A Treacherous Pain: IgG4-related disease relapsing as Sclerosing Cholangitis

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Introduction

- Immunoglobulin G4-related disease (IgG4-RD) is an immune-mediated fibroinflammatory condition resulting in tumor-like masses or organ enlargement from infiltration of IgG4-positive plasma cells.
- Elevated IgG4 serum levels can suggest disease, but tissue biopsy is gold standard for diagnosis.
- IgG4-sclerosing cholangitis (IgG4-SC) involves intrahepatic and extrahepatic bile ducts, is more common in men, and can present with obstructive jaundice.
- Systemic steroids are 1st line of treatment and are effective in inducing remission, but recurrent refractory cases are common.

Case Description

- A 59-year-old male with history of retroocular follicular hyperplasia secondary to IgG-4 related disease, diagnosed by tissue biopsy and on long term steroid therapy until 3 months prior who visited the ER complaining of right upper quadrant abdominal pain, acholia, nauseas and vomiting of 5 days in evolution. He denied new medications or supplements, use of NSAIDs, or travel. Physical exam was remarkable for palpable submandibular nodules and epigastric tenderness upon palpation. Liver chemistries were remarkable for an AST of 830, ALT of 380, total bilirubin of 3.3, and IgG4 serum levels at 1066 mg/dL. An abdominal US showed biliary tree dilatation with CBD >10 mm. An MRCP showed a dilated biliary tree, mucosal wall thickening at proximal to mid extrahepatic CBD with luminal narrowing, consistent with IgG4 related cholangitis. No choledocholithiasis evidenced. The patient was started on prednisone 40 mg daily during hospitalization. He was discharged home three days later to continue steroid therapy. A follow-up MRCP at one month showed a normal caliber common bile duct and interval resolution of mucosal thickening as well of the stenotic and dilated segments. Liver chemistries also normalized.

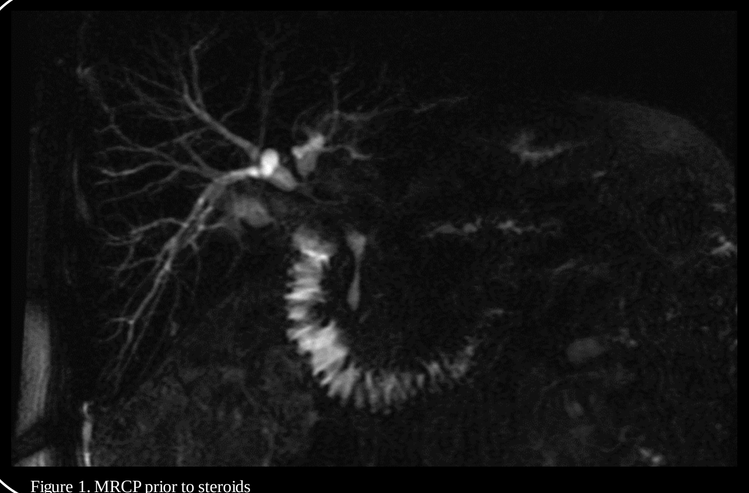

Figure 1. MRCP prior to steroids

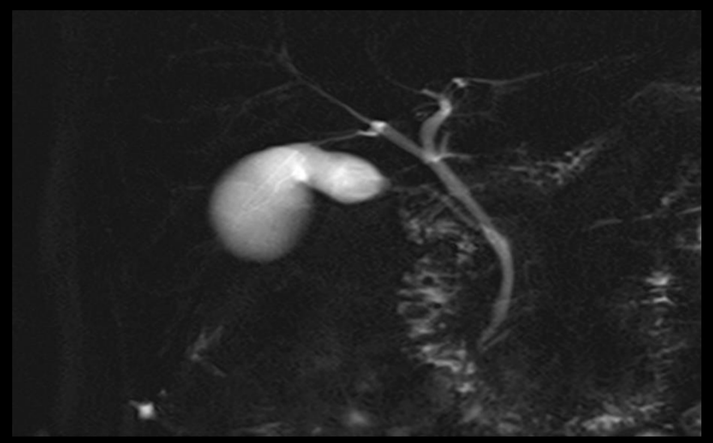

Figure 2. MRCP after one month of steroid therapy

	09/2020	02/2022	09/2022
IgG Subclass 4	39 mg/dL	1066 mg/dL	167 mg/dL

Discussion

- Patients with IgG4-RD respond well to steroids, but relapse is common while tapering or after their withdrawal.
- Studies show that 50% of IgG4-RD patients present with new organ involvement on relapse.
- This case emphasizes the need to be mindful of IgG4-RD relapse to new organ involvement even if the primary site is controlled.
- An elevated serum IgG4 may aid in the rapid recognition of recurrent disease and prompt initiation of steroids can reduce organ inflammation, preserve function, and limit multiorgan involvement.

References

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