Vascular and Biliary Abnormalities Mimicking Cholangiocarcinoma in Patients With Cavernous Transformation of the Portal Vein

Role of Color Doppler Sonography

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Objective. The purpose of this study was to describe and evaluate the sonographic and color Doppler features of tumorlike biliary and venous changes in patients with cavernous transformation of the portal vein (CTPV). Methods. The sonographic studies of 24 patients with CTPV were reviewed. Sonographic evaluation of the biliary system included measurement of intrahepatic and extrahepatic biliary duct caliber changes and common bile duct (CBD) wall thickening and character. Color Doppler features of the porta-portal collateral circulation at various locations (intrahepatic, periportal, gallbladder, pancreatic, and gastric regions) were carefully evaluated. Results. Biliary abnormalities were detected in 13 (54%) of 24 patients with CTPV. All 13 patients (100%) had intrahepatic biliary dilatation; 11 patients (85%) had CBD abnormalities: diffuse CBD wall thickening causing diffuse narrowing of the true lumen in 7 (54%) and CBD dilatation proximal to the focal area of narrowing due to pericholedochal compressing venous collaterals in 4 (30%). A tumorlike solid mass appeared on the gray scale images of 2 patients (8%): 1 at the porta hepatitis and the other at the pancreatic head level. Color Doppler imaging evaluation showed venous-type flow, suggesting a bulk of varicosities. Conclusions. Porta-portal collaterals in patients with CTPV may alter the biliary and venous systems, causing biliary wall thickening, stenosis, intrahepatic and extrahepatic dilatation, and pseudotumors. Detailed sonographic and color Doppler imaging assessment can show and facilitate the correct diagnosis of those changes, thus avoiding the need for a more invasive modality such as endoscopic retrograde cholangiopancreatography or a more expensive investigation such as magnetic resonance cholangiopancreatography. Key words: biliary changes; cavernous transformation; Doppler sonography; portal biliopathy; portal vein thrombosis.

Abbreviations
CBD, common bile duct; CDI, color Doppler imaging; CTPV, cavernous transformation of the portal vein; ERCP, endoscopic retrograde cholangiopancreatography

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abnormalities seen in patients with portal hypertension. Portal biliopathy is often asymptomatic but occasionally may cause clinical signs of biliary obstruction and even cholangitis. Biliary abnormalities include common bile duct (CBD) wall thickening, CBD lumen compression, biliary stricture and dilatation, focal ischemic stenosis, and secondary obstruction.4–8

Awareness of this entity is crucial because biliary changes suggestive of biliopathy are quite common and may be seen in up to 80% of patients with portal vein thrombosis. In addition, CBD narrowing and cavernoma formation may mimic cholangiocarcinoma.8–11 Although sonography and color Doppler imaging (CDI) are very commonly used for the primary evaluation of patients with CTPV, sonographic findings of portal biliopathy were described only in a small series.15 Our purpose was to demonstrate the efficiency of sonography with CDI to show the types and frequency of biliary abnormalities in patients with CTPV. The use of sonography with CDI may decrease the dependency on endoscopic retrograde cholangiopancreatography (ERCP), magnetic resonance cholangiopancreatography, and endoscopic sonography, considered the methods of choice for these abnormalities.4–14 These methods are expensive, less available, and less comfortable for the patient and sometimes use ionizing radiation (ERCP).

Materials and Methods

A retrospective review of 24 patients with CTPV and non-neoplastic disease, referred for sonographic evaluation by the Department of Gastroenterology between January 2003 and June 2006 was performed. The study was approved by our Institutional Review Board. Informed consent was waived because this is not required in retrospective studies. The age and sex of the patients and the CTPV etiology were evaluated.

Sonographic studies were performed with commercially available systems: HDI 5000 SonoCT (Philips Medical Systems, Bothell, WA) using a 2- to 5-MHz convex transducer. Sonographic evaluation included gray scale as well as CDI and pulsed Doppler imaging of the liver in all patients. Multiple parameters were evaluated, including portal vein patency, portal collateral circulation, intrahepatic and extrahepatic biliary dilatation and stenosis, CBD wall thickness, the character of CBD wall thickening (symmetric and regular or asymmetric and irregular), and the presence of biliary stones. Sonographic scanning was performed by sonographers and by experienced sonography staff physicians (A.K. and M.G.); sonographic images were interpreted on hard copy films or on picture archiving and communication systems (Centricity version 2.0; GE Healthcare, Milwaukee, WI) by 2 experienced sonography staff physicians (A.K. and M.G.).

