

CASE REPORTS

Early primary sclerosing cholangitis – a challenging diagnosis

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Received: May 4, 2020

Accepted: June 28, 2020

Online Published: July 14, 2020

DOI: 10.5430/crim.v7n3p5

URL: <https://doi.org/10.5430/crim.v7n3p5>

ABSTRACT

Primary sclerosing cholangitis (PSC) is a rare autoimmune disorder of the biliary system. PSC is fulminant in its course and leads to jaundice and biliary cirrhosis with specific radiological findings of the beading of larger ducts and pruning of smaller ducts. Though cholangiogram is the gold standard for PSC diagnosis, it may be inconclusive in patients with an early disease or small duct PSC or overlap with autoimmune hepatitis. These conditions may require a liver biopsy for confirmation of the underlying pathology. In this case report, we discuss one of these rare instances when a 24-year-old male was admitted with jaundice. During hospitalization, extensive workup, including cholangiograms, remained inconclusive, but based on a high suspicion of PSC, the patient underwent a liver biopsy, which confirmed our diagnoses.

Key Words: Primary sclerosing cholangitis, Autoimmune biliary Disorder, PSC

1. INTRODUCTION

Primary sclerosing cholangitis is a chronic progressive disorder of unknown etiology which is characterized by inflammation, fibrosis, and stricturing of bile ducts in the intrahepatic and extrahepatic biliary tree.^[1,2] PSC has an incidence of approximately 1 case per 100,000 person-years, with 70% being men. It holds a strong association with inflammatory bowel disease and is a severe risk factor for cholangiocarcinoma.^[3] The diagnosis of PSC is typically established by the demonstration of characteristic multifocal stricturing and dilation of intrahepatic and extrahepatic bile ducts on cholangiography. Endoscopic retrograde cholangiopancreatography remains the gold standard for diagnosing PSC, and the only definite treatment is liver transplantation.^[4]

2. CASE PRESENTATION

A 24-year-old male with no past medical history presented to the hospital complaining of less than a week of jaundice, nausea, vomiting, right upper quadrant discomfort, which progressively worsened. The patient denied any recent illness, fever, medication use, or drug/alcohol abuse. The patient had a total bilirubin level of 8.4, direct bilirubin of 5.6, alkaline phosphatase of 208, normal AST/ALT, and INR of 1.6. Acetaminophen, salicylate, lactate dehydrogenase, and ferritin levels were negative. Infectious workup for acute hepatitis panel, EBV antibody, heterophile antibody, HSV-1/HSV-2 antibody, Cytomegalovirus IgM, HIV, urine Chlamydia and Gonorrhea were all negative as well. The patient had a normal right upper quadrant ultrasound and a contrast-enhanced

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CT scan of the abdomen and pelvis. Further evaluation with MRCP was also nondiagnostic. Meanwhile, total bilirubin, direct bilirubin, alkaline phosphatase peaked at 14.4, 9.2, and 480, respectively. The patient had normal antinuclear antibodies, anti-neutrophilic cytoplasmic antibodies, and anti-mitochondrial antibodies.

Then the patient underwent ERCP, which revealed a mildly dilated entire main bile duct with the largest diameter of 8 mm. A biliary sphincterotomy was performed, and only sludge was found on sweeping the biliary tree. However, fluoroscopic imaging of ERCP revealed irregularity of the intrahepatic and extrahepatic biliary tree with no focal filling defects or high-grade stricturing. These findings lead to liver core biopsy. Pathology results of liver biopsy revealed ductopenia and marked bile ductular proliferation with cholangitis suggestive of early primary sclerosing cholangitis. On discharge, the patient was started on Ursodeoxycholic acid, scheduled for a colonoscopy, and referred for liver transplantation.

3. DISCUSSION

Diagnosing PSC early during the disease process can be challenging. Cholangiogram using magnetic resonance cholangiopancreatography (MRCP), endoscopic retrograde cholangiopancreatography (ERCP), or percutaneous transhepatic cholangiography (PTC) are used to establish a diagnosis of PSC.^[5] MRCP being non-invasive, is the first test of choice. ERCP is better than MRCP at detecting early stages of PSC and large-duct PSC, but for patients with suspected small

duct PSC, a percutaneous liver biopsy is required.^[6] Detection of aggregated neutrophils in a bile duct (ductopenia) on a liver biopsy specimen is diagnostic of PSC. However, secondary causes of sclerosing cholangitis must be excluded before confirming the diagnosis of PSC.^[7]

PSC eventually leads to cholestasis and hepatic failure.^[8] Up to 90 percent of patients with PSC also have ulcerative colitis (UC). Because of the strong association between PSC and UC, evaluation for inflammatory bowel disease is strongly recommended.^[3] Median survival without liver transplantation after diagnosis is 10 to 12 years.^[9] The role of any medical therapy is unproven so far. Patients need to be regularly screened for cholangiocarcinoma, gall bladder cancer, colorectal cancer, and if they have cirrhosis, hepatocellular carcinoma.^[10] Liver transplantation is now the treatment of choice for patients with advanced liver disease secondary to PSC.^[7]

4. CONCLUSION

Diagnosing early PSC or variants of PSC like small duct PSC or overlap with autoimmune hepatitis can be extremely difficult. In patients with early stages of PSC, bile ducts may solely present with superficial ulcerations rather than the characteristic strictures. For the detection of such shallow ulcerations, ERCP is the test of choice. And if ERCP remains inconclusive, a liver biopsy is warranted to diagnose underlying pathology.

CONFLICTS OF INTEREST DISCLOSURE

The authors have declared no conflicts of interest.

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