Choledochal cyst type IA with drainage through the ventral duct in pancreas divisum

Choledochal cyst is a rare disease in Western adults. It is frequently associated (about 70%) with an abnormal pancreaticobiliary junction, but a choledochal cyst occurring with pancreas divisum is a very rare condition. This is, to our knowledge, the first published case of a choledochal cyst draining into the ventral duct system in cases of pancreas divisum.

A 28-year-old female patient was referred to our department with cholestatic jaundice (bilirubin 15 mg/dL) and abdominal pain. She was obese but had been healthy all her life. During the first emergency endoscopic retrograde cholangiopancreatography (ERCP), stenosis was seen in the distal part of the common bile duct (CBD), and the proximal part showed signs of cystic dilation (**Fig. 1**). We placed a 6-Fr nasobiliary catheter and scheduled a second ERCP.

During the second ERCP, we found the distal part of the CBD to have small side branches like a pancreatic duct (**Fig. 2**). Also, a minor papilla was found and cannulated, and showed a normal dorsal pancreatic duct (**Fig. 3**). So our endoscopic diagnosis was a type IA choledochal cyst with an abnormal biliopancreatic junction. Drainage of the CBD was achieved through the ventral duct of the pancreas divisum.

To minimize the risk of cancer and because biliary drainage was impossible we referred our patient to surgery. Biliary cysts are associated with cholangiocarcinoma: 10%–30% of adults with biliary cysts present with cancer [1]. A pancreaticobiliary maljunction may contribute to the high cancer risk due to reflux of pancreatic enzymes [2]. Because of the risk of malignant degeneration of the cyst, current standard therapy is surgical resection [3]. Our patient underwent laparotomy with surgical resection of the choledochal cyst and hepaticojejunostomy (**Fig. 4**). Histology showed chronic inflammation but no signs of cholangiocarcinoma. By postoperative day 10 she had completely recovered and was discharged from hospital.

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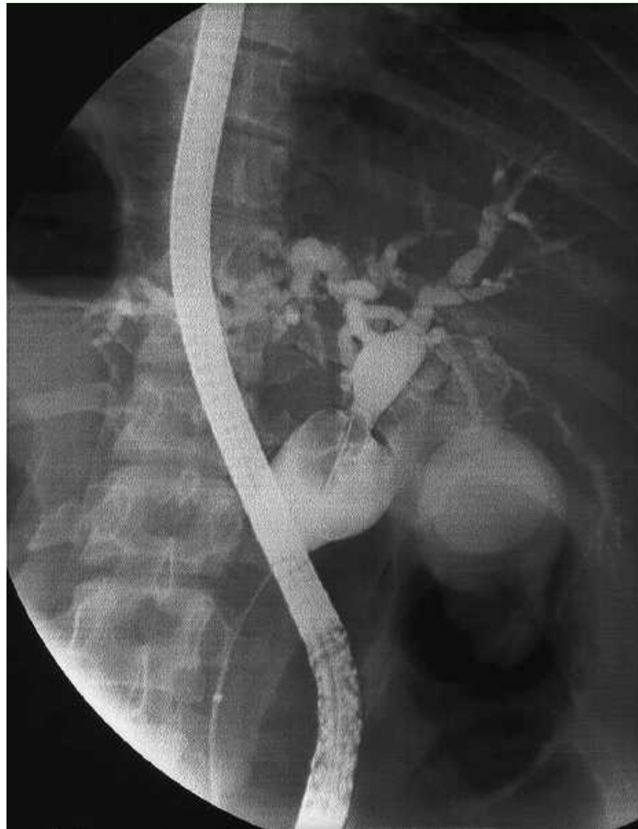

Fig. 1 Initial appearance of the common bile duct (CBD): stenosis with pre-stenotic dilation of the CBD and a large stone.

