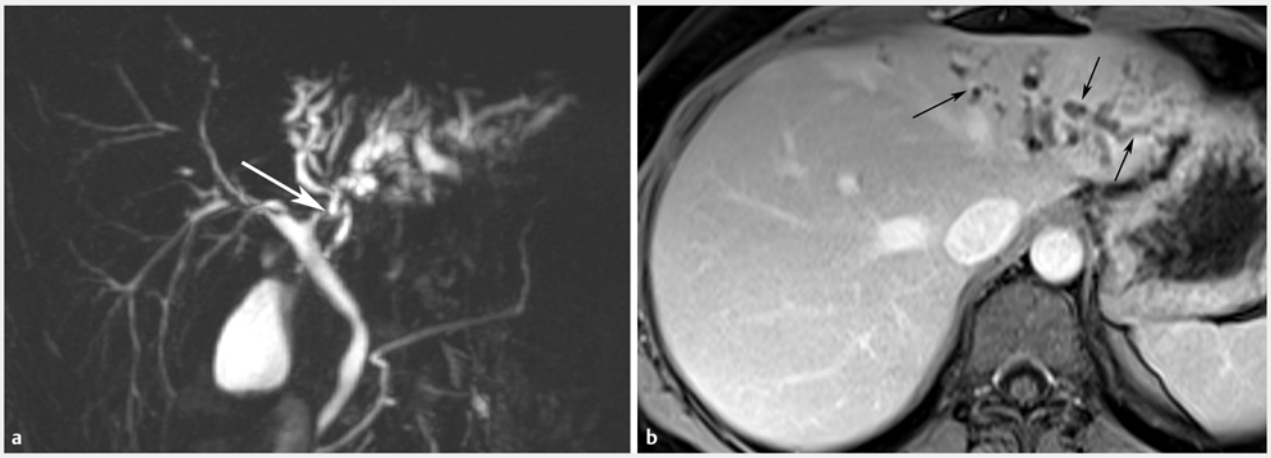
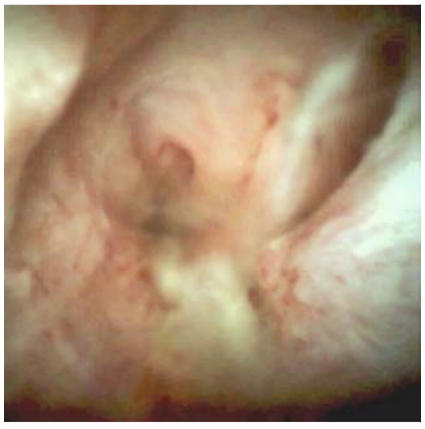
IgG4-related sclerosing cholangitis presenting as an isolated intrahepatic stenosis: a rare presentation of a rare disease

Aymeric Becq, Marine Camus, Lionel Arrivé, Thevy Hor, Doryan Amoodadashi, Louis Buscail, Ulriikka Chaput

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



▶ **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.

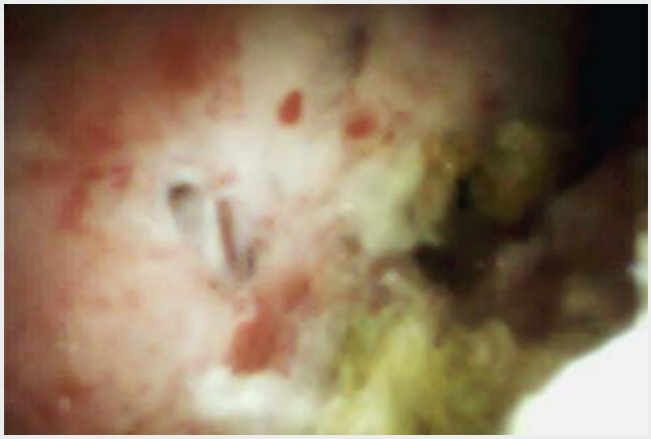
A 36-year-old woman with no significant past medical history presented with fatigue and a 5-kg weight loss. Blood work-up showed isolated elevated γ-glutamyl-transferase (3 × upper limit of normal). Magnetic resonance cholangiopancreatography (MRCP) demonstrated a short stricture of the left main intrahepatic 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.

