Portal Cavernoma Cholangiopathy Presenting With Acute Cholangitis

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Introduction

Portal cavernoma cholangiopathy (PCC) is defined as abnormalities in the biliary tree due to a portal cavernoma. Most cases occur in non-cirrhotic patients who remain asymptomatic. 5-30% of patients develop symptomatic disease and less than 8% of symptomatic cases present with acute cholangitis. This case describes PCC as an uncommon cause of acute cholangitis.

Case Presentation

•A 26-year-old male with a medical history of acute lymphocytic leukemia in remission, cryptogenic cirrhosis, and portal vein thrombosis (PVT) on warfarin presented with 2-weeks of worsening right upper quadrant abdominal pain, darkening of urine and 10lb weight loss.

•Vitals: 101°F

•Physical Exam: scleral and palatal icterus. Tenderness to palpation of his RUQ abdomen.

•Labs revealed white blood count: 13.5 x10^3/mcL, total bilirubin 7.5mg/dL, & ALP 398 U/L.

•MRCP: biliary dilation related to extensive portal vein cavernous transformation and narrowing in the common bile duct (CBD) due to compression by cavernous vessels in porta hepatis. (Figure 1A)

•Endoscopic ultrasound (EUS): PVT and collateral flow. (Figure 1B)

•Endoscopic retrograde cholangiopancreatography (ERCP) revealed biliary narrowing and beading. These findings were diagnostic of cavernous transformation with cholangiopathy. (Figure 1C)

Figures/Pictures

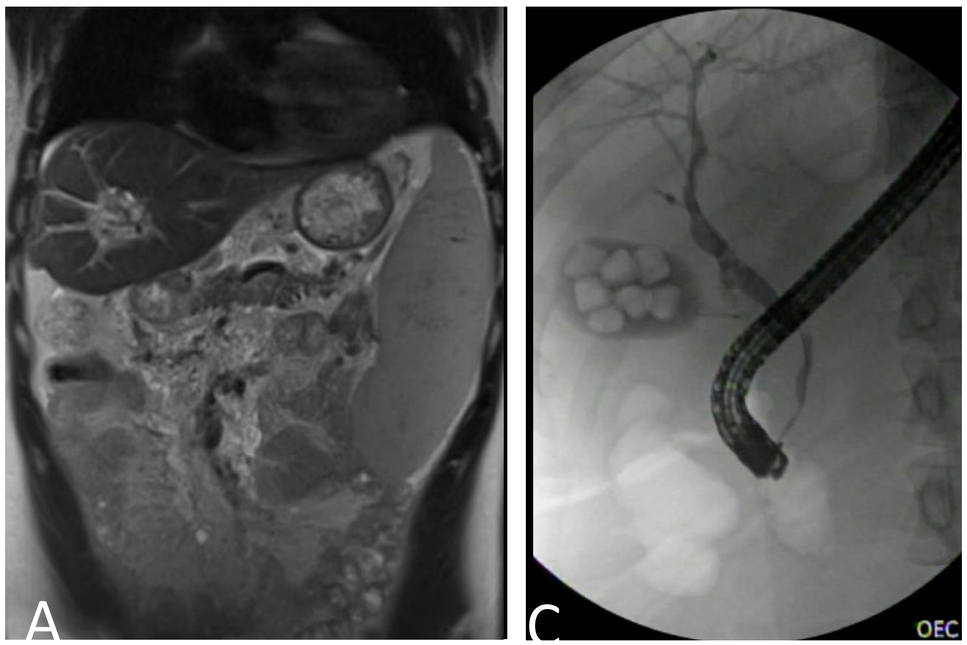

Figure 1:

A) MRCP: Cavernous transformation of the portal vein. B) EUS: A portal vein hrombus involving area of portal vein confluence and collateral flow consistent with cavernous transformation. C) ERCP: Abnormal areas of narrowing and beading throughout the biliary system with extrinsic stenosis.

Hospital Couse

Patient was started empirically on piperacillin and tazobactam for anaerobic coverage. ERCP stent was placed, and he was discharged from the hospital.

On follow up patient had repeat stenting 6 months later.

There was a discussion about TIPS and Portal vein reconstruction. However, the patient was not able to follow through due to changing insurances.

Discussion

PCC is a rare complication of PVT occurring in 0.5-1.0%

Portal cavernoma cholangiopathy is categorized into preclinical, asymptomatic, symptomatic, and complicated stages.

MRCP is gold standard to elucidates liver vascular anomalies to diagnose, classify and grade PCC

Pathological components of PCC can be split into reversible or fixed. Portosystemic shunt creation, TIPS, or ERCP sphincterotomy can resolve reversible findings via decompression of collaterals.

PCC the mainstay of management is now endoscopy. Further endoscopic intervention, surgery or TIPS are considered for persistent biliary obstruction..

References

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