Dramatic imaging changes of a biliary stricture within 8 months in a rare case

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QUESTION

A 62-year-old man sought treatment due to intermittent upper abdominal pain accompanied by jaundice for 5 days. The patient was healthy and had no history of hypertension or diabetes. Liver function examination revealed the following: aspartate aminotransferase level, 138 U/L (1-49 U/L); alanine aminotransferase level, 275 U/L (1-49 U/L); total bilirubin (TBIL) level, 65.5 μmol/L (9-30 μmol/L); direct bilirubin (DBIL) level, 30.0 μmol/L (0-6.8 μmol/L); alkaline phosphatase (ALP) level, 499 U/L (20-125 U/L); gamma-glutamyl transferase (GGT) level, 860 U/L (3-69 U/L); and cancer antigen 19-9 (CA19-9) level, 253.0 U/mL (normal level, <27 U/mL). Abdominal ultrasonography showed common bile duct dilatation, and the results of abdominal magnetic resonance cholangiopancreatography (MRCP) (Figure 1a, b) and enhanced computed tomography (CT) (Figure 1c, d) indicated localized lower bile duct stenosis and upper dilatation, suggesting that a malignant biliary tumor should not be excluded. As the patient's maximum fasting blood glucose level, at admission, was 30 mmol/L, we decided to conduct a follow-up diagnosis after adjusting the blood glucose to a normal level. The jaundice, however, subsided during the treatment, and the patient's bilirubin decreased to a normal level. Additionally, the follow-up examination showed a CA19-9 level of 34.4 U/mL. Therefore, the evidence for a clinical diagnosis of a tumor was insufficient. Regrettably, the patient refused to undergo endoscopic ultrasonography-guided fine needle aspiration (EUS-FNA) and was discharged and followed up in the outpatient clinic. Contrast-enhanced abdominal CT scan conducted at 2 months after discharge showed that the localized extrahepatic bile duct wall was significantly thicker than before, with no other significant changes.

Ten days later, the patient experienced sudden upper abdominal pain with yellowing of the sclera and skin. A liver function examination revealed the following: TBIL, 116.5 μmol/L; DBIL, 69.8 μmol/L; ALP, 252 U/L; GGT, 260 U/L; and CA19-9, >1,000 U/mL. MRCP (Figure 2a,b) and enhanced abdominal CT (Figure 2c,d) revealed diffuse common bile duct wall thickening with obvious hepatic hilar and distal bile duct stenoses, gallbladder wall thickening, and pancreatic head enlargement.

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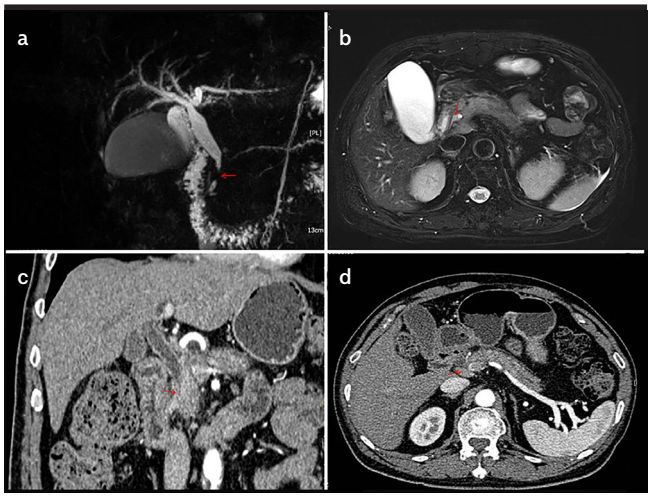

Figure 1. a-d. Abdominal MRI+MRCP and enhanced abdominal CT: a) MRCP shows localized stenosis (red arrow) in the lower segment of the common bile duct and slight dilatation in the upper segment. b) T2-weighted imaging (T2 WI) staging shows localized thickening (red arrow) of the bile duct wall in the pancreatic segment and a slightly enlarged pancreatic head with an uneven signal; the possibility of a tumor was not excluded. c) CT with coronal reconstruction with thickening of the common bile duct wall in the lower segment (red arrow). d) Inflammatory changes in the pancreas, with enlarged lymph nodes behind the common bile duct (red arrow).

