

An unusual cause of extrahepatic cholestasis associated with solid liver lesions: a case report

L. Caverenne¹, L. Weichselbaum¹, M. Van Hoof¹, P. Deltenre^{1,2}

(1) Department of Gastroenterology and Hepatology, Clinique St Luc, Bouge, Belgium; (2) Department of Gastroenterology, Hepatopancreatology, and Digestive Oncology, C.U.B. Hôpital Erasme, Université Libre de Bruxelles, Brussels, Belgium.

Abstract

IgG4-related sclerosing cholangitis is a special type of cholangiopathy often associated with autoimmune pancreatitis. In this article, we report an unusual case of IgG4-SC limited to the common hepatic duct and associated with pseudo tumoral liver lesions, but without evidence of pancreatic involvement. Corticosteroid therapy was rapidly effective and allowed normalization of liver tests. (*Acta gastroenterol. belg.*, 2023, 86, 490-492).

Keywords : IgG4-related sclerosing cholangitis, cholangiocarcinoma.

Abbreviations: IgG4-SC, IgG4-related sclerosing cholangitis.

Clinical case

A 55-year-old man with a history of moderate mental retardation, dyslipidemia and hypertension treated with fenofibrate, quinapril, bisoprolol and hydrochlorothiazide presented to the emergency department reporting several days of abdominal pain, nausea and jaundice. One month before admission, he had an episode of spontaneously resolving cholestasis. At this time, abdominal ultrasound showed two solid lesions of the hepatic parenchyma with no sign of biliary tract obstruction (Fig 1). At admission, he had normal vital parameters and jaundice. Biological analyses showed cholestasis associated with a moderate inflammatory syndrome. AST and ALT were measured at 248 and 363 U/L, respectively. Alkaline phosphatase was at 104 U/L. Bilirubin level was 10.67 mg/dL (Fig. 2). Serum lipase was normal.

Abdominal CT scan and MRI showed dilation of central intrahepatic bile ducts, hypervascularization of the common bile duct wall (Fig. 3) and two solid lesions in the liver (Fig. 4). There was no sign of pancreatitis. Echo-endoscopy confirmed the thickening of the common hepatic duct with a stone visible upstream. During ERCP, 2 stones were extracted after dilation of the stenosis of the common hepatic duct (Fig. 5). A biopsy of the stricture was performed. Bile was purulent and antibiotic therapy was initiated. In spite of the stone removal, the patient's cholestasis kept increasing during the following days. A second ERCP was thus performed to insert a plastic stent over the stenosis of the common hepatic bile duct two days after the first ERCP, which led to a partial regression of cholestasis during the following week.

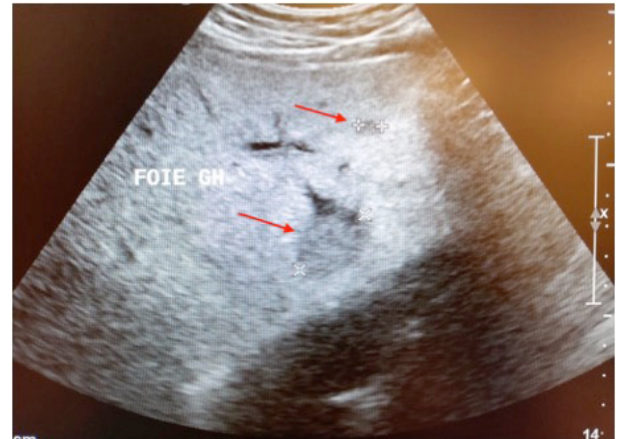


Figure 1. — Two liver lesions on the abdominal ultrasound performed one month before admission.

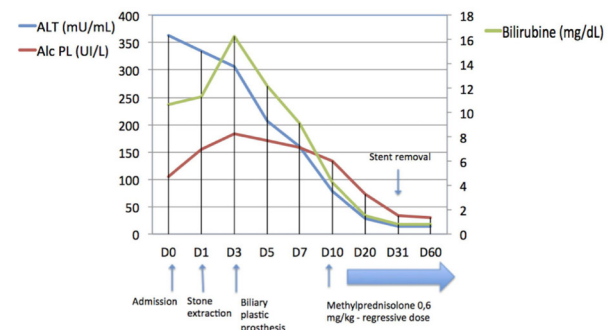


Figure 2. — Liver tests evolution by days in a patient with IgG4-SC at diagnosis and after treatment.

Histological analysis of the common hepatic duct stenosis showed subacute fibro-inflammatory remodeling with infiltrates of IgG4 plasma cells. There was no sign of malignancy. Serum IgG4 level was 1.5 g/L (ULN < 1.26 g/L).

