An Unusual Presentation of IgG4-Related Disease mimicking cholangiocarcinoma

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Background

IgG4-related disease (IgG4-RD) is a fibroinflammatory systemic autoimmune disease.[1]

Affects pancreatico-hepatobiliary system presenting as sclerosing cholangitis, Type I AIP autoimmune, both, or rarely presents as IgG4-related hepatopathy.[2]

Here we present a challenging case of a patient with IgG4-RD.

Initial Presentation and Hospital Stay

43M presents with painless jaundice and diarrhea for 1 month. PE + for scleral icterus but negative for Murphy's sign.

Initial evaluation revealed ALP of 237 and total bilirubin of 10.4.

MRCP: Diffuse intrahepatic biliary dilation with obstruction at the confluence of the CHD

ERCP: Stenosis of the upper third of the right main bile duct. One 8.5 Fr by 15 cm transpapillary temporary stent was placed into the right hepatic duct. Biopsies showed normal ductal mucosa with no malignant cells.

Clinical Course

Patient experienced worsening jaundice, elevated CA-19-9 (3995), serum IgG levels (2582), and total bilirubin (6.7).

Repeat ERCP showed CHD stricture.

CT abdomen showed increased intrahepatic biliary ductal dilation and persistent soft tissue thickening at confluence of the right and left hepatic ducts.

EUS showed normal pancreatic parenchyma and a 3cm mass at the peripheral area.

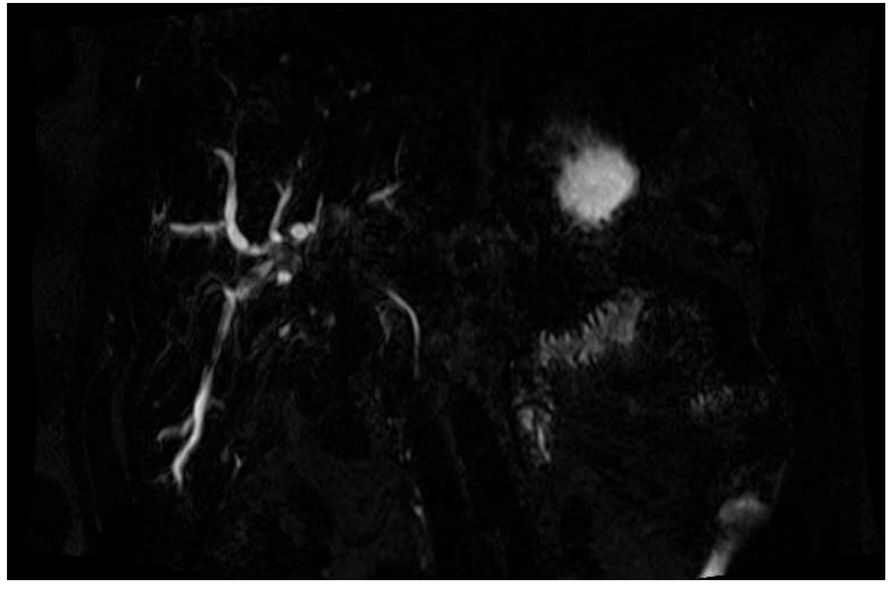
Multiple biopsies were negative for CCA.

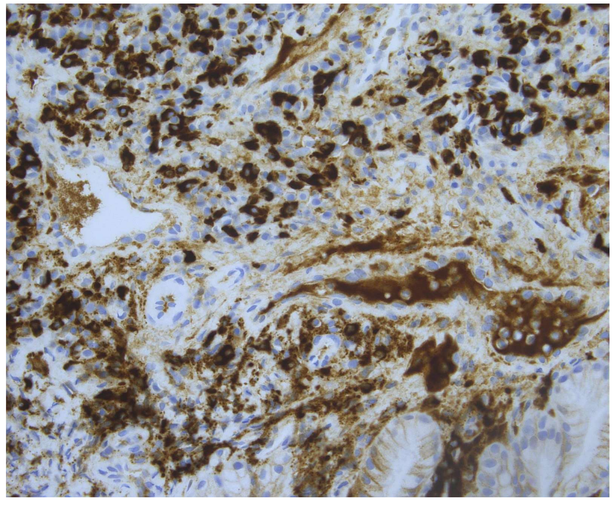
ERCP demonstrated nodular CHD, and biopsies of the biliary duct showed increased IgG4-positive plasma cells and an IgG4:IgG plasma cell ratio of 27%.

After re-examination, it revealed an area with >20 plasma cells positive for IgG4, consistent with IgG4-related disease.

The patient started on Prednisone 40mg once daily with a prolonged taper and improved significantly.

Figures 1 (MRCP Image Showing CHD Stricture) and Figure 2 (Histology)





Immunohistochemical staining for IgG4 in a high-power field showing markedly increased expression of IgG4 (up to 50 per high power field at 400X magnification).

Upper portion of image shows the positive plasma cells with cytoplasmic and membranous expression of IgG4.

Discussion

IgG4 sclerosing cholangitis is the most common extrapancreatic manifestation of Type I AIP (>70%).[3]

This case is suggestive of IgG4 sclerosing cholangitis without evidence of acute pancreatitis presenting as a hilar mass mimicking CCA.

While biopsies failed to identify a neoplastic or infectious agent, it wasn't until biopsies were stained with IgG4 revealing the etiology of the disease.

Be aware of the GI manifestations of IgG4-RD and consider it in the differential when addressing unrelenting symptomatic jaundice.

Early confirmation of >20 plasma cells HPF in biopsy avoids unnecessary procedures and allows for early treatment.

References

1. IgG4 Related Disease - StatPearls - NCBI Bookshelf. https://www.ncbi.nlm.nih.gov/books/NBK499825/.

2. Chen, L. Y. C., Mattman, A., Seidman, M. A. & Carruthers, M. N. IgG4-related disease: what a hematologist needs to know. *Haematologica* 104, 444–455 (2019).

3. Ideaki, H. *et al.* High Serum IgG4 Concentrations in Patients with Sclerosing Pancreatitis. *https://doi.org/10.1056/NEJM200103083441005* 344, 732–738 (2001).