Diagnostic challenge: Mirizzi syndrome with anomalous pancreaticobiliary junction

A 35-year-old man was admitted to our hospital with abdominal pain after eating. A double bile duct structure was revealed by magnetic resonance cholangiopancreatography. The relationship and alignment of the double bile ducts appeared to be poorly represented (▶Fig. 1). Endoscopic retrograde cholangiopancreatography (ERCP) was performed, and showed two separate openings in the zone of the major duodenal papilla (▶Fig. 2, ▶Video 1). The cholangiogram revealed that the patient had two common bile ducts (CBDs) that converged in the middle section of the bile duct, and the stones were disclosed in both ducts. Meanwhile, the pancreatic duct converged into the left CBD (▶Fig. 3). We diagnosed type II double common bile duct (DCBD) [1] with anomalous pancreaticobiliary junction (APBJ) and cholelithiasis (▶Fig. 4 a).

The patient underwent laparoscopic Roux-en-Y hepaticojejunostomy due to the risk of cholangiocarcinoma. Interestingly, we discovered during the operation that the left CBD shown by the cholangiogram was in fact the cystic duct (▶Fig. 5, ▶Video 1). This indicated that the cystic duct formed a fistula with the CBD, and the pancreatic duct met the cystic duct. This suggested a deviation in our initial diagnosis. Based on the information above, we concluded that the cystic duct around the CBD on the left drained independently into the duodenum while forming a fistula with the duct in the middle. In fact, the cystic duct was where the pancreatic duct converged. The patient was eventually diagnosed with type II Mirizzi syndrome [2] in conjunction with a specific type of APBJ (▶Fig. 4 b).

Both Mirizzi syndrome and DCBD are rare diseases of the biliary system, and ERCP is the current gold standard for diagnos-
ing them [1, 3]. However, this is the first report of Mirizzi syndrome in conjunction with APBJ that challenged our initial diagnosis; maybe choledochoscopy can make up for the lack of diagnosis [4, 5].

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Competing interests

The authors declare that they have no conflict of interest.

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Fig. 5 Laparoscopic operation findings. 

a A right channel (green circle) with bile and a left channel (white circle) with colorless fluid could be clearly seen after cutting the common bile duct. 

b The left channel extended into the gallbladder and the right channel extended into the liver.