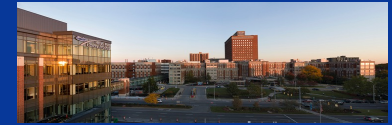




A Unique Case of Small Duct Primary Sclerosing Cholangitis Leading to Recurrent Cholestatic Hepatitis

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Background and Introduction

Small Duct Primary Sclerosing Cholangitis (sdPSC) is the narrowing of intrahepatic bile ducts which often leads to cholestasis and cirrhosis.

Types of Primary Sclerosing Cholangitis¹

1. Classic – affects both large and small ducts
2. Small Duct – only affects small ducts
3. Associated with autoimmune hepatitis – affects large and small ducts

Comparison to PSC¹

sdPSC has similar histological features to PSC and may evolve into PSC. sdPSC generally has a better prognosis than PSC.

Diagnosis¹

1. Liver biopsy consistent with PSC
2. Normal ERCP
3. Abnormal liver function tests
4. Exclusion of other liver diseases

Past Medical History

- Hypertension
- GSW with associated colectomy.
- Cholecystectomy
- Episodes of abdominal pain, jaundice, hyperbilirubinemia, diarrhea, and weight loss occurring three times a year for three-month intervals.
- Previously diagnosed with benign recurrent intrahepatic strictures.

Labs upon Admission

| | |
|----------------------|----------|
| Alkaline Phosphatase | 368 IU/L |
| AST | 56 IU/L |
| ALT | 52 IU/L |
| ANA | 1:320 |

Case Presentation

A 42-year-old male patient presented to Henry Ford Hospital emergency room (Detroit, MI) for abdominal pain associated with recurrent cholangitis.

Hospital Course

- A liver biopsy was taken during admission and revealed mild portal inflammation and cholestasis, bridging fibrosis, and intracytoplasmic iron granules.
- One month later a colonoscopy showed patchy erythema with biopsies demonstrating mild acute proctitis.
- Patient was diagnosed with sdPSC and was placed on liver transplant list.
- His postoperative course was notable for persistently elevated bilirubin and a mild anastomotic stricture.

Treatment:

Surgery

The sdPSC was treated with a cadaveric liver transplant, and upon explant, the patient's liver showed chronic obstructive cholangiopathy

Procedure

Upon ERCP, the transplant liver showed a mild anastomotic stricture and tortuous duct which was treated with a stent. However, the stent was removed because of lack of frank stricture.

Medication

Transplanted liver was managed with a prednisone taper, mycophenolate, tacrolimus, and ursodiol
sdPSC was managed with ursodiol

Discussion

Progressive Decline Lends Clinical Suspicion

A 42-year-old male presented with recurrent episodes of abdominal pain, bone pain, hyperbilirubinemia, diarrhea, and jaundice leading to a previous diagnosis of benign recurrent intrahepatic strictures (BRIC). Given the progressive decline of the patient, a degree of clinical suspicion of the BRIC diagnosis was held. Further tests to determine the diagnosis included a colonoscopy, ERCP, and MRCP.

Differential Diagnosis

Clinical features of the patient endorsed a diagnosis of either BRIC or PSC. The liver biopsy performed upon admission revealed bridging fibrosis, which excluded a diagnosis of BRIC. Furthermore, both ERCP and MRCP revealed a lack of extrahepatic ductal fibrosis which also excluded classical PSC.

A Novel Diagnosis

Given cholestatic pattern, + ANA 1:320, recurrent admissions with sepsis, and recent colonoscopy showing evidence of proctitis (biopsy proven), the patient was diagnosed with sdPSC with overlap of autoimmune cholangiopathy.

Acknowledgements

Sources:

1. Broomé U, Glaumann H, Lindstöm E, et al. Natural history and outcome in 32 Swedish patients with small duct primary sclerosing cholangitis (PSC). *Journal of Hepatology*. 2002;36(5):586-589. doi:10.1016/s0168-8278(02)00036-3

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