Six patients underwent ERCP; 7 patients had upper gastrointestinal endoscopy; and 1 patient had both ERCP and endoscopy, as clinically indicated. The results of these studies were correlated with the sonographic findings.

Results

There were 24 patients, 13 female and 11 male, with an age range of 11 to 75 years (mean, 53 years). Eleven of the patients had advanced cirrhosis of various etiologies; 8 had a hypercoagulable state; and 5 had neonatal complications. Neonatal complications included omphalitis in 3 patients and a history of umbilical vein catheterization in 2 patients.

None of the pediatric patients (11–17 years old) had acute symptoms at the time of their evaluation. Of the remaining 19 adult patients, 6 had acute cholangitis.

Portal vein thrombosis with CTPV was confirmed at sonographic examination in all patients. Biliary abnormalities were present in 13 (54%) of the 24 patients. All 13 patients with biliary abnormalities (100%) had intrahepatic biliary dilatation. The CBD was involved in 11 patients (85%). Common bile duct wall thickening with luminal narrowing was present in 7 patients (54%). Two types of CBD wall thickening were observed: symmetric and regular (Figure 1A) in 5 patients versus asymmetric and irregular (Figure 1B) in 2 patients. Color Doppler evaluation of the thickened CBD wall, either regular or irregular, showed flow within the thickened wall consistent with intramural portal vein collaterals (Figure 2).
Four patients (30%) had CBD dilatation due to external compression by engorged venous collaterals in the porta hepatis. The CBD wall was normal in these patients (Figure 3).

Choledocholithiasis in a narrowed CBD segment was diagnosed by sonography in 4 patients (30%). Only 1 of them also had gallbladder stones. In 3 patients, a bile duct stricture was seen on ERCP. The stricture was treated by dilatation and stenting, and the stones were extracted. The diagnosis of the fourth patient

Figure 1. A, Regular thickening of the CBD wall (arrows) secondary to intramural collaterals. B, Irregular thickening of the CBD wall (arrows) secondary to intramural collaterals associated with perihepatic fibrosis, scarring, angulation, and distortion of the CBD wall.

Figure 2. Color Doppler imaging shows collaterals within the regular smoothly thickened CBD wall (A, arrows) as well as within irregular thickening of the CBD wall (B, arrows). PORTA HEP indicates porta hepatis.
was confirmed at surgery. This patient was transferred to our institution after being treated at an outside institution with a cholecystectomy and surgical removal of the CBD stone. This procedure was complicated by substantial bleeding from the porta hepatis venous collaterals and inadvertent ligation of the CBD. After confirmation of the diagnosis of CTPV and CBD ligation at our institution, a stent was percutaneously placed, and a hepaticojejunostomy was performed.

An apparently solid “mass” was detected on gray scale sonography in 2 patients of the whole cohort (8%): 1 at the porta hepatis (Figure 4A) and the other at the pancreatic head region. Both patients were initially suspected to have neoplastic disease. Contrast-enhanced computed tomography (Figure 4B) showed a hyperdense mass, also suspected as representing a neoplasm. However, power Doppler evaluation (Figure 4C), especially spectral Doppler imaging (Figure 4D), showed the mass to be a pseudotumor consisting of a conglomerate of collateral varicosities, representing a cavernoma. Endoscopic retrograde cholangiopancreatography (Figure 4E) of the same patient showed the external compression of these engorged collaterals on the CBD wall, representing as a filling defect. These 2 patients had no evidence of malignancy during a follow-up of 3 years from the time of diagnosis.

Discussion

Portal vein thrombosis is the leading cause of extrahepatic portal hypertension in the western world. It may be the result of multiple etiologies such as neoplastic invasion or compression of the portal veins, hypercoagulable states, sepsis, noninfectious inflammatory processes, and myeloproliferative disorders. Radiologists’ familiarity with the manifestations of portal vein thrombosis is crucial for accurate and timely diagnosis, which is the basis for appropriate treatment and intervention. Most imaging manifestations of portal vein thrombosis have been described extensively.

However, the associated biliary ductal and gallbladder wall abnormalities, also called portal biliopathy, and especially the sonographic imaging findings, have not received enough attention in the radiology literature. To the best of our knowledge, our study is the largest that evaluated patients with CTPV and portal biliopathy with state-of-the-art sonography and Doppler imaging.

Biliary abnormalities are quite frequent in patients with chronic extrahepatic portal vein thrombosis and CTPV. The incidence of biliary abnormalities varies depending on the imaging modality used. In studies using ERCP, biliary changes were shown in 70% to 100% of cases. By magnetic resonance cholangiopancreatography, biliary abnormalities were seen in 92% of cases, and by endoscopic sonography, changes compatible with portal biliopathy were shown in 52% to 76% of cases.

