The Importance of Cholangioscopic Biopsies: An IgG4-Related Sclerosing Cholangitis Story

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ABSTRACT

Introduction: IgG4 related sclerosing cholangitis can be easily confused with cholangiocarcinoma and secondary sclerosing cholangitis and should be on the differential for biliary strictures.

Case presentation: 68 year old male presented for abdominal pain, loss of appetite, weight loss, and elevated liver function tests. Abdomen CT, MRCP, and EUS were consistent with intrahepatic biliary ductal dilation. Cholangioscopy showed fibrotic intraluminal growth in the upper third of common bile duct, and stricture at bifurcation and left intrahepatic duct. Initial cholangioscopic brushings were negative. Cholangioscopic biopsy showed IgG4 cells consistent with IgG4 related sclerosing cholangitis. Patient was started on corticosteroids with adequate response.

Discussion: Our patient presented with obstructive jaundice and isolated dilatation of the biliary tree with a normal serum IgG4 (elevated in about 75% of patients with IgG4-SC) suggestive of underlying malignancy, but also had elevation of total IgG leading to persistent suspicion for an underlying autoimmune disorder. Biliary brushings can be used to exclude malignancy, but generally are not sufficient to diagnose IgG4 disease as its sensitivity is only 36%. The most useful modality of diagnosis is cholangioscopic biopsy as this is positive in about 88% of bile duct biopsy specimens.

Conclusion: Thus, IgG4 sclerosing cholangitis should be considered in patients with an atypical presentation of obstructive jaundice with incongruent lab and imaging findings as this condition is treatable.

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INTRODUCTION

IgG4 related sclerosing cholangitis has generally been associated with autoimmune pancreatitis, retroperitoneal fibrosis, and sialadenitis. However, very few cases are isolated to the biliary tree. In addition, IgG4 related sclerosing cholangitis can be easily confused with cholangiocarcinoma and secondary sclerosing cholangitis. Particular detail should be paid to the typical subset of patients and clinical manifestations of the disease including patients who are elderly males who have obstructive jaundice. IgG4 serum levels more than 135 generally suggest IgG4 related sclerosing cholangitis but lower IgG4 levels do not rule out the disease. Thus, here we present an atypical case of IgG4 related sclerosing cholangitis.

CHOLANGIOSCOPY IMAGES

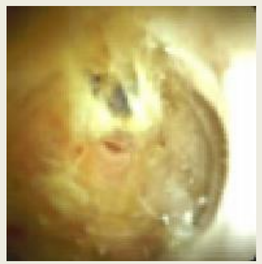

Image 1: Common Bile Duct Stricture

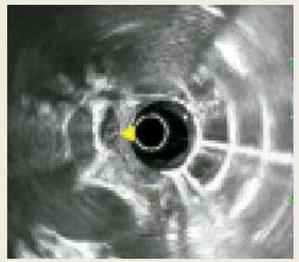

Image 2: Intraluminal Growth in Common Bile Duct

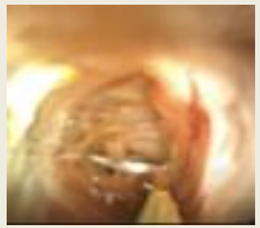

Image 3: Fibrotic Intraluminal Growth

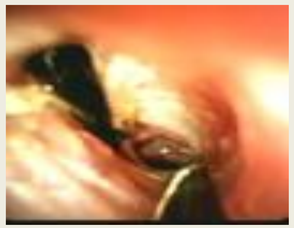

Image 4: Common Hepatic Duct Stone

CASE PRESENTATION

68 year old male with past medical history of gallbladder rupture secondary to trauma and alcohol misuse disorder presented for pruritis, scleral icterus, intermittent vague diffuse abdominal pain, loss of appetite, and 40 pounds of unintentional weight loss for 3 weeks. On admission, AST was 158, ALT was 81, ALP was 713, total bilirubin was 5.8, and direct bilirubin was 4.9. Hepatitis panel was negative, IgG was 3203, IgG4 was 113, ANA was positive 1:40, but rest of autoimmune work up was negative. Abdominal CT and MRCP showed isolated left intrahepatic biliary dilation. EUS/ERCP/cholangioscopy was performed showing fibrotic intraluminal growth in upper third of the common bile duct, long stricture at the bifurcation, and short stricture at left intrahepatic duct. Due to difficulty positioning the side-viewing scope, the cholangioscope was unable to be advanced beyond the initial stricture. Biliary brushings done only showed acute inflammation. A plastic biliary stent was placed for decompression. Due to concern for cholangitis from purulent material drained from the common bile duct, patient was started on meropenem. Liver biopsy was then performed showing benign liver tissue and stage 3 bridging fibrosis with severe cholestasis but was negative for malignancy or autoimmune process. A few weeks later, percutaneous internal-external biliary drain was placed by interventional radiology to facilitate subsequent rendez-vous ERCP. Repeat cholangioscopy showed scarring and fibrosis of the entire biliary tree and segmental stricturing at the bifurcation and left intrahepatic duct. Cholangioscopic forceps biopsies performed showed more than ten IgG4 cells per high powered field indicating IgG4-SC. Patient was discharged with close follow up with hepatobiliary surgery and started on corticosteroids with adequate response.

DISCUSSION

Awareness of IgG4-SC is important as it can mimic cholangiocarcinoma and primary sclerosing cholangitis. Our patient was part of the typical subset of patients of IgG4-SC including elderly men and those with obstructive jaundice. But he had multiple atypical features of disease, as well, including isolation of IgG4 disease to the biliary tree and serum IgG4 less than 135. Our patient presented with obstructive jaundice and isolated dilatation of the biliary tree with a normal serum IgG4 (elevated in about 75% of patients with IgG4-SC) suggestive of underlying malignancy, but also had elevation of total IgG leading to persistent suspicion for an underlying autoimmune disorder. Biliary brushings can be used to exclude malignancy, but generally are not sufficient to diagnose IgG4 disease as its sensitivity is only 36%. The most useful modality of diagnosis is cholangioscopic biopsy as this is positive in about 88% of bile duct biopsy specimens.

CONCLUSIONS

IgG4 sclerosing cholangitis should be considered in patients with biliary tree strictures as this condition is treatable with corticosteroids. In addition, biopsies of the biliary tree should be taken due to atypical clinical, endoscopic, and lab findings that can obscure the diagnosis.

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