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IMAGE OF THE MONTH

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Portal Bilopathy

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Case report: A 63-year-old man was transferred to Keimyung University Dongsan Hospital due to abnormalities in liver function test results. Initial laboratory test on admission showed hemoglobin 14.0 g/dL (normal range, 12-18), white blood cells 6,140/μL (normal range, 5,200-12,400), platelets 188,000/μL (normal range, 130,000-400,000), prothrombin time INR 0.99, total protein 7.4 g/dL (normal range, 6.7-8.3 g/dL), albumin 4.5 g/dL (normal range, 3.2-4.8 g/dL), total bilirubin 0.5 mg/dL (normal range, 0.2-1.2 mg/dL), direct bilirubin 0.2 mg/dL (normal range, 0.0-0.4 mg/dL), AST 51 IU/L (normal range, 5-44 IU/L), ALT 51 IU/L (normal range, 5-44 IU/L), ALP 162 IU/L (normal range, 104-338 IU/L) and BUN/creatinine 18/1.6 mg/dL, HBsAg/Ab negative/negative.

Physical examination revealed soft abdomen without

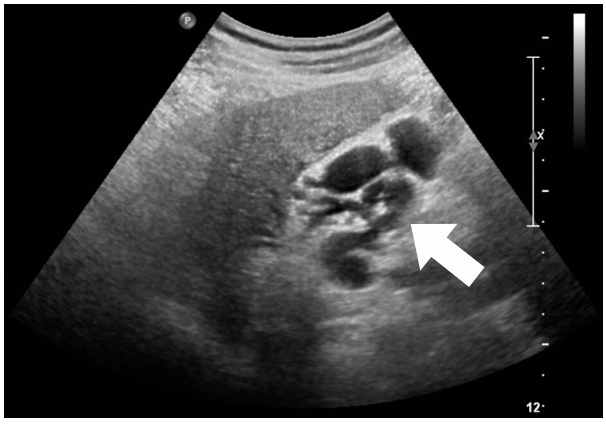

Fig. 1. Abdominal ultrasound examination revealed multisegmental narrowing of extrahepatic duct and cavernous transformation of portal vein (arrow). Intraluminal lesions were not detected in the dilated extrahepatic duct.

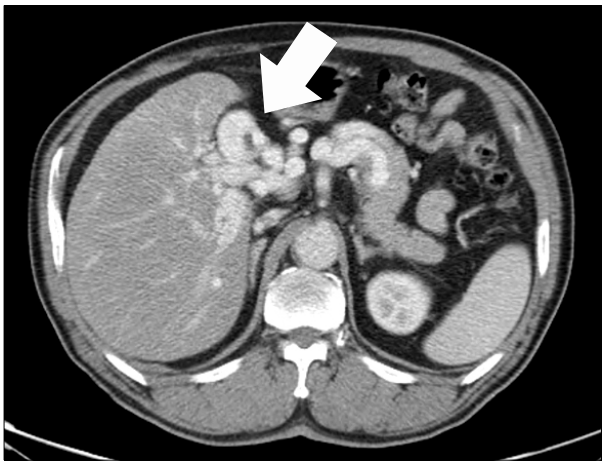

Fig. 2. Abdominal computed tomography revealed atrophied and crenate contoured liver and coarse hepatic parenchyma with fatty change. There showed a cavernous transformation of portal trunk (arrow).

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signs of ascites. There was no history of hepatic encephalopathy.

Abdominal ultrasound examination revealed multisegmental narrowing of the extrahepatic duct and cavernous transformation of the portal vein. Intraluminal lesions were not detected in the dilated extrahepatic duct (Fig. 1).

Abdominal computed tomography revealed the atrophied and crenate contoured liver and coarse hepatic parenchyma with fatty change. There showed a cavernous transformation of portal trunk (Fig. 2).

Follow-up ultrasound examinations at 9 months after the initial visit showed a slightly more prominent dilated common bile duct and an intraluminal filling defect. We, therefore, performed ERCP and MRCP.

ERCP findings revealed a focal filling defect at distal common bile duct (Fig. 3), and MRCP findings revealed a longitudinal filling defect, caused by the cavernous transformation of the portal vein without enlarged lymph nodes or neoplasms (Fig. 4).

Diagnosis: Portal hypertensive bilopathy

Although liver cirrhosis is a main cause of portal hypertension, some portions of portal hypertension occurred as a result of portal vein thrombosis, idiopathic portal hypertension and other rare diseases. Whatever the causes, portal vein occlusion is rapidly followed by compensatory mechanisms such as re-canalization and/or the development of new collateral veins around the occluded portal vein, bile duct or gall bladder. Eventually, portal vein is transformed into a cavernoma, the multiple collateral veins around the obstructed portion of the portal vein.