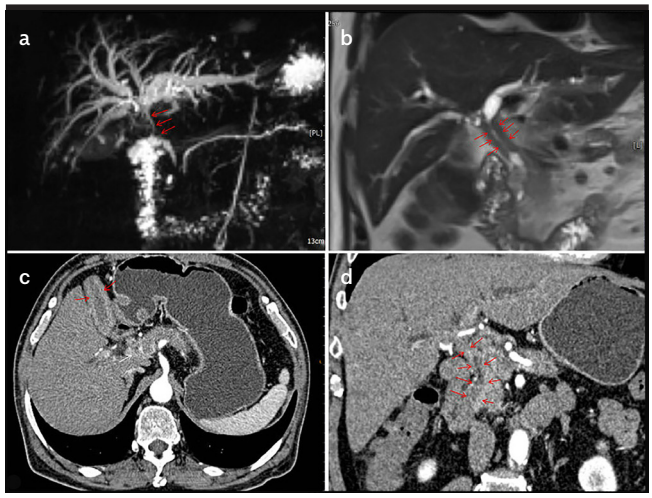

Figure 2. a-d. Abdominal MRI+MRCP and enhanced abdominal CT: a) MRCP reveals obvious stenosis of the common hepatic duct and common bile duct in the upper pancreatic segment (red arrow), with a tail-like change; the main pancreatic duct of the pancreatic tail shows a bead-like change. b) Coronal T2 WI shows diffuse annular thickening of the common hepatic duct and a common bile duct with stenosis (red arrow), eccentric nodules in the lower segment of the common bile duct, and a suspicious tumor in the lower segment. c) Uniform thickening of the gallbladder wall (red arrow), enlarged lymph nodes (white arrow) in the posterior common bile duct, obvious annular thickening of the common bile duct, a full pancreas, and an anterior fibrosis sheath are shown. d) CT with coronal reconstruction shows further extrahepatic bile duct thickening (red arrow), and the lumen is almost occluded.

What is the most likely diagnosis for the patient?

ANSWER

IgG4-related sclerosing cholangitis (IgG4-SC) combined with type 1 autoimmune pancreatitis (AIP)

The patient's jaundice subsided again during treatment, the CA19-9 level decreased to 36 U/mL, and abdominal MRCP revealed a thinning gallbladder and common bile duct walls (Figure 3). The serum IgG4 concentration was 2,040 mg/L (normal level, <1,350 mg/L). EUS-FNA was performed to exclude a malignant pancreatic tumor. The pathological results of the pancreatic biopsy showed small amounts of plasma cells, and no malignant tumor cells were found. IgG4-SC combined with AIP was considered based on the clinical symptoms, disease progression, imaging changes, and pathology and laboratory results. According to the latest guideline for IgG4-SC in Japan (1), this case belongs to type 3, which is characterized by stenosis in the hilar hepatic lesions and lower bile duct, and accounts for approximately 10% of all IgG4-SC patients.

Due to stenosis in the bile duct, recurrent obstructive jaundice occurred; therefore, endoscopic retrograde cholangiopancreatography (ERCP) (Figure 4a) was performed, and intraoperative intraductal ultrasonography (IDUS) (Figure 4b) suggested annular thickening of the entire segment of the bile duct wall. Stenosis in the lower end of the bile duct and head of the pancreatic duct was observed using cholangiopancreatography; thus, biliary and pancreatic duct stents were placed. Oral prednisolone (Prednisolone acetate tablets; Sine pharmaceutical factory, Shanghai, China) (0.6 mg/kg/day) was also started. A contrast-enhanced abdominal CT after 2 months of prednisolone treatment revealed that the gallbladder and bile duct walls were significantly thinned, and an arterial phase scan revealed that the enhanced nodules in the pancreatic head were significantly reduced. The prednisolone dosage was then reduced by 5 mg and gradually tapered by 5 mg every 2–4 weeks, thereafter reaching 5 mg/day over 6 months for maintenance treatment. An abdominal CT review at 4 months after initiation of steroid treatment suggested that the extent of thickening in the gallbladder and common bile duct walls has been reduced, and the

enlargement of the lymph nodes behind the common bile duct had decreased. Subsequently, ERCP was performed to remove the biliary and pancreatic duct stents. No recurrence was observed at the 17-month follow-up.

