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CASE REPORT | BILIARY

Portal Cavernoma Mimicking Pancreatic Malignancy

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ABSTRACT

Portal cavernoma colangiopathy (PCC) is an uncommon cause of portal hypertension, and it is an important differential diagnosis of pancreatic malignancy given the expanded network of collateral vessels. On imaging studies, portal cavernoma can be seen as a hypoechoic mass, possibly associated with distal common bile duct obstruction. Most cases occur in non-cirrhotic patients. During the symptomatic phase, these patients carry a high-risk of complications related to sustained biliary obstruction. We report a unique patient with obstructive jaundice and a presumed pancreatic mass that proved to be a portal cavernoma complicated by PCC in the setting of nodular regenerative hyperplasia of the liver.

INTRODUCTION

Portal cavernoma cholangiopathy (PCC) is characterized by biliary tract abnormalities and is typically seen in non-cirrhotic patients with extrahepatic portal vein obstruction. Chronic portal vein thrombosis may complicate portal cavernoma, leading to portal hypertension. Portal cavernoma refers to the expanded collateral vessels and is considered a preclinical stage of PCC as it can compress the common bile duct (CBD).[1]

CASE REPORT

A 65-year-old white man was hospitalized due to jaundice and abdominal distension. He had a history of portal hypertension secondary to cryptogenic cirrhosis diagnosed 4 years prior, when he was first noted to have thrombocytopenia and large non-bleeding esophageal varices. He did not undergo liver biopsy at that time. As he developed progressive jaundice, muscle wasting, and ascites, he was admitted to a local hospital. Laboratory evaluation showed alkaline phosphatase 674 U/L, total bilirubin 6.8 mg/dL, and transaminase 60 U/L. Abdominal computed tomography showed a low-attenuation lesion measuring 3.7 × 6.4 × 5.1 cm with peripheral nodular enhancement, consistent with hemangioma. Biliary ductal dilatation was also noted, both centrally and peripherally, with the CBD measuring up to 1.5 cm (Figure 1). Magnetic resonance cholangiopancreatography (MRCP) revealed intra- and extrahepatic biliary dilatation, as well as irregular narrowing of the CBD at the level of the porta hepatis surrounded by an ill-defined soft-tissue lesion (Figure 2). The patient underwent liver biopsy, and he was transferred to our hospital for further evaluation before the results of liver biopsy were available.

To better assess the CBD stricture and the unclear "mass at porta hepatis," endoscopic ultrasonography (EUS) was performed. A hypoechoic mass was seen at the pancreatic head region with associated distal CBD obstruction and extensive collateral circulation (Figure 3). Endoscopic retrograde cholangiopancreatography (ERCP) demonstrated a localized biliary stricture with a malignant appearance. A metal stent was placed into the CBD via sphincterotomy. Fine-needle aspiration from the EUS and brush cytology from the ERCP were negative for malignancy. The liver biopsy slides revealed nodular regenerative hyperplasia with minimal fibrosis. Bile ducts were present and without injury. Repeat EUS showed stable vascular lesion in the peripancreatic area. The patient's abdominal

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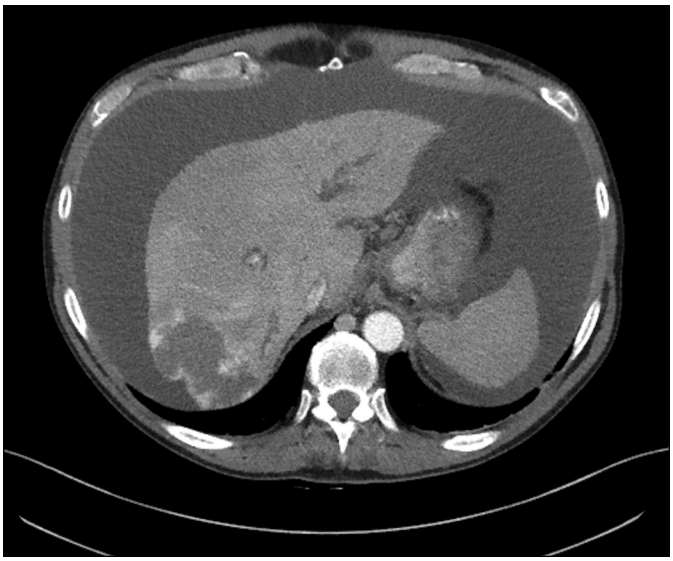

Figure 1. Abdominal computed tomography showing biliary ductal dilatation and a low-attenuation hepatic lesion (3.7 × 6.4 × 5.1 cm) in the right lobe, suggestive of hemangioma.

distension and jaundice improved after the placement of the biliary metal stent. Given the definitive changes from portal cavernoma, a decision was made to keep the stent in place. Serum levels of bilirubin and alkaline phosphatase also decreased after the procedure. Of note, a coagulation work-up performed during this hospitalization was negative for thrombophilias, including factor V Leiden, prothrombin gene mutation, protein C and S deficiency, and antithrombin II deficiency.