Corticosteroids were started. Follow-up was unremarkable with a rapid regression of cholestasis within

Correspondence to: Pierre Deltenre, M.D., Ph.D., Department of Gastroenterology and Hepatology, Clinique St Luc, Bouge, Belgium.
Email: pierre.deltenre01@gmail.com

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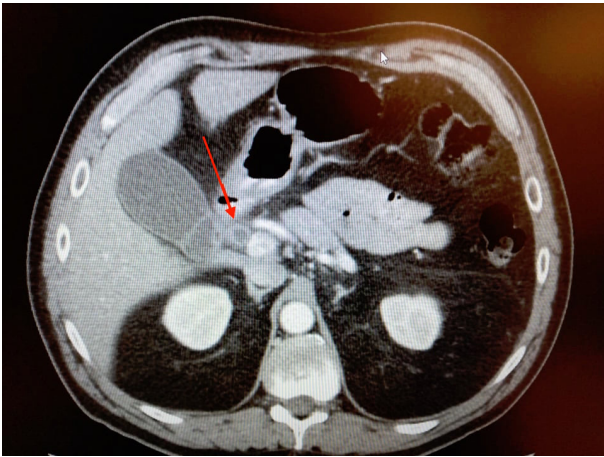


Figure 3. — Hypervascularization of the common bile duct wall on abdominal CT scan.

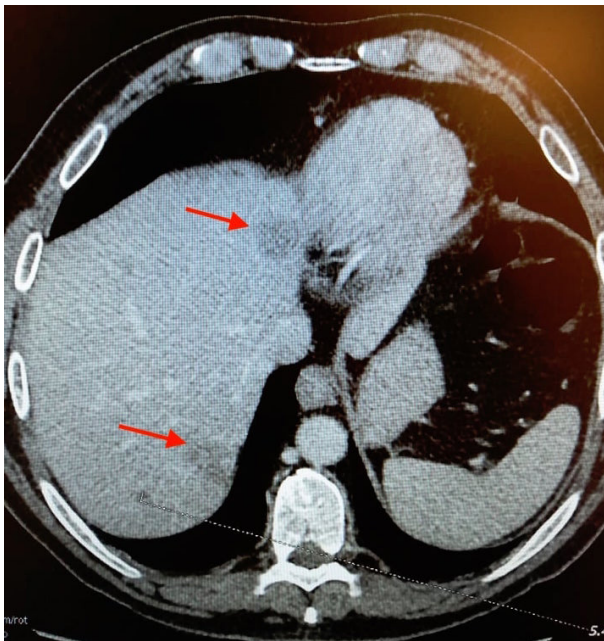


Figure 4. Hepatic lesions on abdominal CT scan.

10 days of the initiation of treatment (Fig. 2). Three weeks after the beginning of the corticosteroid therapy, the biliary stent was removed. At this time, bile duct opacification revealed a nearly total regression of the common hepatic duct stenosis. The cholestasis continued to regress with the treatment. However, hepatic lesions did not respond as quickly as the stenosis of the common hepatic duct. After discussion with the interventional radiologists, a biopsy of the lesions was not performed since their location did not allow a safe transparietal liver biopsy and given the rapid favorable evolution from a clinical, biological and radiological standpoint.

The biological follow-up remained stable one month after removal of the stent and two months after initiation of steroid therapy. A Follow-up liver MRI was scheduled but, unfortunately, the patient died in the mean time. Cause



Figure 5. — Stenosis of the common hepatic duct during ERCP.

of death was an out-of-hospital cardiac event unrelated to the autoimmune pathology or to the treatment.

Discussion

IgG4-related sclerosing cholangitis (IgG4-SC) is a cholangitis characterized by a lymphocytic and IgG4-positive plasma cell infiltration together with a fibrosis of the biliary wall and an elevation of serum IgG4 levels (1). It is frequently associated with autoimmune pancreatitis (AIP). Corticosteroid therapy is often rapidly effective and response to therapy can be used as a diagnostic test. Recent guidelines define four types of IgG4-SC based on the location of biliary duct lesions (2). Type 4 IgG4-SC involves only the common hepatic duct and the beginning of segmental branches and accounts for only 10% of IgG4-SC (3). A biopsy is often required to exclude a cholangiocarcinoma.

As indicated in recent guidelines, hepatic inflammatory IgG4 pseudotumors are often observed when the involvement of the biliary tree is limited to the common hepatic duct (2,4), as is the case here. A recent Japanese multicenter study showed that type 4 IgG4-SC was the most frequent subtype not associated with AIP, which is in line with what we observed in our patient (5).

Conclusion

Here, we reported a case of histologically proven IgG4-SC limited to the common hepatic duct associated

with probable IgG4-related liver lesions but without AIP. Clinical course was rapidly favorable after the initiation of corticosteroid therapy. Clinicians should be aware of this entity that mimics cholangiocarcinoma, especially when liver pseudotumors are associated.

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