



HAL
open science

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

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

► To cite this version:

Aymeric Becq, Marine Camus, Lionel Arrivé, Theyvy Hor, Doryan Amoodadashi, et al.. IgG4-related sclerosing cholangitis presenting as an isolated intrahepatic stenosis: a rare presentation of a rare disease. *Endoscopy*, 2020, 53 (06), pp.E205 - E206. 10.1055/a-1244-9065 . hal-03254189

HAL Id: hal-03254189

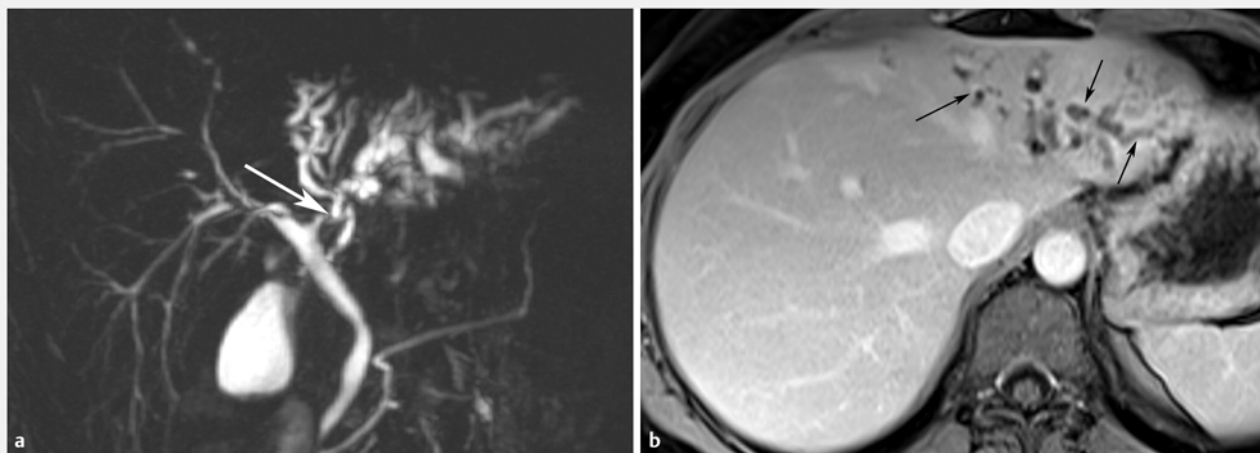
<https://hal.sorbonne-universite.fr/hal-03254189>

Submitted on 8 Jun 2021

HAL is a multi-disciplinary open access archive for the deposit and dissemination of scientific research documents, whether they are published or not. The documents may come from teaching and research institutions in France or abroad, or from public or private research centers.

L'archive ouverte pluridisciplinaire **HAL**, est destinée au dépôt et à la diffusion de documents scientifiques de niveau recherche, publiés ou non, émanant des établissements d'enseignement et de recherche français ou étrangers, des laboratoires publics ou privés.

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 ($3\times$ 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.

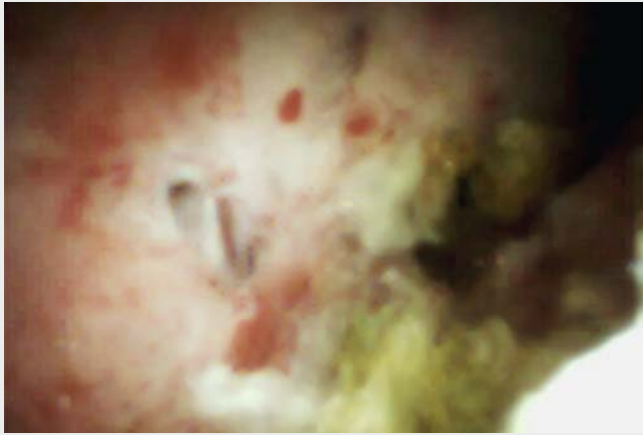
An endoscopic retrograde cholangiopancreatography (ERCP) was performed with retrograde cholangioscopy. The endoscopic appearance of the stricture was worrisome, with irregular pattern and anarchic vascularization (► **Fig. 2**; ► **Video 1**). Different sampling techniques of the stricture, including 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 ($1.3\times$ 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.

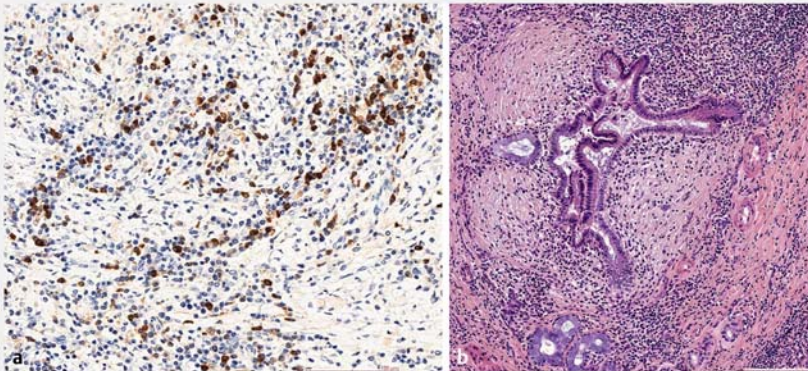
IgG4-related sclerosing cholangitis is an autoimmune biliary tract disease [1,2]. Amongst the IgG4-related diseases, bile duct and kidney involvement are usually associated with manifestations in other organs, specifically the pancreas [3]. The lower bile duct is the most commonly involved [4]. Cholangioscopy 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 Arrivé³, Theyy Hor⁴, Doryan Amodadashi⁵, Louis Buscail⁶, 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, Lourdasamy 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

© 2020, Thieme. All rights reserved.

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

ENDOSCOPY E-VIDEOS

<https://eref.thieme.de/e-videos>



Endoscopy E-Videos is a free access online section, reporting on interesting cases and new techniques in gastroenterological endoscopy. All papers include a high quality video and all contributions are freely accessible online.

This section has its own submission website at
<https://mc.manuscriptcentral.com/e-videos>