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CASE REPORT

Portal Vein Aneurysm Presenting with Obstructive Jaundice

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ABSTRACT

To the best of our knowledge, a portal vein aneurysm presenting with obstructive jaundice has not been reported in the literature. The preferred treatment for these aneurysms is surgical and a shunting procedure should be considered in cases with portal hypertension to preserve portal vein flow when portal hypertension is present or is secondary to the aneurysm itself. In our case, due to patient's advanced age and co-morbidities, an endoscopic biliary stent was placed which led to successful resolution of symptoms of obstructive jaundice.

Key words: Biliary obstruction, obstructive jaundice, portal vein aneurysm

INTRODUCTION

The majority of portal vein aneurysms is acquired and occurs secondary to advanced hepatocellular disease with portal hypertension and increased portal pressures. Other acquired causes include pancreatitis, trauma, post-transplantation, and less commonly portal vein aneurysms seen in the pediatric population without underlying liver disease or portal hypertension. These aneurysms are usually incidental on imaging and asymptomatic. The preferred treatment for these aneurysms is surgical. A shunting procedure should be considered in cases with portal hypertension to preserve portal vein flow when portal hypertension is present or is secondary to the aneurysm itself. In our case, due to the patient's advanced age and associated co-morbidities an endoscopic biliary stent was placed which led to successful resolution of symptoms of obstructive jaundice.

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CASE REPORT

An 89-year-old female presented to the emergency

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department with jaundice, severe pruritis, and low- grade fever of 100° F that had persisted for 3 weeks. Patient had multiple underlying co-morbidities, most notably untreated non-Hodgkin's lymphoma, chronic renal dysfunction from diabetes mellitus, and myelofibrosis. Significant findings on clinical examination included splenomegaly. Initial laboratory analysis revealed total serum bilirubin of 7.5 mg/dl (normal 0.3 to 1.9 mg/dl) (conjugated bilirubin 6 mg/dl), alkaline phosphatase 762 U/L (normal <126 U/L), Aspartate Aminotransferase (AST) 141 U/L (normal <41 U/L), and Alanine Aminotransferase (ALT) 274 U/L (normal <45 U/L). There was thrombocytopenia, platelet count of 90 000/ mm^3. The other parameters such as complete blood count, INR (International normalized ratio), and serum albumin were normal. Hepatitis workup including hepatitis A IgM antibodies, hepatitis B surface antigen, hepatitis B IgM core antibody, and hepatitis C antibodies were also negative. In view of the patient's advanced age and clinical presentation, the working diagnosis was that of Klatskin tumor (hilar cholangiocarcinoma) *versus* lymphoma extending up to the porta hepatis.

An abdominal CT scan, without intravenous contrast at an outside facility was performed (due to underlying renal (dysfunction, estimated glomerular fi lteration rate (GFR) of 38 ml/min), revealed a 4.8 cm soft tissue attenuation mass at the porta hepatis adjacent to the pancreatic head. There was no intrahepatic biliary ductal dilatation reported at the time of initial presentation. Given her multiple co-morbidities, she was transferred to the university hospital for consideration of ERCP (endoscopic retrograde cholangiopancreatography) with biliary stenting. In view of the patient's history of lymphoma, the possibility of confluent periportal lymphadenopathy causing extrinsic biliary narrowing was raised, and an abdominal MRI and magnetic resonance cholangiopancreatography (MRCP) were performed (Siemens Magnetom Avanto 1.5 Tesla, Siemens Medical Solutions, Erlangen, Germany). The scans showed mild intrahepatic biliary dilatation. On axial and coronal T1- and T2-weighted images, a large elliptical flow void was noted at the porta hepatis which on post gadolinium dynamic sequences showed enhancement similar to adjacent venous structures [Figures 1 and 2]. Findings were consistent with an aneurysm of the extra hepatic portal vein. Incidentally noted was an early cirrhotic appearance of the liver with surface nodularity and splenomegaly related to portal hypertension (spleen craniocaudal dimension was 15 cm). No ascites was present. The portal vein aneurysm caused a smooth extrinsic

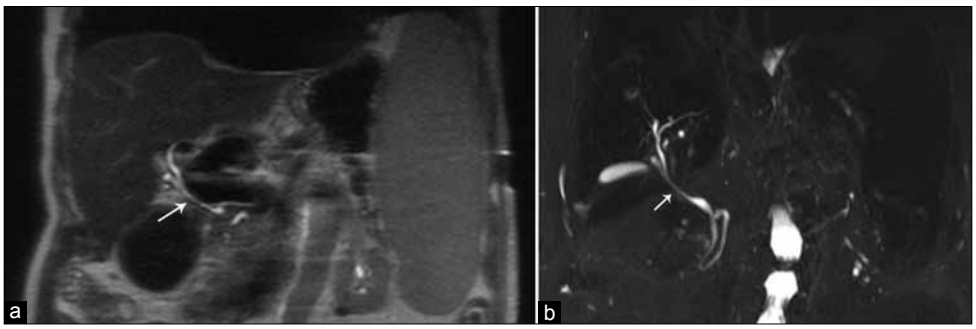

Figure 1: An 89-year-old female with portal vein aneurysm. (a) Coronal Half Fourier Acquisition Single shot Turbo spin Echo (HASTE) image and (b) magnetic resonance cholangiopancreatography (MRCP) image show a 4.8 cm long smooth narrowing of the common bile duct (arrow) at and superior to the porta hepatis secondary to mass effect from the large portal vein aneurysm. Mild intrahepatic biliary ductal dilation was present which is not well appreciated on these images. Marked splenomegaly was present related to portal hypertension from underlying myelofibrosis. No ascites was seen.

