Introduction

- Portal biliopathy is biliary ductal changes in patients with portal hypertension.
- Long standing obstruction of the portal vein leads to replacement of the portal vein by large collaterals along the CBD. These large collaterals compress the pliable CBD.
- Patients can present with icterus, episodes of biliary pain, hepatomegaly, splenomegaly, or ascites.

Case

HPI

36 year-old female who presented with complaints of right upper abdominal dull pain for 3 days. Her pain was non-radiating, with no exacerbating or alleviating factors. It was accompanied by nausea and non-bloody diarrhea.

PMH: laparoscopic sleeve gastrectomy for morbid obesity complicated by portal vein thrombosis, irritable bowel syndrome, atrial fibrillation and history of cholelithiasis

Medications: multivitamin, zofran oral as needed, oxycodone 5 mg oral as needed

Social Hx: Married, 3 children, no tobacco, ETOH or drug use

Family Hx: Father – DM II, hypertension

Physical Exam

T: 98.3 F, HR: 80 bpm, RR: 16, BP: 110/70

General: Alert and oriented

HEENT: No conjunctival pallor, dry mucosa

Gastrointestinal: Soft, moderate tenderness to palpation in epigastrium, No organomegaly

Integumentary: Tanned complexion, no rashes

Musculoskeletal: No arthralgia

Psychiatric: Anxious, flat affect

She was admitted to the hepatobiliary surgery team for concern for symptomatic cholelithiasis and possible cholecystectomy

Further Workup

Laboratory Values

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Imaging

MRI Abdomen day 1

- extensive cavernous transformation of both the intra- and extrahepatic portal veins. Chronic thrombosis and scarring of the mid to distal splenic vein with new onset perisplenic, perigastric, and paraesophageal varices. Fig 1

- Abdominal ultrasound- 2.5 cm gallstone. No sonographic signs of cholecystitis.

Repeat MRI abdomen (on hospital day 3):

Showed sight prominence of the intrahepatic bile ducts. These taper smoothly to the CBD, which is not dilated. However, there is slight narrowing of the central intrahepatic bile ducts at the level of the collaterals, that reflect mild portal biliopathy. Fig 2

Endoscopic ultrasound done on hospital day 8:

- showed dilated submucosal vascular spaces. Fig 3

- Spyglass cholangioscopy showed intraductal varices in lower CBD. Fig 4

Treatment

- TIPS was unsuccessful on the first try due to extensive thromboses in PV. DIPS was successfully placed with successful recanalization of chronic portomesenteric occlusion

- The procedure was complicated by splenic vein rupture, which was stented without issue

- Patient had resolution of her right upper quadrant pain after successful DIPS placement

Discussion

- Cholangiographic abnormalities can occur in patients with portal cavernoma.
- Identifying features Biliary stenosis are wavy appearance of the bile ducts, angulation of the CBD, upstream dilatation of the bile ducts
- Occur in 1-30% of patients with cirrhosis and in 9-40% of patients with non-cirrhotic portal hypertension
- 70%-95% do not manifest with any symptoms of biliary obstruction

Conclusion

In asymptomatic patients, no specific treatment is indicated but serial abdominal ultrasound with dopplers are recommended.

Treatment includes endotherapy dilation of biliary strictures, placement of biliary stents if symptoms persist, surgical portosystemic shunt or TIPS are done for portal venous decompression.

It is important to consider PB in a patient with acute presentation of cholestasis, with a history of chronic vein thrombosis or portal hypertension.

References