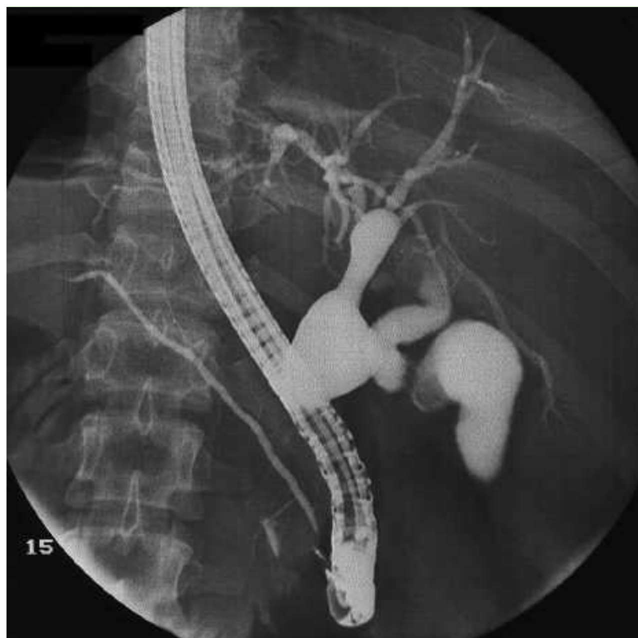

Fig. 2 Cannulation of the minor papilla: the dorsal pancreatic duct in pancreas divisum.

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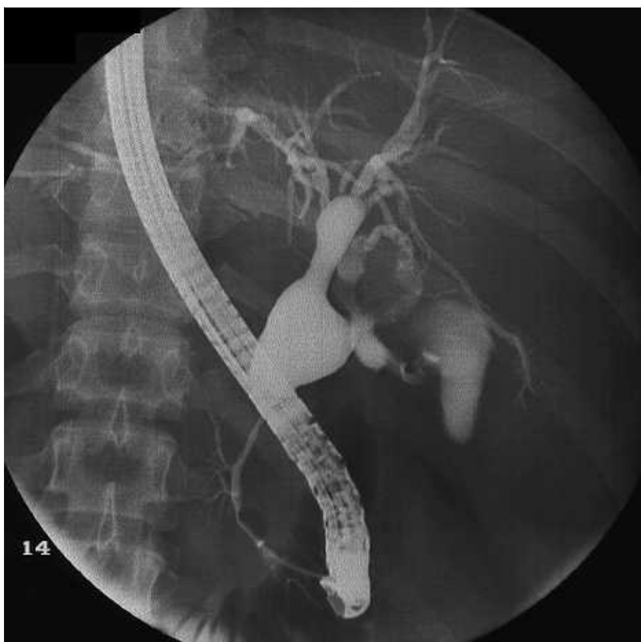

Fig. 3 Type IA choledochal cyst with drainage of the common bile duct through the ventral duct in pancreas divisum.

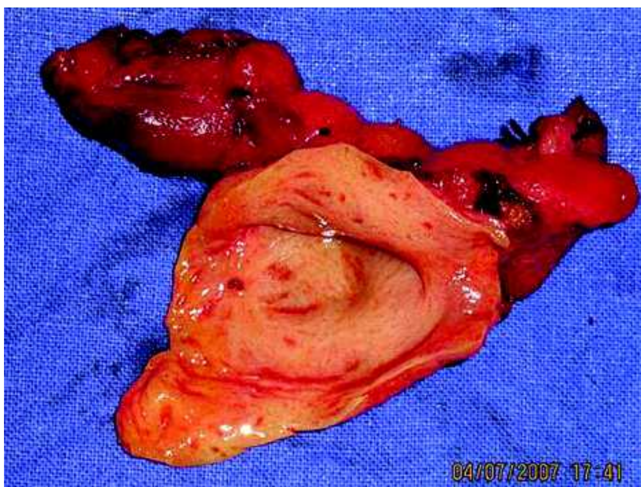

Fig. 4 Surgically resected specimen of the choledochal cyst.

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Bibliography

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