However, the effects of the cavernous transformation of the portal vein on the biliary tract and the pancreatic duct have not been well established yet. Occasionally, cholangiographic appearances mimic biliary tract cancer, the so-called "pseudocholangiocarcinoma" sign.

Portal hypertensive bilopathy (PHB) refers to the abnormalities of biliary tract including intra-and extrahepatic bile ducts, cystic ducts, and gallbladder in patients with portal hypertension. Commonly, the changes of biliary tract by PHB are detected as a form of extrahepatic biliary occlusion.

Gibson et al.[1] first described the relationship between extrahepatic portal vein occlusion (EHPVO) and jaundice in 1965. Williams et al.[2] were the first to report the cholangiographic changes caused by choledochal varices. This pathologic occurrence has also been termed as "portal bilopathy", "cholangiopathy associated with portal hypertension", or "portal cavernoma-associated cholangiopathy".

The frequency of PHB in patients with EHPVO is much greater than in patients with liver cirrhosis (0-33%)[3-5] or idiopathic portal hypertension (9-40%).[4,5] Prospective studies have shown that 80-100% of patients with EHPVO have PHB in ERCP. However, only a small portion of patients have such symptoms as chronic cholestasis, biliary pain or acute

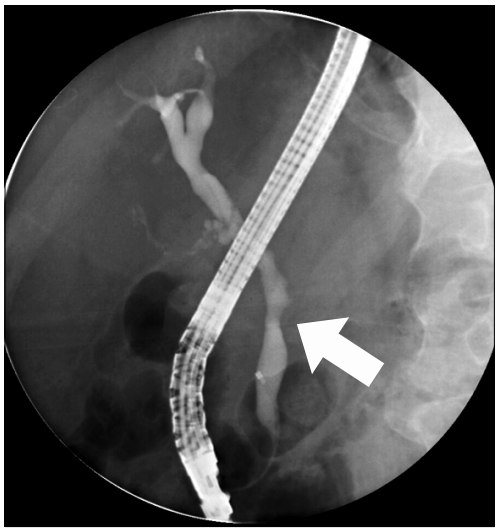

Fig. 3. Endoscopic retrograde cholangiopancreatography revealed a focal filling defect at the distal common bile duct (arrow).

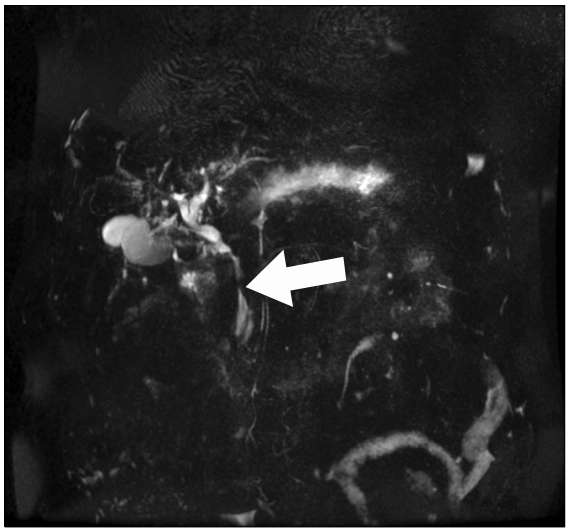

Fig. 4. Magnetic resonance cholangiopancreatography revealed a longitudinal filling defect, owing to a cavernous transformation of portal vein (arrow).

cholangitis.[4,6-8] This is probably caused by longstanding portal hypertension that results in the formation of large collaterals in the biliary regions with the formation of a typical portal cavernoma.

The exact pathogenesis of PHB is not well known. It has been postulated that the external pressure of portal cavernoma and/or ischemia may play a role.[9-11]

MRCP with portography should be the initial choice of an investigation tool for the evaluation of PHB.[12] Endoscopic ultrasound with Doppler is evolving and could provide useful extrahepatic bile duct information, especially on the causes of biliary obstruction.[13,14]

There is no consensus about the optimal treatment for PHB, as the current data regarding various forms of therapy are inconclusive. No treatment is necessary for asymptomatic patients. However, a patient with recognized symptoms should be treated as determined by the characteristics of the patient, and the treatment should focused on the management of portal hypertension and the relief of obstructive jaundice.

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