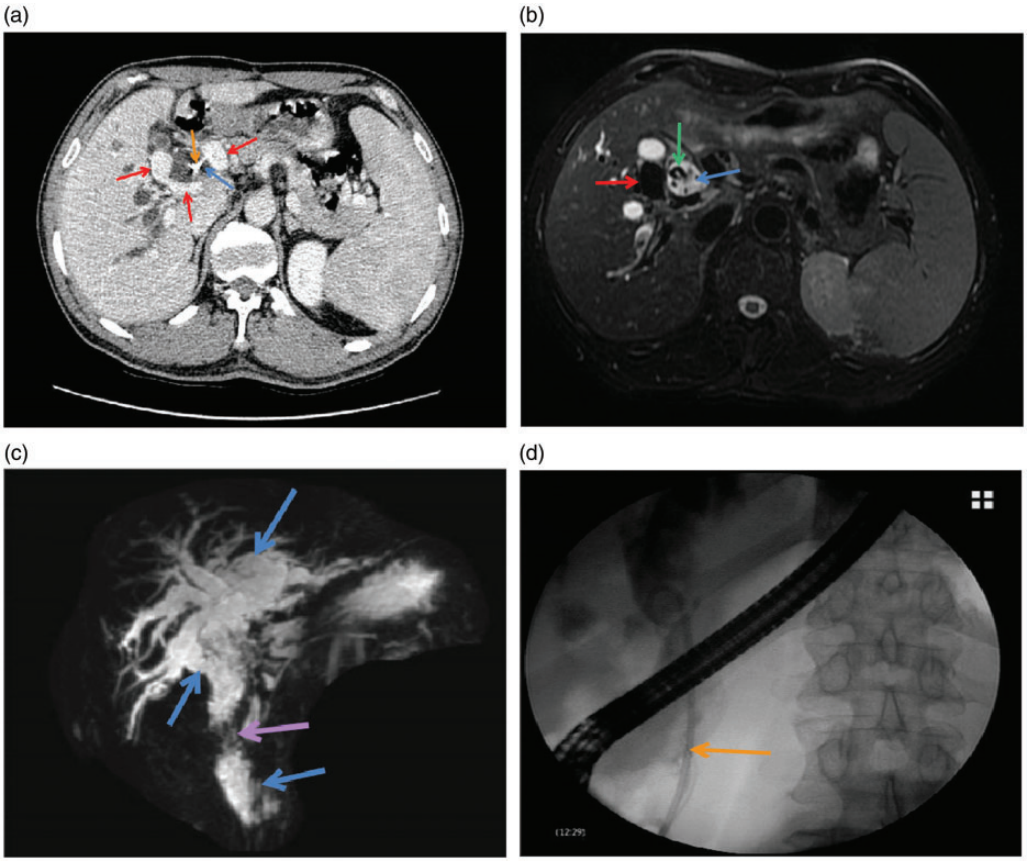

Figure 2. Computed tomography and magnetic resonance imaging + magnetic resonance cholangiopancreatography images reviewed prior to the most recent endoscopic retrograde cholangiopancreatography. (a) Computed tomography showed the dilatated collateral pathway of the portal vein, partially enclosing the bile duct (red arrows). (a–c) The intrahepatic bile duct and common bile duct were dilated (blue arrows). (a) The drainage tube shadow could be seen in the common bile duct (yellow arrow). (c) Magnetic resonance cholangiopancreatography showed obvious expansion of the intrahepatic and extrahepatic bile duct and common bile duct (blue arrows), and the lower segment of the common bile duct was relatively narrow (purple arrow). (b) The collateral pathway of the portal vein was dilatated, branched, and disorganized (red arrow), and a stone shadow could be seen in the common bile duct (green arrow). (d) In the most recent endoscopic retrograde cholangiopancreatography procedure, a new plastic stent was placed (yellow arrow).

