Locally Invasive Cholangiocarcinoma Presenting as Portal, Splenic, and Mesenteric Vein Thrombosis

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Introduction

- Portal vein thrombosis is fairly uncommon and etiology can be difficult to determine. Etiology includes cirrhosis, hepatobiliary malignancy, abdominal infection, myeloproliferative malignancy, and inherited prothrombotic disorders.[3]
- Cholangiocarcinoma is cancer of the bile duct. It is the second most common primary hepatic malignancy representing 10-20% of cases.[2]
- This case looks at patient who initially presents with abnormal LFTs with initial work up revealing portal, splenic, and mesenteric vein thrombosis without obvious etiology who is later found to have cholangiocarcinoma.[3]

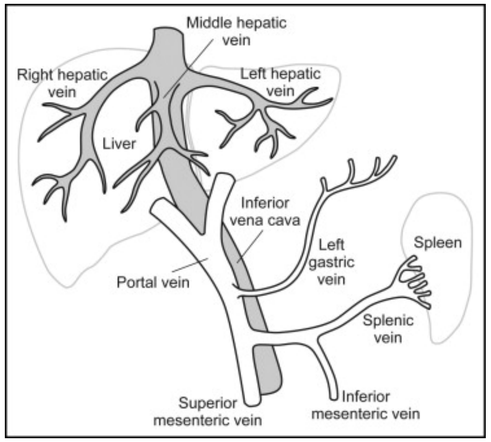

Image 1: Normal anatomy of the portal vein and its tributaries[3]

Case Presentation

A 79-year-old woman with a past medical history significant for hyperlipidemia and breast cancer presenting with LFT abnormalities. She was asymptomatic and work up revealed chronic PVT, splenic and superior vein thrombosis with gastric and esophageal varices. She underwent banding of esophageal varices. Evaluation for inherited prothrombotic disorders was unrevealing. MRI and CT of the abdomen and pelvis did not reveal signs of malignancy. Three months later she represented to the hospital with RUQ pain with associated jaundice, acholic stool, and dark urine. She denied weight loss or anorexia.

Physical Exam: BP 158/70, pulse 72, temp 98.5°F, scleral icterus, jaundice, abdominal exam without tenderness or hepatosplenomegaly

Admission labs: LFTs (AST 167, ALT 190, alkaline phosphatase 672, total bilirubin 9.2, direct bilirubin 7.2), CBC WNL, BMP WNL

RUQ US with duplex: occlusion of the portal splenic confluence and main portal vein, non-visualization of the right and left portal veins, patent hepatic veins, mild splenomegaly

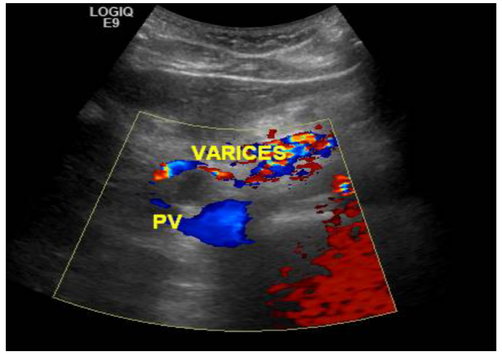

Image 2: RUQ ultrasound showing the occlusion of the portosplenic confluence

Clinical Course

She underwent MRI pancreas showing moderate diffuse enhancement of the periportal connective tissues and soft tissues suggesting underlying cholangiopathy.

Subsequently, EUS was remarkable for a mass in the middle third of the main bile duct. She underwent stent placement of a malignant-appearing biliary stricture, biliary sphincterotomy, dilation of the upper third of the main bile duct, and biopsy of the bile duct. There was minimal improvement of direct bilirubin, AST, ALT, and alkaline phosphatase post-procedure. Biopsy revealed adenocarcinoma.

Once discharged, she followed up with medical oncology and she elected against surgery or chemotherapy.

She was readmitted one month later for acute cholangitis. Shortly after she decided to pursue hospice care and passed away.

Discussion

- Portal vein thrombosis can present with abdominal pain, nausea, and fever if acute. If chronic, patients may present with signs and symptoms of portal hypertension (variceal bleeding, ascites, and hypersplenism).[2]
- This patient has chronic PVT with resulting varices. Work up for malignancy in this patient was initially negative which prompted evaluation for inherited prothrombotic disorders. It is important identify etiology as this leads to an underlying cause that may need to be treated.
- Diagnosis of early stages of cholangiocarcinoma can be difficult because many patients are asymptomatic. It is usually diagnosed at advanced stages.
- Malignant strictures within the right or left intrahepatic bile ducts may experience weight loss, abdominal discomfort, and malaise. Tumors that originate in the hilar and distal biliary ducts that obstruct biliary flow are associated with jaundice, pruritis, and acholic stools.[2]
- It is can also be discovered as an incidental finding or during evaluation of abnormal liver function test.[1]

Conclusions

- The purpose of this case presentation was to demonstrate uncommon presentation of cholangiocarcinoma.
- This case shows that there must be high index of suspicion for abdominal malignancy as a cause of portal vein thrombosis.
- Perhaps EUS should be considered despite normal abdominal MRI if other work up is negative.

References

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