In our study, portal biliopathy was shown in 54% of patients with CTPV by gray scale sonography and CDI. We found only 1 prior study using CDI to evaluate portal biliopathy. In this study of 46 patients with extrahepatic portal vein thrombosis, a 7% incidence of biliary changes was noted. However, not all patients had CTPV, possibly suggesting a less severe degree of portal hypertension. This may explain their low incidence in comparison with most other reports as well as our own study results.

Biliary changes in patients with CTPV are postulated to result from extrinsic compression of the collateral vessels on the bile ducts. In addition, there is associated pericholedochal fibrous

Figure 3. Color Doppler imaging shows engorged collaterals around the CBD, causing external compression, stasis, and subsequent ductal dilatation. Note that there is no effect on the thickness of the CBD wall itself (arrows).
Figure 4. A, Mass (m) in the porta hepatitis initially interpreted as a tumor. B, Contrast-enhanced computed tomography in the portal phase shows an enhancing mass in the porta hepatitis (between arrows) with a differential diagnosis of a tumor versus a conglomerate of blood vessels. C and D, Power Doppler (C) and pulsed Doppler (D) sonography show this mass to be a network of blood vessels, all with venous type of flow, consistent with abundant collaterals forming a cavernoma pseudotumor. E, Endoscopic retrograde cholangiopancreatography shows a serpentine filling defect on the distal CBD (arrows), representing the engorged collaterals’ impression on the CBD wall.
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...that may distort the bile duct anatomy, sometimes causing angulation of the bile ducts with possible ischemic injury. This can result in stricture formation, bile duct irregularity, and proximal dilatation.

Two venous systems run along the extrahepatic bile ducts: the paracholedochal and the epicholedochal veins. The paracholedochal veins run parallel to the bile ducts and when dilated may cause extrinsic compression on the bile duct lumen. Dilatation of the epicholedochal veins, which constitute a reticular mesh running on the surface of the bile duct itself, may cause regular or irregular ductal wall thickening.4–8

We found some degree of biliary dilatation in all patients with portal biliopathy. Biliary tract abnormalities included CBD narrowing and proximal dilatation at the extrahepatic as well as intrahepatic levels. Two types of CBD narrowing were observed: (1) extrinsic, probably due to external compression of the CBD by surrounding engorged collaterals, in 4 patients; and (2) intrinsic, caused by intramural collaterals, as clearly shown by CDI in 7 patients. The latter type, intrinsic collaterals, manifested itself as wall thickening of either a regular (5 patients) or an irregular (2 patients) shape. We assume that the irregular wall thickening seen in these 2 patients represented pericholedochal fibrosis with associated scarring, angulation, and distortion of the CBD wall.

Our study shows the ability of abdominal sonography using CDI to depict these different types of pathologic collateral systems and their effect on the biliary system. Irregular CBD wall thickening may mimic cholangiocarcinoma on a gray scale display and has been called the pseudocholangiocarcinoma sign. However, the enlarged varicose veins can be easily identified with CDI evaluation.

When collaterals are large and abundant, they may form a pseudotumoral mass at the porta hepatitis or at the pancreatic head region, as seen in 2 of our cases. Biopsy or resection of this suspected mass may result in massive bleeding.16

Using Doppler imaging and awareness of this manifestation of CTPV enabled the diagnosis of a cavernoma presenting as a pseudotumor.

Spectral Doppler imaging should be always used in addition to CDI and power Doppler imaging to show that flow within this plexus of abnormal vasculature is all portal venous type flow and that indeed no arterial signal is present. Therefore, that would be a good way to differentiate cavernous transformation from tumor neovascularity, which would show arterial flow.

Choledocholithiasis may develop in 5% to 20% of cases17 and is usually removed at ERCP. Open surgery is usually avoided because of the high risk of life-threatening bleeding from collaterals. In our study, 4 (17%) of the 24 patients had choledocholithiasis, which was treated by ERCP or surgery.

Strictures may be treated by dilatation or stent placement. In selected severe cases of biliary obstruction secondary to portal biliopathy, surgical portosystemic shunting can be performed for portosystemic decompression of the engorged collaterals, thus reducing the effect of these collaterals on the biliary system.16–18

In conclusion, sonography is a useful tool in the evaluation of patients with CTPV, complementing other modalities used to evaluate these patients. It allows diagnosis of the biliary changes frequently associated with CTPV. In addition, CDI may enable the diagnosis of pseudotumoral masses, thus avoiding possible biopsy and patient morbidity.

References


