The Difficult case of Elevated liver Chemistries and Biliary Ductal Dilatation: IgG4-seronegative Autoimmune Cholangiopathy with Autoimmune Pancreatitis

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INTRODUCTION

- IgG4-sclerosing cholangitis (SC) is a type of steroid responsive SC frequently associated with autoimmune pancreatitis (AIP).
- Diagnosis of IgG4-SC is challenging as its presentation can mimic primary sclerosing cholangitis (PSC), cholangiocarcinoma, and pancreatic cancer.
- IgG4-seronegative autoimmune cholangiopathy (AIC) with pancreatic involvement is even rarer, with only 4 prior cases reported.

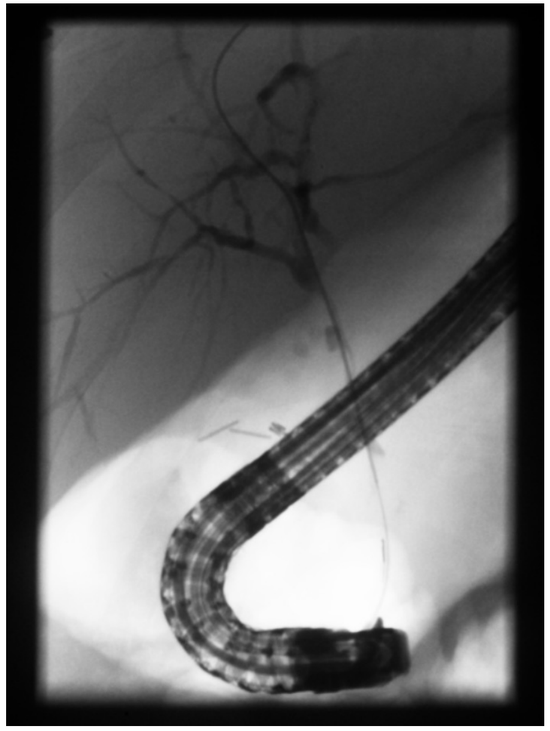

Figure 1: ERCP fluoroscopy image demonstrating long bile duct stricture

CASE DESCRIPTION

- A 27-year-old man presented in 2011 with 4 months of bloating. Infectious work-up was negative but liver tests were elevated (aspartate aminotransferase 40U/L, alanine transaminase 57U/L, alkaline phosphatase 158 U/L). Magnetic resonance imaging (MRI) abdomen revealed mild intrahepatic and common bile duct thickening without dominant stricture. He was lost to follow-up until 2017, when he presented with fever concerning for cholangitis. Endoscopic retrograde cholangiopancreatography (ERCP) identified severe ductopenia characterized by thin-caliber, irregular, beaded ducts with alternating segments of mild stenosis and dilation without dominant stricture. Liver biopsy was negative for autoimmune hepatitis and IgG4-related liver disease. Total IgG, IgG4, anti-mitochondrial and anti-smooth muscle antibodies were normal.
- Over the next 3 years he had multiple admissions for cholangitis with bacteremia. Imaging demonstrated a common hepatic stricture, leading to multiple ERCPs (**Figure 1**) requiring stent placement and exchanges. Due to frequent recurrent cholangitis in the setting of PSC, he was listed for liver transplantation with intent for live donor transplant. Repeat serum IgG4 testing was normal. In early 2021, surveillance MRCP revealed a sharply demarcated T2 moderately hyperintense, T1 hypointense appearance of the distal 4 cm of the pancreatic tail with early hypoenhancement and delayed hyperenhancement suggestive of inflammation and fibrosis. There was also marked periductal enhancement of the common hepatic duct with long segment stenosis consistent with acute on chronic SC (**Figure 2**). These imaging findings were suggestive of IgG4-seronegative autoimmune cholangiopathy with autoimmune pancreatitis. Endoscopic ultrasound with fine needle biopsy failed to obtain sufficient tissue sample. Given the findings, the patient was placed on prednisone 40 mg daily.
- Over the following 5 months, the patient experienced resolution of episodes of cholangitis and liver test normalization. There was mild improvement on MRI after 3 weeks of corticosteroids. Recent ERCP showed his biliary tree strictures had markedly improved. All biliary stents were removed, and the patient has tapered to low dose prednisone and remains on azathioprine.

WORKS CITED

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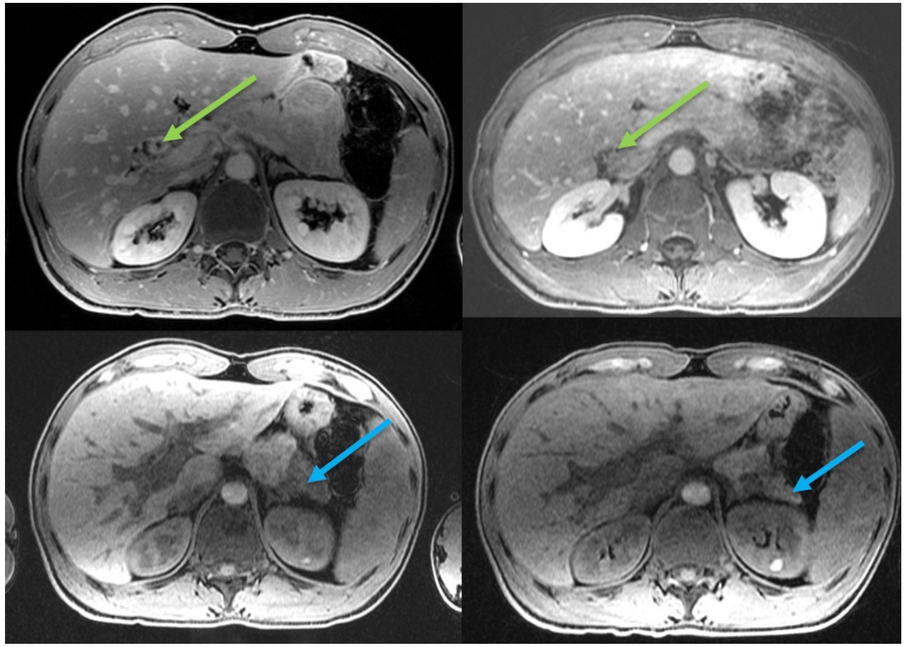

Figure 2: Left, top: Delayed post-contrast T1 weighted fat-suppressed image demonstrates marked peribiliary thickening and enhancement around the common hepatic duct (**green arrow**). Left, bottom: Axial T1 weighted pre-contrast fat-suppressed image reveals sharply demarcated abnormal hypointense T1 signal in the pancreatic tail (**blue arrow**). Right top: Delayed post-contrast T1 weighted fat-suppressed image obtained 6 months after initiation of steroids demonstrates resolution of peribiliary thickening and enhancement around the common hepatic duct (**green arrow**). Right, bottom: Delayed post-contrast T1 weighted fat-suppressed image obtained 6 months after initiation of steroids shows normalization of T1 signal in the pancreatic tail (**blue arrow**)

CONCLUSIONS

- IgG4-seronegative AIC and AIP can be difficult to diagnose due to its ability to mimic PSC and lack of IgG4 on serology, and often remains a diagnosis of exclusion.
- Given its responsiveness to steroids, it is important to maintain awareness of this condition to help patients avoid unnecessary liver transplantation or surgery.