A Case of IgG4-related sclerosing cholangitis in a patient with chronic pancreatitis

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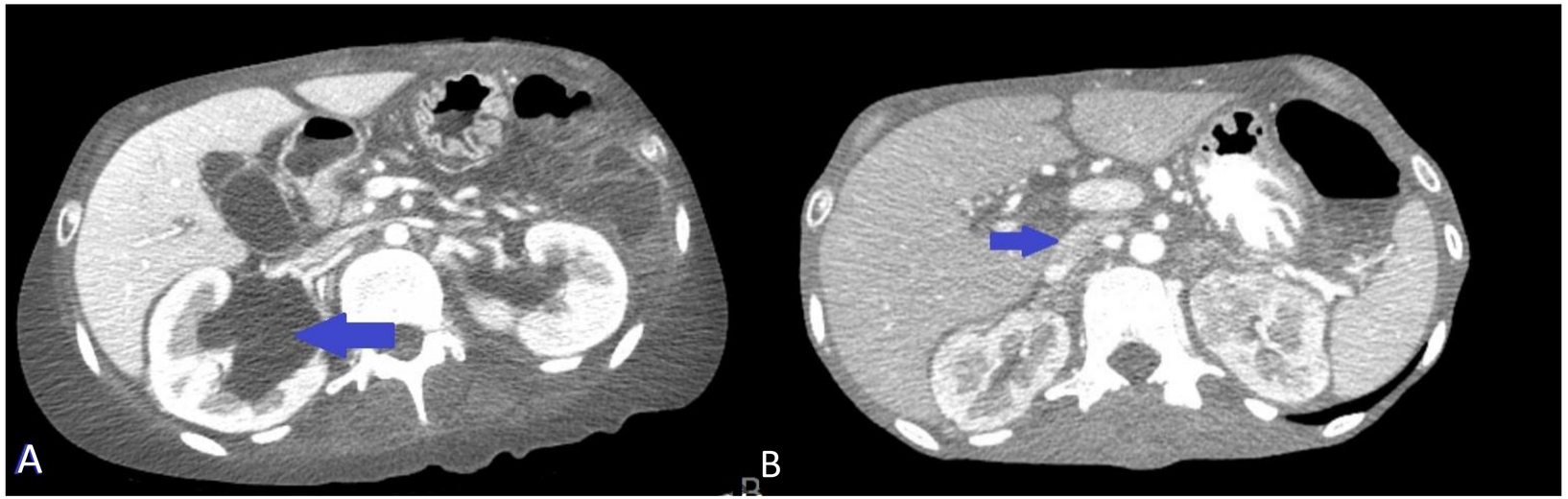
Introduction

Immunoglobulin G4-related disease (IgG4-RD) is an immune-mediated fibroinflammatory condition that is capable of affecting multiple organs [1,2]. IgG4-related sclerosing cholangitis is a form of sclerosing cholangitis that is clinically different from primary sclerosing cholangitis (PSC) and occurs usually as a part of the IgG4-RD. Additionally, it is also the most common extrapancreatic manifestation of type 1 AIP (IgG4-related) and is present in over 70% of such patients [3]. It rarely occurs in the absence of pancreatitis.

Case Presentation

We present a case of a 43-year-old female patient with medical history of chronic pancreatitis and secondary bacterial peritonitis status post exploratory laparotomy and gastrostomy tube (G tube) insertion who presented with the complaint of abdominal pain and leakage around the G tube site. Computed tomography abdomen and pelvis was suggestive of mild intrahepatic ductal dilatation. Laboratory results showed elevated ALP (>1200) and GGT levels. Ultrasound abdomen showed gallstones with sludge and bilateral hydronephrosis. Magnetic resonance cholangiopancreatography (MRCP) was unremarkable. Further workup showed elevated IgG4 levels (143 mg/dL). The autoimmune workup (ASMA, AMA, Anti-cardiolipin, LKMA) were negative. Patient had normal A1AT levels and viral hepatitis panel was negative. Patient underwent a liver biopsy which showed findings suggestive of chronic hepatitis with mild activity, secondary mild hemosiderosis and the presence of lymphoplasmacytic inflammation. IgG4 immunostaining highlighted scattered lgG4 positive plasma cells in portal tracts with up to 3 lgG4 positive plasma cells/ HPF.



A. CT Abdomen showing Bilateral Hydronephrosis (Right more than the left).
B. CT Abdomen showing Pancreatic Calcifications with findings suggestive of Chronic Pancreatitis

Discussion

Treatment is usually initiated with prednisone (0.6 mg/kg/day) and then tapered to discontinuation over a two-month period. Rituximab is generally considered in patients who do not respond to up to 40 mg/day of prednisone or cannot be tapered to <5 mg daily as well as in patients who have strong relative contraindications to glucocorticoid therapy in these doses. Prognosis of this condition is not well established. In some cases, spontaneous improvement can be seen, however disease often recurs without the treatment. Most patients respond to initial therapy with glucocorticoids, but relapses are not uncommon once therapy is discontinued.

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References

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