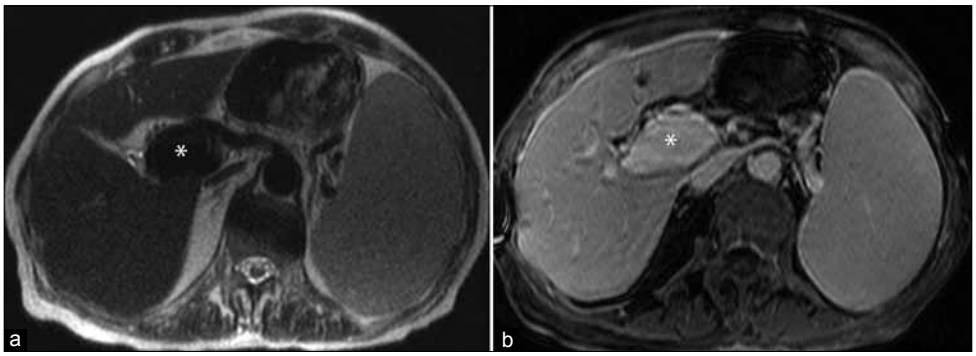

Figure 2: An 89-year-old female with portal vein aneurysm. (a) Axial T2-weighted and (b) post gadolinium T1-weighted MR images show a large enhancing aneurysm of the main portal vein (enhancing to the same degree as the IVC (inferior vena cava) and splenic vein) (asterix), causing extrinsic mass effect on the adjacent common bile duct with narrowing.

mass effect upon the common hepatic duct at the porta hepatis with significant narrowing of the common bile duct (CBD) to a diameter of 3 mm.

Color Doppler and duplex US were performed at the request of the referring gastroenterology service to confirm the suspicion of a portal vein aneurysm compressing the common hepatic duct (CHD). Transverse color Doppler US images demonstrated a large aneurysm of the portal vein measuring 3.2 cm in diameter and extending over a length of 4.8 cm, with turbulent flow noted within the aneurysm [Figure 3]. The aneurysm causes significant extrinsic compression upon the adjacent common hepatic duct (CHD) and common bile duct (CBD) with subsequent mild intrahepatic biliary dilatation, better appreciated on real time imaging. Marked splenomegaly was again noted. No ascites was present.

Subsequent ERCP showed mild intrahepatic biliary ductal dilatation with a short segment of smooth significant narrowing of the CHD and proximal CBD secondary to extrinsic mass effect from the portal vein aneurysm [Figure 4]. A sphinterotomy was performed and a biliary stent was placed with resolution of intrahepatic biliary ductal dilatation. There was slow normalization of elevated serum bilirubin (likely related to underlying myelofibrosis), liver enzymes, and overall improvement of patient's clinical symptoms over the next week. Total serum bilirubin was 0.6 mg/dl (normal 0.3 to 1.9 mg/dl) after 7 days of biliary stent placement. The other liver function tests were also normal.

DISCUSSION

Portal vein aneurysms are relatively uncommon. The majority of portal vein aneurysms is acquired and occurs secondary to advanced hepatocellular disease with portal hypertension and increased portal pressures such as in our case, but presentation with obstructive jaundice is rare.[1] Other acquired causes include pancreatitis and trauma.[2] Release of pancreatic enzymes in the vicinity of the porta

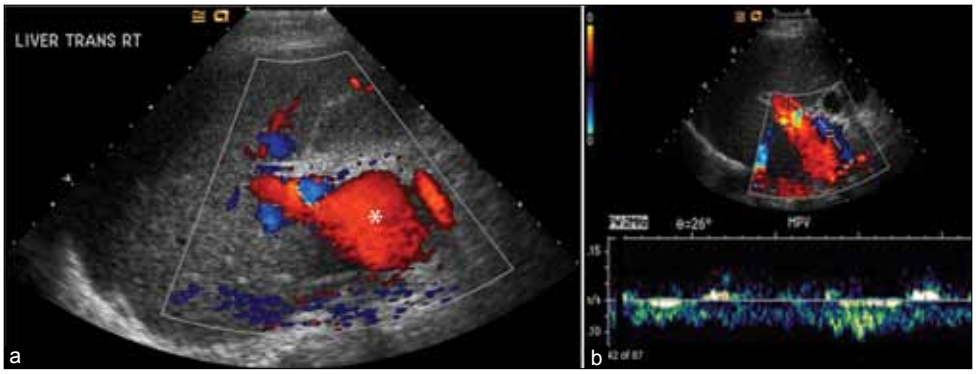

Figure 3: An 89-year-old female with portal vein aneurysm. (a) Color Doppler and (b) duplex US images show a large elliptical aneurysm of the main portal vein with turbulent to and fro flow (Ying-Yang sign) within the aneurysm.

