IgG4-related sclerosing cholangitis involving the gallbladder mimicking a hilar cholangiocarcinoma

A 76-year-old man presented with abdominal pain, jaundice, and fever. Abdominal computed tomography demonstrated wall thickening of the gallbladder with gallstones surrounding a large low-density lesion communicating with the gallbladder and a thickened, enhancing wall at the hepatic hilum and common hepatic duct (CHD). Magnetic resonance cholangiopancreatography (MRCP) showed a stricture at the hilum extending to the CHD, suggestive of a hilar cholangiocarcinoma (Fig. 1). Percutaneous catheter drainage for liver abscess and endoscopic retrograde cholangiopancreatography with stent placement for biliary drainage and biopsy was performed. Histology did not show malignancy. Same-session endoscopic ultrasound (EUS)-guided tissue acquisition using a 22-gauge needle (Fig. 2) was performed for the biliary lesion. EUS revealed symmetrical and smooth wall thickening of the hilum and CHD. EUS-guided tissue acquisition was performed using a 22-gauge needle (Video 1). POCS (SpyGlass DS; Boston Scientific, Natick, Massachusetts, USA) demonstrated a stricture with dilated vessels and hyperemic, edematous mucosal surface, and a papillary-appearing mucosal projection. SpyBite forceps biopsy specimens were taken from the abnormal mucosal lesions (Video 1). Specimens were obtained using the SpyBite biopsy forceps (Boston Scientific). The pathology results from EUS-guided tissue acquisition and SpyBite forceps biopsy showed stromal fibrosis with lymphoplasmacytic infiltration and more than 10 IgG4-positive plasma cells per high-power field (Fig. 3).
cells per high-power field (HPF) (▶ Fig. 3). Serum IgG4 level was 185 mg/dL. Laparoscopic cholecystectomy was performed, and on histology the wall of the gallbladder showed multifocal lymphoplasmacytic infiltrations with more than 10 IgG4-positive plasma cells per HPF. The patient received steroid treatment at a dosage of 40 mg/day. After 4 weeks of steroid treatment, MRCP demonstrated improved luminal narrowing of the hilum and CHD (▶ Fig. 4). Therefore, IgG4-related sclerosing cholangitis involving the gallbladder was diagnosed. The patient was placed on long-term low-dose steroid treatment, the biliary stent was removed, and the patient has now had no recurrence of the cholangitis for over 1 year.

IgG4-related sclerosing cholangitis is difficult to differentiate from malignancy [1]. EUS-TA and POCS may be a useful modality for evaluating an indeterminate hilar stricture [2, 3].

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Competing interests
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