Discussion

PHB is common in patients with EHPVO and occurs in the late stage of portal hypertension. Most patients with PHB are asymptomatic; symptoms are present in only a minority of patients and include abdominal pain, fever, and jaundice.[7] Most patients have mild hyperbilirubinemia; a markedly increased bilirubin concentration is seen in only 15% of patients, half of whom have twice the normal level of serum alkaline phosphatase.[1] Age and the duration of EHPVO are important risk factors for symptomatic PHB. Gallstones and CBD stones are other major risk factors.[8]

In the late stage, some patients may progress to cirrhosis, which manifests as hypoalbuminemia, ascites, and coagulation dysfunction.[3]

The diagnosis of PHB relies on imaging examination to detect morphological changes in the biliary tract, portal vein thrombosis, cavernous transformation of the portal vein, and atrophy of the pancreas. ERCP is the main diagnostic method for PHB.[9] One study showed that patients with PHB who underwent ERCP often had a variety of bile duct shapes such as stenosis of varying degrees and lengths, paracholedochal collaterals of the bile ducts, angulation or displacement of ducts, dilatations, and irregular walls.[10] Stone formation can be observed in some cases.[7] The extrahepatic bile duct exhibits abnormalities in 60% to 97% of patients, while the left hepatic duct shows more severe abnormalities than the right.[11] Abdominal Doppler ultrasound can display dilatation of the intrahepatic biliary radicles and changes in the portal vein.[12] Endoscopic ultrasonography can show compression of the biliary system by the collateral circulation of the portal vein and can differentiate the biliary strictures caused by PHB from those due to malignant tumors or chronic pancreatitis.[13] CT can clearly show the formation of lateral branches and cavernous transformation of the portal vein after obstruction. Clinical practice has demonstrated that MRCP and ERCP have highly similar diagnostic value for PHB.[14] The imaging features on MRCP include biliary dilations, strictures, wavy appearance of the bile ducts, angulation of the CBD, and varicose veins around the bile duct or gallbladder.[15] MRCP is widely used for diagnosis because of its noninvasive nature, while ERCP is mainly used for the treatment of PHB.

Treatment is not recommended for asymptomatic PHB. Endoscopic therapy is often used as the first-choice treatment of symptomatic PHB because of its high safety and effectiveness. Percutaneous or surgical treatment also can be considered in patients with severe symptoms.[6] The ideal therapeutic goal is to control the portal venous pressure. Multiple complications caused by portal hypertension, such as bleeding from esophageal and gastric varices and refractory ascites, can be treated effectively by TIPS. Additionally, TIPS can help to lessen the symptoms of PHB such as abdominal pain and recurrent cholangitis.[16] Bayraktar et al.[17] described a 38-year-old woman with an incomplete main portal vein web and complete thrombotic occlusion of the left portal vein causing portal hypertension. She was treated by TIPS, which resulted in disappearance of the collaterals and clinical recovery. However, TIPS for the treatment of PHB has a low success rate, and more clinical studies are needed to evaluate its efficacy.

In the present case, we were unable to determine the cause of the cavernous transformation of the portal vein, although the clinical testing excluded some genetic and acquired factors related to thrombosis. After reviewing the patient's history, we speculated that his portal vein changes might have been related to an umbilical cord infection at birth (he had been born at home). Such patients often develop symptoms at a late stage of the disease because of the long-term portal hypertension. The expanded collateral circulation exerted pressure on his bile duct, and the portal cavernous transformation led to bile duct ischemia, which ultimately resulted in poor bile excretion and stone formation. CT and MRCP confirmed the change in the bile duct morphology in this patient. Thus, the treatment was extremely challenging. All surgical procedures would have been very dangerous if decompression of the portal vein had not been performed in advance. We anticipated that TIPS would relieve the pressure; however, because of the cavernous transformation

of the portal vein, the operation was difficult and ultimately failed.

At the time of this writing, the patient still had intermittent symptoms of cholangitis and was returning to our hospital for replacement of the plastic stent by ERCP every 3 to 6 months on average. However, this is an impermanent and symptomatic therapy. We hope to find a more ideal treatment to resolve the patient's disease. A self-expandable metallic stent may be an option, but the use of an uncovered self-expandable metallic stent is associated with a high risk of uncontrollable bleeding, and safety cannot be guaranteed. A fully covered self-expandable metallic stent is expensive, and the patient refused this option because of economic difficulties. Performance of TIPS under guidance by percutaneous insertion of a balloon in the portal vein should also be considered. All of these methods are costly and carry certain risks. A reasonable plan for future treatment must be devised after detailed communication with this patient and his family.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

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ORCID iD

Zhenguo Qiao https://orcid.org/0000-0002-9079-956X

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