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Portal Biliopathy: A Rare Diagnosis Mimicking Cholangiocarcinoma And Cholangitis

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ABSTRACT

Portal biliopathy is a term to describe the spectrum of abnormalities of the entire biliary tract or gallbladder associated with portal hypertension. It is a rare disorder, which mostly presents as sub-clinically. The most common cause of portal biliopathy is extra-hepatic portal vein obstruction (EHPVO). PB may mimic cholangiocarcinoma or sclerosing cholangitis. Misdiagnosis can be avoided using appropriate imaging modalities to prevent complications. We report a case of portal biliopathy, diagnosed in a patient who presented with complaints of abdominal pain.

Key-words: Portal vein thrombosis, Common bile duct, Collaterals, Cavernous transformation of portal vein

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INTRODUCTION:

Portal biliopathy has been described as abnormalities arising anywhere in the walls of the biliary tree as a result of portal hypertension.[1] These include intra- and extrahepatic bile duct strictures and dilatation, ischemic injury, choledochal varices and portal cavernoma formation. It is most commonly associated with portal hypertension secondary to portal vein thrombosis. Studies have shown that 80-100% of patients with extrahepatic portal vein obstruction (EHPVO) manifest clinical or radiological evidence of the wall changes typical of portal biliopathy.[2] PB may mimic cholangiocarcinoma, sclerosing cholangitis, or choledocholithiasis.[3,4] Misdiagnosis can be avoided using appropriate imaging modalities to prevent complications. We report a case of Portal Biliopathy, a rare diagnosis, in a patient referred to our department with nonspecific complaints of pain.

CASE PRESENTATION:

A 40 year old male patient presented with complaints of abdominal pain. Physical examination revealed normal vitals. On systemic examination, he was found to have mild abdominal distension, mild spleenomegaly and engorged veins over the anterior abdominal wall. Biochemical investigations revealed normal liver function tests. Ultrasound imaging of the patient revealed narrowing in the proximal part of CBD with upstream mild dilatation of the proximal biliary tract; diffuse regular wall thickening of common bile duct, common hepatic duct and its branches, neck and body of gall bladder;nonvisualisation of the portal vein; mild ascites and mild spleenomegaly. Colour Doppler imaging revealed a network of serpentine vessels which demonstrated low velocity, continuous waveform suggestive of mural collaterals within the thickened part of CBD, CHD,

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its branches and, neck and body of gall bladder and multiple collaterals in peripancreatic region. Triple phase CT scan revealed multiple collaterals in peribiliary location and gallbladder varices. There was a narrowing of CBD caused by compression due to collaterals, causing dilatation of the proximal biliary tract. Portal vein and spleenic vein were not separately visualised with multiple collaterals in peripancreatic region and perigastric region and with mild spleenomegaly. MRCP showed narrowing of the common bile duct with upstream mild dilatation of common, right and left hepatic ducts.

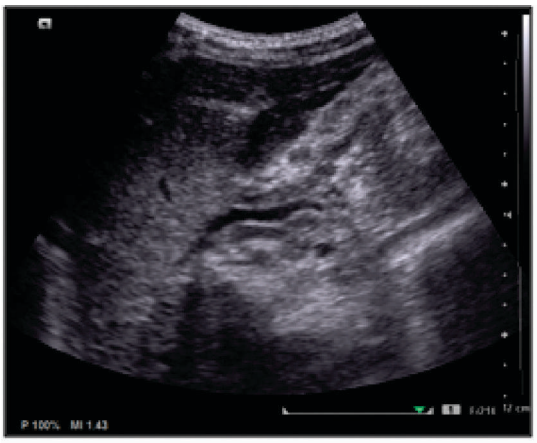

Figure 1 : B Mode Ultrasound image showing smooth regular thickening and mild dilatation of common hepatic duct

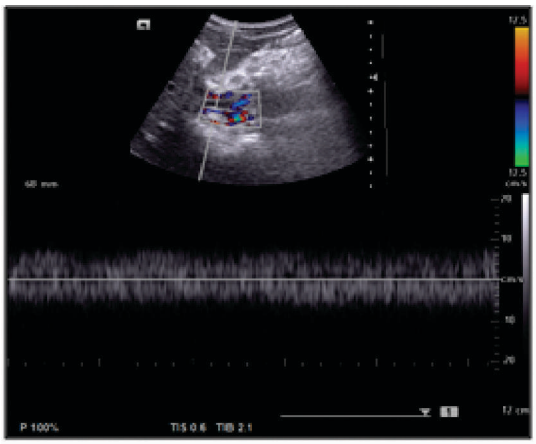

Figure 2 : Colour Doppler imaging of the thickening showing a network of serpentine vessels which demonstrated low velocity continuous waveform suggestive of mural collaterals

DISCUSSION:

The term portal biliopathy was first coined in the 1990s,[5] used to describe abnormalities in the intrahepatic and extrahepatic biliary tract, gallbladder and cystic duct secondary to portal hypertension.

