CASE REPORT

Portal Biliopathy Diagnosed Using Color Doppler and Contrast-enhanced Ultrasound

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Abstract

Portal biliopathy is a morphological abnormality of the biliary ductal and gallbladder wall associated with portal hypertension. A patient with essential thrombocythemia was initially diagnosed with extrahepatic portal vein obstruction (EHPVO). The contrast-enhanced computed tomography (CT) findings were similar to those of cholangiocarcinoma or sclerosing cholangitis. However, color Doppler and contrast-enhanced ultrasound (US) were more specific. The paracholedocheal veins around the bile ducts appeared as beads soon after the injection of contrast medium, followed by linear enhancement of the epicholedochal veins and the gradual enhancement of the whole bile ducts. These findings led to a diagnosis of portal biliopathy, which prevented the patient from having to endure hazardous procedures such as bile duct biopsies. Color Doppler and contrast-enhanced US findings are useful for diagnosing or ruling out portal biliopathy in patients who present with EHPVO.

Key words: extrahepatic venous obstruction, essential thrombocythemia, pseudocholangiocarcinoma


Introduction

Portal biliopathy can arise in patients with extrahepatic portal vein obstruction (EHPVO), which can be caused by clotting disorders, abdominal postoperative complications, malignant tumors, dehydration and neonatal umbilical vein catheterization (1). Pseudosclerosing cholangitis and pseudocholangiocarcinoma are biliary ductal and gallbladder wall abnormalities associated with portal hypertension. If portal biliopathy is misdiagnosed as pseudosclerosing cholangitis or pseudocholangiocarcinoma, performing a bile duct biopsy and pancreatic head resection is necessary but hazardous. Such procedures can be avoided using imaging modalities to prevent complications (2).

We herein describe the case of a patient with portal biliopathy diagnosed on color Doppler and contrast-enhanced ultrasound (US), which eliminated the need for biopsy or surgery.

Case Report

Upper gastrointestinal endoscopy revealed bleeding from esophageal varices in a 55-year-old woman with melena who was referred to our hospital for treatment. The patient was in a hypercoagulable state resulting from essential thrombocythemia (Table); however, her medical history and that of her family were otherwise unremarkable and did not include abdominal surgery.

The patient was alert at the time of admission with a normal liver function. Contrast-enhanced computed tomography (CT) of the abdomen revealed a slightly enhanced solid mass around the common bile duct and ductal narrowing in the portal phase (Fig. 1). The intrahepatic bile duct and pancreatic duct were not dilated, although the gallbladder wall had thickened. We considered the major portal and splenic
Figure 1. Contrast-enhanced computed tomography of the abdomen. The images show the superior or mesenteric, portal and splenic veins at the arterial (A) and portal (B) phases. The mass in the common bile duct is slightly enhanced in the portal, but not the arterial, phase (white arrow). The intrahepatic bile duct is not dilated. The gallbladder neck is enhanced (black arrow), whereas the absence of enhancement in the major portal trunk (white arrowhead) indicates portal obstruction by a thrombus.

Figure 2. T2-weighted magnetic resonance cholangiography images. The common bile duct is narrow.
Discussion

Portal biliopathy includes abnormalities of the entire biliary tract, including the intrahepatic and extrahepatic bile and cystic ducts, as well as the gallbladder, in patients with portal hypertension. The drainage veins of the common bile duct form an epicholedochal and paracoledochal venous plexus (2-4). Hypertrophy of both collateral plexuses produces a scalloped or smooth indentation in the ductal lumen of the bile duct, which can progress to more dramatic narrowing, stenosis and kinking. The development of gallbladder varices as a collateral pathway via the cystic vein is also a characteristic feature of portal biliopathy (3). Although the prevalence of this condition, and of EHPVO in particular, is still not understood in detail, elevated hepatobiliary enzymes and cholangiographic abnormalities are considered to be
Cholangiographic abnormalities, liver dysfunction and commonly associated in patients with portal hypertension. Cholangiographic abnormalities, liver dysfunction and cholangitis are found in 81%-93%, 40%-67% and 5%-18% of patients with EHPVO, respectively (5-7). The cholangiographic findings sometimes mimic bile duct cancer, with cavernomas appearing as solid tumors that are considered to be a sign of pseudocholangiocarcinoma (8). Although vascular diseases can be diagnosed on dynamic CT, the contrast patterns of the collateral and normal vessels differed in this patient. Vascular abnormalities could not be differentiated in our patient due to a weak enhancement in the portal phase (Fig. 1). The reasons for the poor enhancement remain obscure; however, we speculated that the amount of contrast medium would be diminished and would reach the epicholedochal plexus, which comprises a mesh of venous vessels of <1 mm in diameter, at different times. Moreover, such inconsistencies might also be responsible for the irregularity and tortuosity of the paracholedochal venous plexus. If the findings indicate bile duct cancer, then the patients might have to undergo risky maneuvers, such as biliary biopsies.

Patients should initially be examined using various diagnostic imaging modalities such as color Doppler and contrast-enhanced US to avoid the need for hazardous and unnecessary procedures (2). Color Doppler (3) and contrast-enhanced US can also avoid misdiagnosing porto-portal collateral veins as solid masses on gray-scale images by defining the vascular nature of the abnormalities. The drainage veins of the common bile duct form an epicholedochal plexus that appears as a mesh of venous vessels with a diameter of <1 mm on the surface of the common bile and hepatic ducts, and a paracholedochal venous plexus runs parallel to the bile duct. Contrast-enhanced US can distinguish between these two collateral plexuses (Fig. 6). Hypertrophic changes in the venous plexus have not been reported as far as we can determine. The resolution of color Doppler US is too low to distinguish these venous plexuses, whereas contrast-enhanced US can clearly differentiate them. Gallbladder varices appear as tortuous dilated vessels in or around the gallbladder wall and can be easily diagnosed using color Doppler or contrast-enhanced US.

Figure 5. Histological findings of the liver specimen. The findings of Hematoxylin and Eosin staining (A) and silver (B) staining are normal (magnification ×40).

Figure 6. Contrast-enhanced ultrasound imaging findings. Dilation of the epicholedochal and/or paracholedochal venous plexuses indicates portal biliopathy. The paracholedochal veins around the bile ducts initially became enhanced to resemble beads (arrowheads), then the epicholedochal veins immediately became linearly enhanced (white arrows, A). Thereafter, the whole bile ducts gradually became enhanced (B, C). Contrast-enhanced US clearly differentiates dilation of the paracholedochal (arrowheads) and epicholedochal (white arrows) venous plexuses.
Our patient developed a biliary abnormality without exhibiting any other symptoms, such as pruritus, jaundice, fever, abdominal pain or fever. Only the esophageal varices were treated with sclerosing therapy. The symptoms of portal biliopathy are associated with a more advanced age and a longer disease duration (9). Providing follow-up is necessary because more symptoms might emerge over time.

In conclusion, we herein described a case of portal biliopathy that was visualized on contrast-enhanced and color Doppler US. When patients present with portal hypertension, both contrast-enhanced and color Doppler US are thus considered to be useful modalities for making a differential diagnosis of portal biliopathy.

The authors state that they have no Conflict of Interest (COI).

References