A Case of IgG4 Sclerosing Cholangiopathy After Cholecystectomy

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CASE PRESENTATION

HISTORY

- An 83-year-old Caucasian male presented to clinic for anorexia, abdominal pain, and elevated liver function tests (LFTs).
- The patient had a history of cholelithiasis and was status-post cholecystectomy eight months prior, but otherwise had no history of elevated liver enzymes.
- Upon presentation to clinic, his AST was 205, ALT 143, and alkaline phosphatase 253, with normal bilirubin.
- Autoimmune work-up as well as serologies for Hepatitis A, B, and C were negative.
- Serum alpha fetoprotein, alpha-1-antitrypsin, ceruloplasmin and ferritin levels were in the normal range.

FINDINGS

- Magnetic resonance cholangiopancreatography (MRCP) revealed intrahepatic bile duct dilation, but normal common bile duct.
- Endoscopic retrograde cholangiopancreatography (ERCP) revealed strictures and dilations consistent with a sclerosing cholangitis (Figure 1).
- Initially, there was a concern for an ischemic cholangiopathy secondary to the prior cholecystectomy, however his IgG4 returned at 1140 mg/dl, specific for IgG4-SC.
- The patient was started on high-dose prednisone.

CLINICAL COURSE

- Repeat MRCP showed improvement of biliary ductal dilation and chronic IgG mediated pancreatic inflammation.
- Patient had resolution of anorexia, abdominal pain, and his liver function tests and IgG4 downtrended.

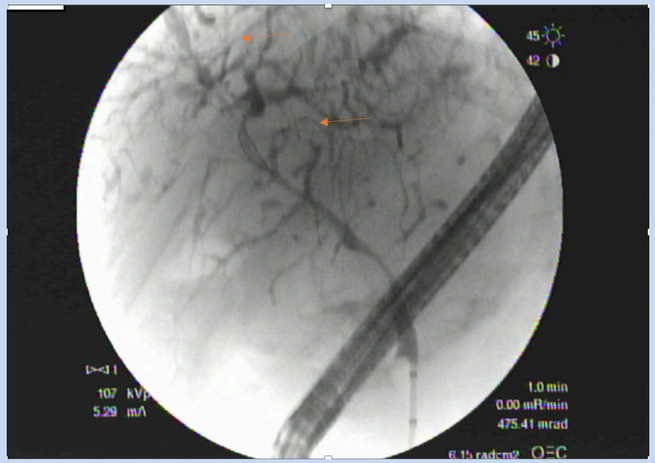

Figure 1: Intrahepatic duct dilation with strictures proximally (red arrows).

DISCUSSION

- IgG4-SC is a chronic inflammatory disease of the biliary system that typically occurs in association with other manifestations of IgG4-related disease.
- Most patients present in their 7th or 8th decade of life and the disease has a male predominance.
- The presenting symptoms include jaundice, pruritis, abdominal pain, and weight loss. IgG4-SC is diagnosed via a combination of imaging, laboratory, serological, and histopathological findings.
- When IgG4 levels are greater than 250 mg/dl, the specificity of IgG4-SC is 90%. IgG4-SC can be differentiated from primary sclerosing cholangitis with elevated serum levels and steroid responsiveness.
- Rarely is a tissue diagnosis necessary.
- The mainstay of treatment is corticosteroids with patient response shown by normalization of LFT's, reduction in serum IgG4 levels and improvement in imaging.
- Relapse can occur and is typically treated with immunomodulators.