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 and peroral cholangioscopy (POCS) were performed. EUS revealed symmetrical and smooth wall thickening of the hilum and CHD. EUS-guided tissue acquisition was performed using a 22-gauge needle (Fig. 2). POCS (SpyGlass DS; Boston Scientific, Natick, Massachusetts, USA) demonstrated a stricture with dilated vessels and hyperemic and papillary-appearing mucosal projection. SpyBite forceps biopsy specimens were taken from the abnormal mucosal lesions.

Video 1 Peroral cholangioscopy reveals a stricture with dilated vessels, a hyperemic, edematous mucosal surface, and a papillary-appearing mucosal projection. SpyBite forceps biopsy specimens were taken from the abnormal mucosal lesions.

Fig. 1 Magnetic resonance cholangiopancreatography (MRCP) showing bile duct stenosis (arrowheads) in the hepatic hilum extending to the common hepatic duct, suggestive of a hilar cholangiocarcinoma.

Video 1

Fig. 2 Endoscopic ultrasound (EUS) showing symmetrical and smooth wall thickening (thin arrows) of the hepatic hilum and CHD, and EUS-guided tissue acquisition using a 22-gauge needle (open arrow) was performed for the biliary lesion.

Fig. 3 Histology of endoscopic ultrasound-guided fine-needle aspiration/biopsy and SpyBite forceps biopsy: a stromal fibrosis with lymphoplasmacytic infiltration (H&E, ×200); b more than 10 IgG4-positive plasma cells per high-power field (IgG4 stain, ×400).
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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