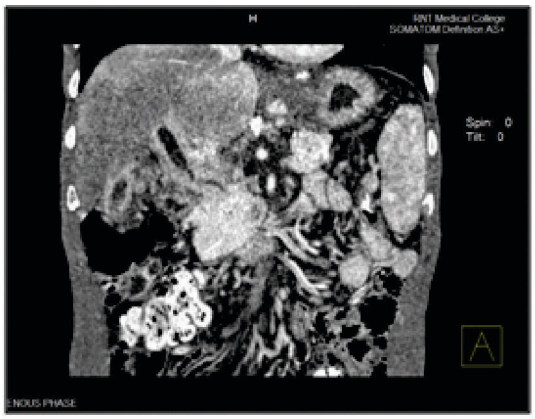

Figure 3 : Portal venous phase CT scan showing peribilary collaterals causing compression and narrowing of CBD with upstream dilatation of CHD; and multiple collaterals in peripancreatic, pericholecystic region.

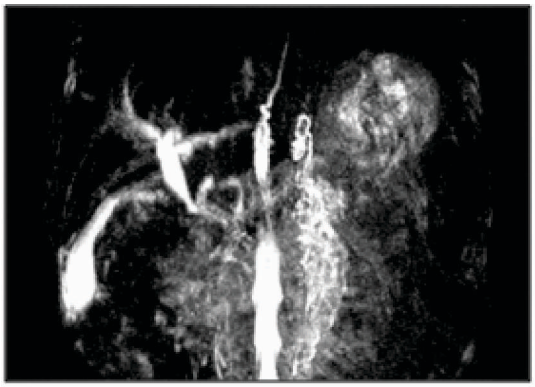

Figure 4 : MRCP image showing narrowing of CBD with proximal dilatation of CHD

It is the biliary obstruction associated with enlarged collateral veins in the setting of chronic portal vein thrombosis or cavernous transformation of the portal vein. The smooth appearance of the stenosis has prompted some to refer to this entity as *pseudosclerosing cholangitis* or *pseudocholangiocarcinoma*,but now the

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preferred term is portal biliopathy. Portal biliopathy is predominantly associated with extrahepatic portal venous obstruction. The condition has also been described in patients who have non-cirrhotic portal fibrosis and cirrhosis, but in smaller numbers. In response to chronic occlusion of the portal vein and superior mesenteric vein, several new venous collaterals are recruited in the porta hepatitis to compensate the diminished hepatopetal portal flow. These collateral veins dilate forming huge varices that completely surround the biliary tree producing a "portal cavernoma", thus providing an alternate route for portal mesenteric blood flow to enter the hepatic parenchyma.[6]

Early changes of PB occur with engorgement of the paracholedochal and the epicholedochal venous plexus, which lie adjacent to and within the biliary duct walls, respectively. Venous compensation through the paracholedochal veins in the setting of chronic PVT results in scalloped or smooth indentations in the ductal lumen, while engorgement of the smaller epicholedochal veins results in thickened, enhancing bile duct walls on cross-sectional images.[7] More dramatic ductal displacement, kinking, and further narrowing leading to proximal biliary dilatation may also occur as the larger paracholedochal and pancreaticoduodenal veins enlarge and elongate around the distal CBD. This can progress to secondary biliary cirrhosis in later stage of the disease.

Prominent changes include: irregularity and strictures of CBD and hepatic ducts which may be smooth and tapering,strictures may be single or multiple, short or long. Differential diagnosis should include hepatic hilum neoplasia, such as cholangiocarcinoma or metastatic neoplasms, as well as other causes of intrahepatic bile ducts involvement, such as sclerosing cholangitis. The diagnosis is made by imaging methods.

The main utility of Colour Doppler Ultrasound is to distinguish gallbladder and bile duct inflammatory or neoplastic wall thickening from mural varices. Doppler sonography demonstrates the portoportal collaterals as a network of serpentine vessels with hepatopetal flow. It is also capable of detecting luminal narrowing of the CBD as well as biliary duct dilatation proximal to the focal area of stenosis due to compressing venous collaterals.[8]

MDCT imaging shows similar vascular findings in the porta hepatitis. CT clearly depicts cavernous transformation of the portal vein, marked dilatation of the intra and extrahepatic portions of the parabiliary and peribiliary plexuses, and gallbladder varices.[9] MDCT can show secondary biliary ductal dilatation caused by the portal collaterals thus excluding a cholangiocarcinoma or extrinsic compression by metastatic adenopathies as the cause of obstruction. The MDCT is also useful for evaluating portal vein obstruction excluding neoplastic causes such as tumoral thrombosis, and showing ancillary vascular findings related with portal hypertension such as splenorenal shunts, gastric or esophageal varices.

MRI is the diagnostic procedure of choice for PB. MRI is also helpful in clarifying the cause of bile duct obstruction, allowing distinction between ductal wall ischemic fibrosis and cholangiocarcinoma. MRC abnormalities of the biliary system include a wavy appearance of the bile ducts, biliary ducts and gallbladder wall thickening, focal biliary stenosis, proximal dilatation, CBD angulation and lithiasis.[9,10] Shin et al classified the MRI features of the PB patients into three types-as varicoid, fibrotic, or mixed; depending on the

appearance of the bile duct at the point of obstruction.[10] MRC is useful in determining the sites of stenosis due to PB, guiding therapeutic interventions and allowing noninvasive follow up of these patients. Biliary stasis proximal to dominant strictures predisposes to lithiasis. Stones are seen as hypointense round or ovoid filling defects within dilated ducts on T2 weighted images.

CONCLUSION:

Portal Biliopathy is a condition which presents as bile duct wall thickening which can mimic cholangiocarcinoma and sclerosing cholangitis on imaging. However accurate diagnosis and evaluation can be done by various imaging modalities, which is crucial in preventing complications like bleeding from varices while attempting biopsies through ERCP.

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