Portal hypertensive biliopathy presenting as acute jaundice with suspected intrahepatic biliary strictures and stones

A 58-year-old woman with liver cirrhosis, previously decompensation with ascites, presented with jaundice. Abdominal ultrasound identified a shrunken, cirrhotic liver with intrahepatic duct dilatation within the left liver lobe, gallbladder calculi, and a normal bile duct. Subsequent magnetic resonance cholangiopancreatography identified dilated left and right intrahepatic ducts containing several filling defects, with strictureing of the left main duct and a normal-caliber common bile duct (▶Fig. 1). Endoscopic retrograde cholangiopancreatography (ERCP) with cholangioscopy was performed to assess the stricture and remove presumed intrahepatic stones. ERCP revealed similar features, with strictureing of the left and right main hepatic ducts and significant dilatation of the left intrahepatic ducts, which contained apparent filling defects (▶Fig. 2). Cholangioscopy was performed with the Spy-Glass DS II (Boston Scientific). The scope was passed to the left and right intrahepatic ducts, where vascular structures were identified protruding into the lumen with focal dilatations occupying more than 75% of the lumen (▶Video 1). These structures followed the route of the bile ducts, consistent with choledochal varices. The common bile duct was spared and there were no biliary calculi throughout the intrahepatic and extrahepatic ducts.

The patient was started on a nonselective β-blocker (carvedilol) and the jaundice improved slightly (from 65 to 45 µmol/L). Abnormalities of the biliary tree relating to portal hypertension can be referred to as “portal hypertensive biliopathy” and are commonly seen in patients with extrahepatic portal venous obstruction (81%–100%), but less commonly in patients with cirrhosis (0%–33%) [1]. The abnormalities described include strictures, caliber irregularities, segmental dilatation, and indentations. There is also a predisposition to involvement of the left intrahepatic ducts (55%–100%) more than the right (40%–56%) [1]. The focus of treatment is to ensure adequate biliary drainage by endoscopy, or, if this fails, to consider portosystemic shunt.

Competing interests

The authors declare that they have no conflict of interest.
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