IgG4-related disease is an autoimmune disease involving multiple organs and often, the biliopancreatic system. Overall, 92%-95% of IgG4-SC cases are combined with type 1 AIP (2). The diagnosis of this disease is difficult. In clinical practice, more than 40% of AIP cases are treated as pancreatic malignancies and undergo pancreatectomy (3). The imaging manifestations of the bile duct need to be distinguished from those of primary sclerotic cholangitis (PSC) and cholangiocarcinoma, and the imaging changes of the pancreas should be differentiated from those of pancreatic cancer. The biliary tract wall thickening of IgG4-SC is characterized by uniform annular thickening of at least 0.8 mm in biliary stricture and non-stricture areas, mostly combined with thickening of the gallbladder wall. These imaging features are different from those of cholangiocarcinoma and PSC (3). The intraoperative observation of the papillary morphology during ERCP can also help to further distinguish sclerosing cholangitis with autoimmune pancreatitis (SC-AIP) and PSC. A study by Kubota K. demonstrated that 17 (63%) of 27 SC-AIP cases exhibited morphological characteristics of duodenal papilla swelling, and the papillae in 12 PSC cases were essentially in contracture status (4). Some AIP and pancreatic cancer may accompany diabetes (2). Endo T et al. (5) suggested that amylase alpha-2A autoantibodies can be used as a new marker for the diagnosis of AIP and can be differentiated from pancreatic tumors. Additionally, a comprehensive judgment should be made based on medical history, clinical manifestations, laboratory tests, imag-

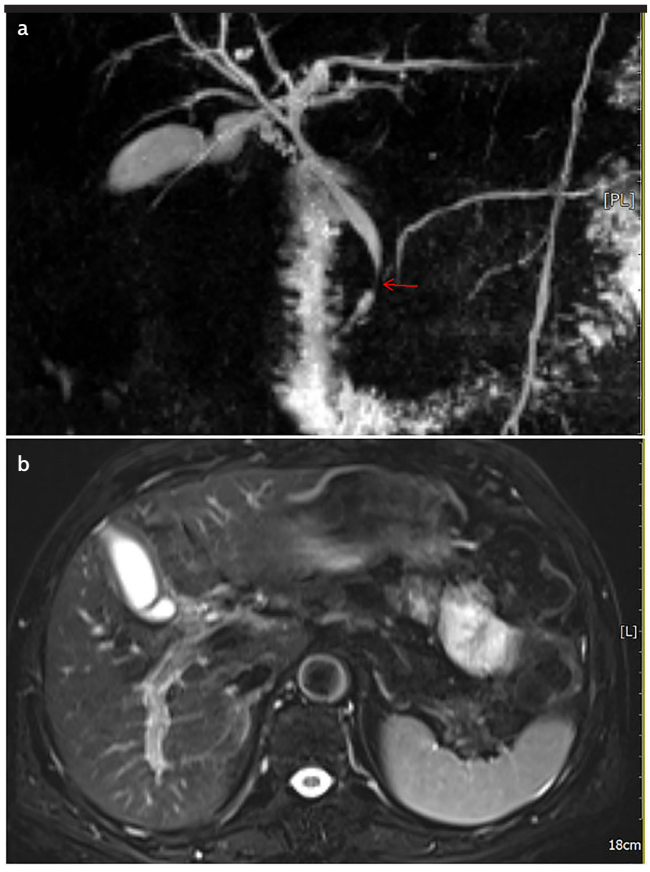

Figure 3. a, b. Abdominal MRI+MRCP: a) MRCP shows the extent of reduction of the extrahepatic bile duct stenosis (red arrow), with dry branch-like changes in the intrahepatic bile duct with uneven thickness; the thickness of the main pancreatic duct also shows uneven changes. b) The results of T2 WI fat saturation sequencing show thinning of the gallbladder and common bile duct wall, a reduced extent of bile duct stenosis, and slightly reduced lymph nodes behind the common bile duct.