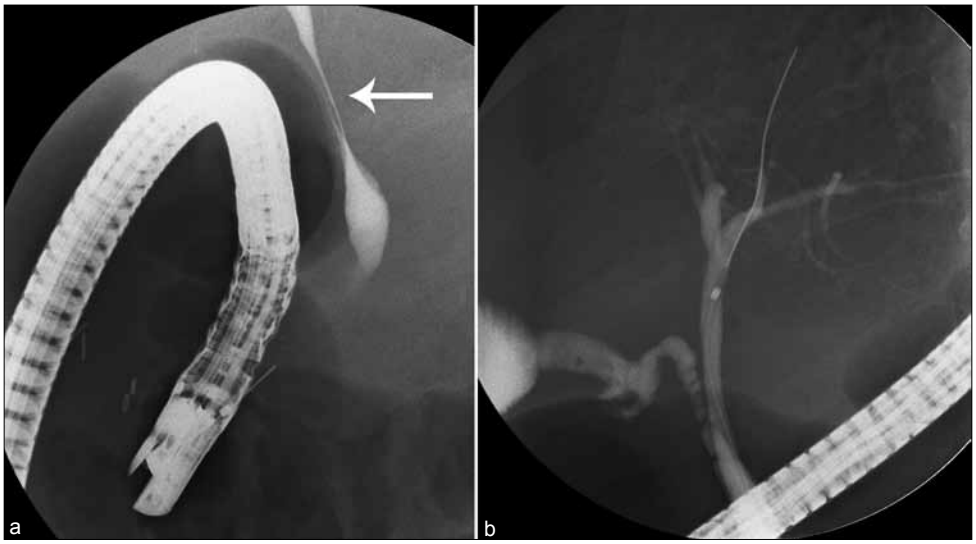

Figure 4: An 89-year-old female with portal vein aneurysm. Endoscopic retrograde cholangiopancreatogram (ERCP) images (a) pre- and (b) post biliary stent placement show a smooth long segment narrowing of the common bile duct (arrow) secondary to the portal vein aneurysm, (not seen on ERCP) treated with endoscopic spincterotomy and biliary stent placement.

hepatis in acute pancreatitis causes a weakening of the adjacent vessel walls leading to development of portal vein aneurysm and aneurysms of adjacent arteries. A case of acquired portal vein aneurysm, secondary to gastric adenocarcinoma invading the portal venous system, has also been reported in the literature.[3] A smaller number of portal venous aneurysms are congenital, supported by the fact that portal vein aneurysms have been reported in children and young adults without evidence of liver disease or portal hypertension and have also been diagnosed in utero. Congenital portal vein aneurysms are likely the result of an underlying congenital weakness of the portal vein wall.[4] Portal vein aneurysms are being seen with increasing frequency in patients following orthotopic liver transplantation. These aneurysms are classically seen at the surgical portal vein anastomosis.[4] Acute thrombosis of the portal venous systems may sometimes lead to aneurysmal dilatation.[5,6] Portal vein aneurysms following cholecystectomy have been reported. These are likely due to inadvertent injury during operative procedure. Portal vein aneurysms often present in conjunction with major gastrointestinal variceal bleeding, but may occur with minimal or no symptoms. Thrombosis, rupture, and pressure effects are major complications although clinical presentation is varied with vague right upper quadrant pain being a common presenting symptom. Most are asymptomatic and discovered incidentally. It is rare for a portal vein aneurysm to be present with obstructive jaundice. The diagnosis is usually confirmed with Duplex-doppler US, CT imaging, or MRI. Most reported extrahepatic portal vein aneurysms occur at the confluence of the superior mesenteric and splenic veins. Intrahepatic aneurysms tend to occur at vascular bifurcations, a potentially weak site (80% of reported cases). It is unclear whether vascular confluences and bifurcations are more susceptible to variations in intraluminal pressure than linear vessels.[7]

The preferred treatment for these aneurysms is surgical, depending on aneurysm size, symptoms, complications, and overall clinical status of the patient.[8] In our patient endoscopic biliary stent placement bridging the site of narrowing was deemed sufficient, taking into account patient's advanced age and co-morbidities. In symptomatic patients, surgical resection entails longitudinal incision of the aneurysm with reconstruction of the portal vein from the aneurysm segment with longitudinal oriented sutures with wrapping of the remaining aneurysm wall around the portal vein.[9] A shunting procedure is also an option to preserve portal vein flow. A case of acutely thrombosed portal vein aneurysm treated with thrombectomy and aneurysmorrhaphy, with successful 10-year follow-up, has been reported in the literature.[10]

To the best of our knowledge, an extrahepatic portal vein aneurysm presenting with obstructive jaundice has not been reported. There have been no prior reports detailing stenting of the common bile duct in this scenario with fairly rapid alleviation of patient's symptoms. In our case, the portal vein aneurysm was the likely result of longstanding increased blood flow through the splenoportal system due to splenic enlargement secondary to untreated lymphoma.

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