DISCUSSION

Portal cavernoma is thought to be the result of chronic portal vein thrombosis, caused by different conditions such as hypercoagulable states, myeloproliferative disorders, or pancreatitis. Approximately 30% of patients have no identifiable cause for the portal vein thrombosis.[2] Diagnosing PCC in older patients is challenging because the presentation and imaging findings are concerning for malignancy. Differentiation between these entities is key. Radiologic changes of PCC develop as a complication of long-standing portal cavernoma and compression to the biliary tree in 80–100% of patients with portal cavernoma, although symptoms are seen in only 5–50% of patients.[3,4] In a consensus statement published by Dhiman et al, 3 requisites are necessary to make the diagnosis of PCC: the presence of portal cavernoma, typical cholangiographic changes, and the absence of other causes of biliary irregularities.[4]

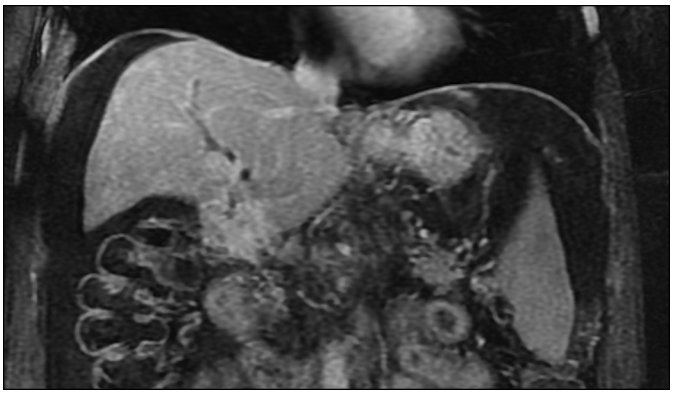

Figure 2. Magnetic resonance cholangiopancreatography revealing intra- and extrahepatic biliary dilatation, as well as an irregular narrowing of the common bile duct (CBD) at the level of the porta hepatis and surrounded by an ill-defined soft tissue lesion.

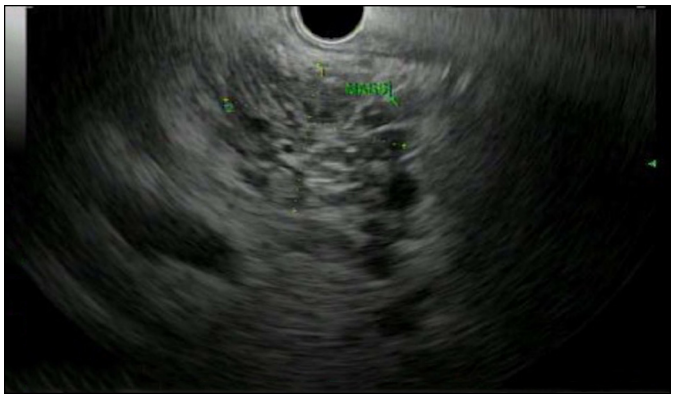

Figure 3. Endoscopic ultrasound showing a hypoechoic mass seen at the pancreatic head region with associated distal CBD obstruction and extensive collateral circulation.

Most patients with PCC are asymptomatic. Symptoms usually appear during late stages of portal hypertension, indicating long-term obstruction.[5-7] Portal cavernoma on sonography appears as an anechoic structure, described as a sponge-like mass. Magnetic resonance imaging with MRCP can detect the wavy appearance of the biliary duct related to the extrinsic compression by the portal cavernoma, which is a common finding.[7] The biliary abnormalities in PCC can be caused by direct compression of the biliary tree by the portal cavernoma or by a large collateral vessel. These changes are well-defined for non-cirrhotic patients, in whom malignant causes have been excluded. The hypoechoic mass at the peripancreatic region and biliary stricture are radiological features of PCC and is often unrecognized due to low clinical suspicion.

Asymptomatic PCC may be associated with irregularity of the bile ducts, sometimes with multiple stenoses and dilatations, and can resemble mild ischemic cholangiopathy. Patients with advanced cholangiopathy are at high risk for sustained biliary obstruction complications, including secondary biliary cirrhosis.[5] Symptoms can be triggered by biliary stasis, and patients with cholestatic jaundice are also at increased risk for cholangitis.[8-10]

Although diagnosis can be made in an asymptomatic phase with imaging studies, treatment should be offered only for symptomatic patients. The goal of the therapy is to reduce portal hypertension and resolve biliary obstruction.[3] Some cases can be relieved by porto-systemic shunts when mechanical obstruction by a large collateral vessel is the main contributing factor. However, when the injury to the biliary tree presents with an ischemic component, patients may require multiple interventions with the placement of stents throughout their lifetime.[2,11] Our patient's symptoms improved after placement of a metal stent in the biliary tree. Because we don't expect the portal cavernoma to regress, the stent was left in place, and no attempts have been made to remove it.

Nodular regenerative hyperplasia is an unusual cause of non-cirrhotic portal hypertension, resulting from obstructive vasculopathy or from irreversible injury to the sinusoids. It has been associated with systemic disorders, including autoimmune diseases and hematological malignancies, as well as with some drugs, such as azathioprine.[12] Barge et al reported that 35% of cases were not associated with any disease, and one third of patients developed portal hypertension.[13] In our patient, nodular regenerative hyperplasia likely contributed to the development of portal hypertension, although the association between PCC and nodular regenerative hyperplasia remains unclear. Jaundice and biliary tree abnormalities prompted the work-up for pancreatic malignancies. Although an uncommon cause for these findings, portal cavernoma can be an unrecognized diagnosis and should be included among the differential diagnoses for extrahepatic biliary obstruction.

DISCLOSURES

Author contributions: VM Ussui wrote the manuscript. L. Goldstein gathered clinical information. E. Souto reviewed the manuscript. C. Levy critically reviewed the manuscript and is the article guarantor.

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Informed consent was obtained for this case report.

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