Steroid- Refractory IgG4- Related Disease Presenting as Sclerosing Cholangitis

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INTRODUCTION

- IgG4-related disease (IgG4-RD) is characterized by tumefactive infiltration of IgG4-bearing plasmablasts and other lymphocytes, along with storiform fibrosis, into one or multiple organs, with resultant organ enlargement, fibrosis, and dysfunction.
- Although initial reports of IgG4-RD concerned involvement of the pancreas, other organs such as the biliary tract can be involved. We present a case of steroid refractory IgG4- RD sclerosing cholangitis (IgG4-SC).

CASE DESCRIPTION

- A 67-year-old man with hypertension presented with a two-week history of unintentional weight loss, painless jaundice, and pruritus. Exam showed jaundice and scleral icterus.
- Laboratory revealed elevated liver tests with an obstructive pattern, with ALP, GGT, & Bilirubin of 431 U/L, 438 IU/L, 17.6mg/dL, respectively.
- MRCP, ERCP, and subsequent EUS demonstrated diffuse pancreatic parenchymal hypo-echogenicity, long segment strictures at the distal common bile duct with proximal duct dilation, dilated common hepatic duct, and multiple segmental intrahepatic biliary duct strictures.
- Biliary sphincterotomy and stent placement resolved symptoms. Biopsy of the Ampulla of Vater revealed chronic inflammation with ulceration. Further work-up uncovered an elevated serum IgG4 of 266 mg/dL with a negative ANA, SMA, AMA, and tumor markers.
- He was then diagnosed with IgG4-SC and completed a course of prednisone. Relapse of symptoms prompted a repeat ERCP which uncovered biliary tree pruning and irregularity, necessitating biliary stent replacement.
- Due to poor tolerance with rituximab infusion, he was treated with Mycophenolate Mofetil which accomplished a year of disease quiescence.

IMAGING

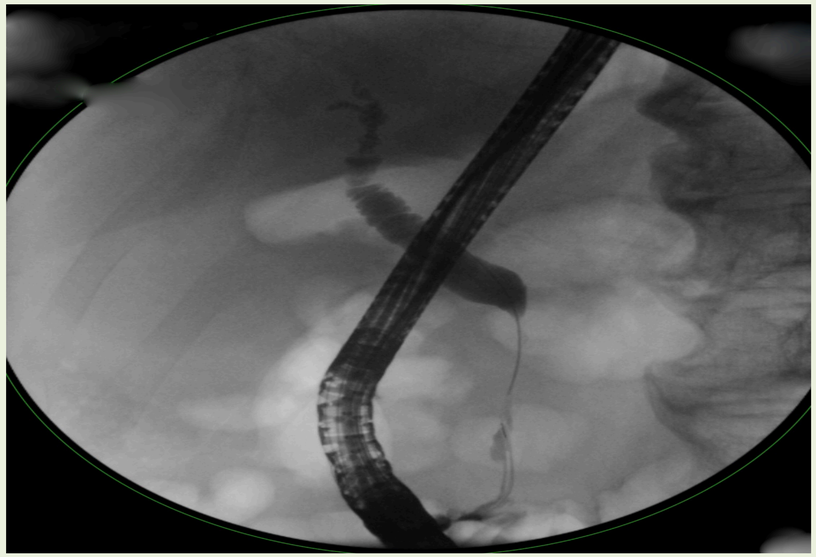

Figure 1. Long segment stricture

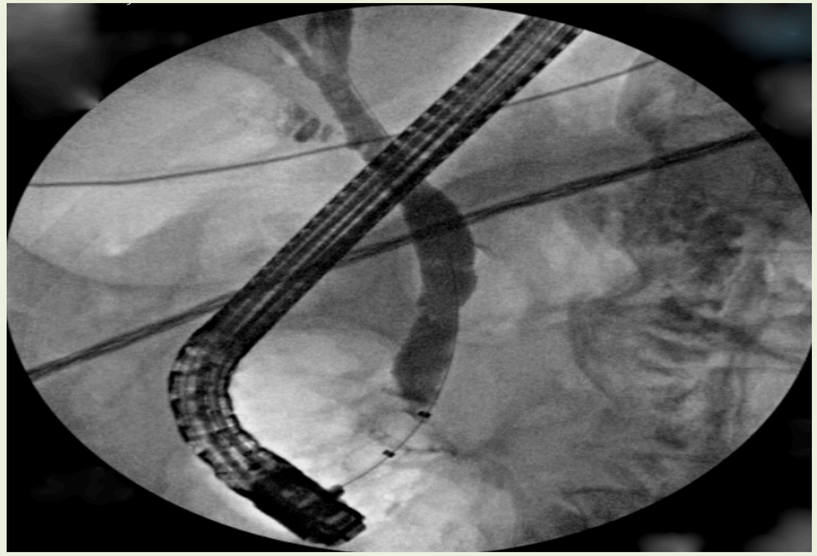

Figure 2. Resolved CBD stricture

DISCUSSION

- IgG4-related disease is a set of newly recognized fibroinflammatory conditions whose prime gastrointestinal manifestations include type 1 AIP and IgG4-SC and are often associated with each other.
- Urgent treatment with steroids is recommended to prevent infectious cholangitis and permanent fibrosis. Patients are at highest risk of relapse during steroid taper or within 6 months of completion, as seen in our case, with a relapse rate of 15-60%.
- Although our patient did not have the classic predictive factors of relapse, such as proximal strictures or serum IgG4 >280 mg/dL, he had a suboptimal response to steroids which prolonged his clinical course and exposed him to repeated procedures.
- Through this case we hope to highlight the need for further research in predictive factors and the role for possible combination therapy (steroids & immunosuppressive therapy) to prevent relapse in high-risk individuals.

REFERENCES

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