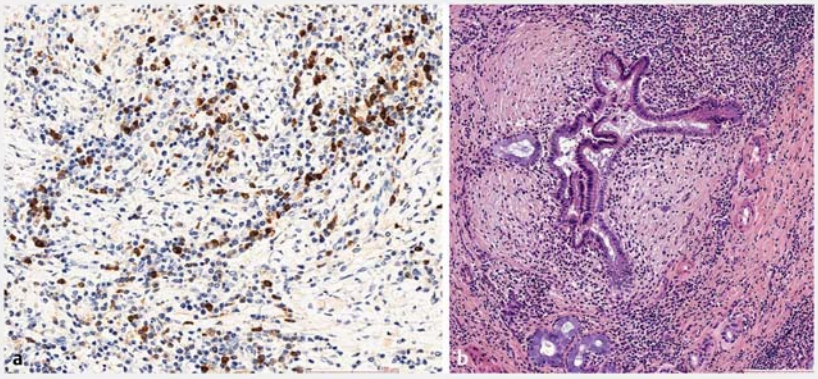
IgG4-related sclerosing cholangitis is an autoimmune biliary tract disease [1, 2]. Amongst the IgG4-related diseases, bile duct and kidney involvement are usually associated with manifestations in other organs, specifically the pancreas [3]. The lower bile duct is the most commonly involved [4]. Cholangioscopy 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



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



▶ **Fig. 3** Histopathological appearance of the resected tissue showing: **a** 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).

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.

The authors

Aymeric Becq[1,2], Marine Camus[1,2], Lionel Arrivé[3], Thevy Hor[4], Doryan Amoodadashi[5], Louis Buscail[6], Ulriikka Chaput[1,2]

1 Department of Endoscopy, Saint-Antoine Hospital, Paris, France
2 Sorbonne University, AP-HP, Saint-Antoine Hospital, Paris, France
3 Department of Radiology, Saint-Antoine Hospital, Paris, France
4 Department of Digestive Surgery, Saint-Antoine Hospital, Paris, France
5 Department of Pathology, Saint-Antoine Hospital, Paris, France
6 Department of Gastroenterology and Pancreatology, CHU Rangueil, University of Toulouse III, Toulouse, France

Corresponding author

Aymeric Becq, MD
Department of Endoscopy, Saint Antoine Hospital, 184 Rue du Faubourg, Saint Antoine, Paris 75012, France
Fax: +33-1-49282970
aymeric.becq@aphp.fr

References

[1] Lee CW, Ronnekleiv-Kelly S. Autoimmune diseases of the biliary tract: a review. Surg Clin North Am 2019; 99: 185–201
[2] Culver EL, Chapman RW. IgG4-related hepatobiliary disease: an overview. Nat Rev Gastroenterol Hepatol 2016; 13: 601–612
[3] Inoue D, Yoshida K, Yoneda N et al. IgG4-related disease: dataset of 235 consecutive patients. Medicine (Baltimore) 2015; 94: e680
[4] Goodchild G, Pereira SP, Webster G. Immunoglobulin G4-related sclerosing cholangitis. Korean J Intern Med 2018; 33: 841–850
[5] Navaneethan U, Hasan MK, Lourdusamy V et al. Single-operator cholangioscopy and targeted biopsies in the diagnosis of indeterminate biliary strictures: a systematic review. Gastrointest Endosc 2015; 82: 608–614.e2

Bibliography

Endoscopy 2021; 53: E205–E206
DOI 10.1055/a-1244-9065
ISSN 0013-726X
published online 11.9.2020

Georg Thieme Verlag KG, Rüdigerstraße 14, 70469 Stuttgart, Germany