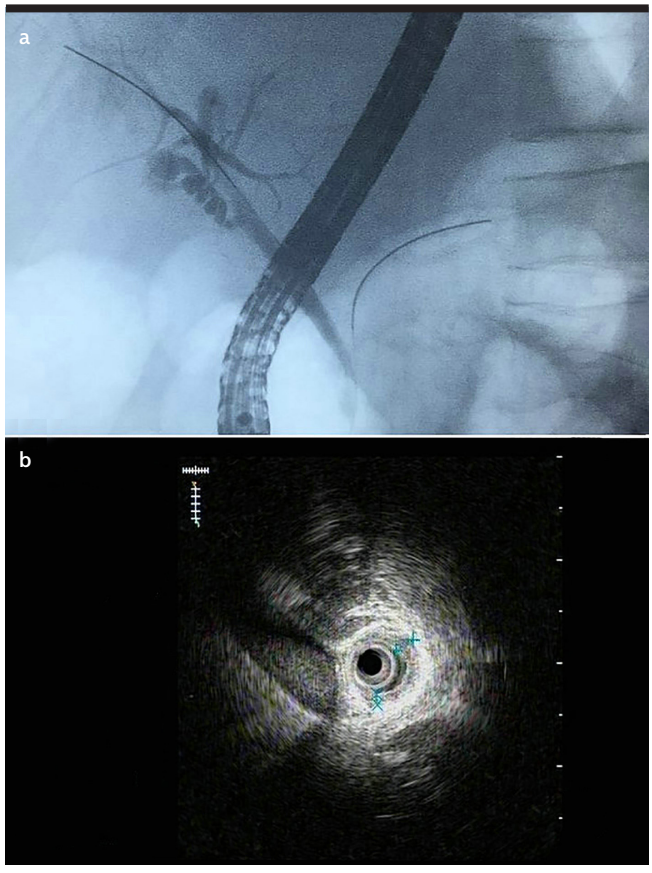

Figure 4. a, b. Imaging findings during ERCP and IDUS: a) ERCP shows localized stenosis at the lower end of the biliary tract and no dilatation of the common bile duct. b) IDUS: The wall of the entire common bile duct is obviously thickened; the thickest part is 2.1 mm.

ing findings, and pathological results. The natural disease progression during the first 4 months and subsequent imaging changes over a period of 8 months after the diagnosis and intervention with ERCP were demonstrated, and a steroid therapy was administered to the patient.

Traditionally, cases of AIP with obstructive jaundice require placement of biliary stents using ERCP to reduce jaundice before the steroid treatment; however, in the study by Bi Y., 15 cases of AIP with jaundice were directly treated with steroids, without the placement of biliary stents (6). Due to the small sample size, determination of whether the placement of biliary stents is necessary for all AIP cases with jaundice will require further study. It is noteworthy that IgG4-SC can progress to end-stage liver disease within a few months; therefore, the diagnosis should be confirmed, and effective treatment should be provided in a timely manner. Cholangiocarcinoma resulting from IgG4-SC is thought to be a rare complication, and rarely, they both may be present in the same patient. As a relatively new concept for clinicians, IgG4-SC is not considered a risk factor for cholangiocarcinoma (1). The differential diagnosis should receive sufficient attention.

Ethics Committee Approval: Ethics committee approval for this study was received from the Ethics Committee of the first hospital of lanzhou university (Date: Sep 10, 2018; Number: LDYYLL2018-172).

Informed Consent: Written informed consent was obtained from the patients who participated in this study.

Peer-review: Externally peer-reviewed.

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Conflict of Interest: The authors have no conflicts of interest to declare.

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