IgG4-related sclerosing cholangitis presenting as an isolated intrahepatic stenosis: a rare presentation of a rare disease

A 36-year-old woman with no significant past medical history presented with fatigue and a 5-kg weight loss. Blood work-up showed isolated elevated γ-glutamyltransferase (3 × upper limit of normal). Magnetic resonance cholangiopancreatography (MRCP) demonstrated a short stricture of the left main intrahepatic duct (IHD) with upstream dilatation of the IHDs (▶Fig. 1). The biliary tract was otherwise unremarkable. An endoscopic retrograde cholangiopancreatography (ERCP) was performed with retrograde cholangioscopy. The endoscopic appearance of the stricture was worrisome, with irregular pattern and anarchic vascularization (▶Fig. 2; ▶Video 1). Different sampling techniques of the stricture, including brush cytology, bile aspiration, and multiple targeted biopsies, were used. Pathology showed non-specific signs of chronic inflammation. The patient’s serum IgG4 levels were non-significantly elevated (1.3 × normal). The case was discussed at a multidisciplinary meeting and it was decided to perform surgery, given the patient's weight loss, asthenia, and the suspicion of underlying neoplasia. A left hepatectomy with lymphadenectomy was performed. Final pathology showed lesions of sclerosing cholangitis at the level of the IHDs, with significant inflammation and IgG4 infiltration of the stenotic region, but no tumor cells (▶Fig. 3). A positron emission tomography (PET) scan showed no signs of involvement of other organs. The patient has had no further events during 1 year of follow-up. IgG4-related sclerosing cholangitis is an autoimmune biliary tract disease [1,2].

Amongst the IgG4-related diseases, bile duct and kidney involvement are usually associated with manifestations in other organs, specifically the pancreas [3]. The lower bile duct is the most commonly involved [4]. Cholangioscopy has a high specificity and fair level of sensitivity for the diagnosis of cholangiocarcinoma [5] and can therefore be helpful in differentiating malignant from benign strictures [4]. This case illustrates how challenging the diagnosis of an indeterminate isolated IHD stricture is. Endoscopic characterization of IgG4 sclerosing cholangitis lacks sensitivity. Furthermore, detection of IgG4-positive plasma cells in cholangioscopy-targeted biopsies remains difficult.

Endoscopy_UCTN_Code_CCL_1AZ_2AZ

▶Fig. 1 Radiographic images showing: a on magnetic resonance cholangiography, a marked stenosis of the left main hepatic duct (arrow) with dilatation of left intrahepatic ducts (the right intrahepatic ducts and common bile duct are normal); b on gadolinium-enhanced T1-weighted magnetic resonance imaging, dilatation of left intrahepatic ducts with contrast enhancement of the biliary ducts walls (arrows).

▶Fig. 2 Cholangioscopic view of the left main intrahepatic duct stricture showing an irregular vascularity and pattern.
Competing interests

Marine Camus is a consultant for Boston Scientific and Cook Medical. Ulriikka Chaput is a consultant for Boston Scientific. The remaining authors declare that they have no conflict of interest.

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Endoscopy 2021; 53: E205–E206
DOI 10.1055/a-1244-9065
ISSN 0013-726X
published online 11.9.2020
© 2020. Thieme. All rights reserved.
Georg Thieme Verlag KG, Rüdigerstraße 14, 70469 Stuttgart, Germany

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Video 1 Cholangioscopy showing an isolated left main intrahepatic duct stricture with worrisome features.

Fig. 3 Histopathological appearance of the resected tissue showing: a bile duct with periductal fibrosis and plasma cell-rich inflammation on hematoxylin and eosin (H&E) staining; b immunostaining with IgG4 highlighting numerous IgG4-positive plasma cells (>50 per high power field).