A 62-year-old woman presented with marked cholestasis (bilirubin 9.8 mg/dL), and ultrasound suggested intrahepatic biliary dilation down to the hilum and a solid-filled gallbladder. Claustrophobia precluded magnetic resonance imaging, but computed tomography imaging excluded distant metastases. Endoscopic ultrasound suggested an anomalous pancreaticobiliary junction (APBJ) with extra-mural pancreaticobiliary confluence, a solid gallbladder mass, and an independent, vague 12 mm hypoechoic hilar lesion, thus suggesting double carcinoma formation in the biliary tract [1, 2].

Endoscopic retrograde cholangiopancreatography (ERCP), under antibiotic coverage with oral levofloxacin for 3 days, revealed a long common channel of 12 mm with associated APBJ. Of note, the pancreaticobiliary ductal junction remained functionally uninterrupted during sphincter contraction, thus favoring true pancreaticobiliary maljunction (PBM) without biliary dilation over high confluence of pancreaticobiliary ducts (HCPBD) (▶Fig. 1).
Bile aspirate analysis from the middle portion of the common bile duct indicated high amylase levels of 8987 U/L. Video 1 shows the in vivo functional characterization of pancreaticobiliary reflux (PBR). Consistent with the pro-carcinogenic effects on the biliary mucosa related to PBR, a high-grade Klatskin lesion was shown on cholangiography, and was confirmed by aspiration and brush cytology (▶Fig. 2). After bilateral stenting with complete contrast media drainage and slow bilirubin normalization, the patient was referred for hepatobiliary surgery.

APBJ is rare in Western populations and, relative to functional separation capacities of the sphincter apparatus, dichotomizes into PBM (with or without biliary dilation) or HCPBD. In vivo characterization of PBR as backflow of pancreatic juice into the biliary system in PBM had been reported via minor papilla access pancreatography but not by standard, though specifically dedicated, ERCP [3,4].

Fig. 2 Malignant hilar stricture complicating pancreaticobiliary maljunction. a Cholangiography indicated an extensive high-grade hilar stricture with interrupted right–left communication consistent with a Bismuth Corlette III lesion. b Aspiration and brush cytology confirmed the presence of adenocarcinoma.

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Competing interests

None