International Journal of Radiology Research
www.radiologyjournal.in
E-ISSN: 2663-4562, P-ISSN: 2663-4554
Received: 01-03-2024; Accepted: 23-03-2024; Published: 15-04-2024
Volume 6, Issue 1, 2024, Page No. 18-20

Imaging of portal gastroduodenopathy: A case report

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Abstract
Portal Cavernoma Cholangiopathy (PCC) is a rare condition characterized by biliary changes secondary to extrahepatic portal vein obstruction and portal cavernoma formation. We present a case of a 40-year-old patient who presented with gastric outlet obstruction, wherein imaging studies revealed extensive alterations involving the biliary, vascular, and gastrointestinal systems. Computed Tomography (CT) demonstrated diffuse circumferential symmetric thickening of the stomach wall, with measurements of thickening noted at various regions. Additionally, mild intrahepatic and extrahepatic biliary dilatation were observed, along with smooth narrowing of the common bile duct, likely attributed to external compression by the portal cavernoma. Multiple dilated tortuous porto-systemic collaterals were evident in various regions, indicative of portal biliopathy. Concurrent findings of cholelithiasis and possibly thrombosed hepatic artery branches further complicated the clinical picture. This case underscores the multifaceted nature of PCC, highlighting the importance of comprehensive imaging assessment for accurate diagnosis and management planning.

Keywords: Portal cavernoma, porto- systemic collaterals, peripancreatic cavernoma, splenorenal collateral.

Introduction

Portal cavernoma cholangiopathy (PCC) refers to the biliary changes which occur in the setting of extrahepatic portal vein obstruction and secondary portal cavernoma formation causing vascular changes in the form of portosystemic collaterals and biliary changes in the form of extrinsic impressions and strictures. In this article we describe the non cirrhotic portal cavernpma with vascular and biliary, stomach & duodenum changes associated with PCC on US, CT. 40 yr old pt presented to us for gastric outlet obstruction for CT. CT shows diffuse circumferential symmetrically thickening of the mucosa of stomach wall noted along the fundus13mm, greater curvature 11mm, & lesser curvature 9mm extending into pylorus causing mild narrowing of the stomach wall with heterogenous enhancement. mild central & peripheral IHBR & extrahepatic biliary dilatation with smooth narrowing ofCBD mid-distal part likely due to external compression by portal cavernoma & multiple periportal collaterals s/o of portal biliopathy.

Portal cavernoma with multiple dilated tortuous porto-systemic collaterals in peri-portal, perigastric, paraesophageal, splenic hilum & paravertebral lumber regions. Cholelithiasis with possibly THAD in segment V1/V11.

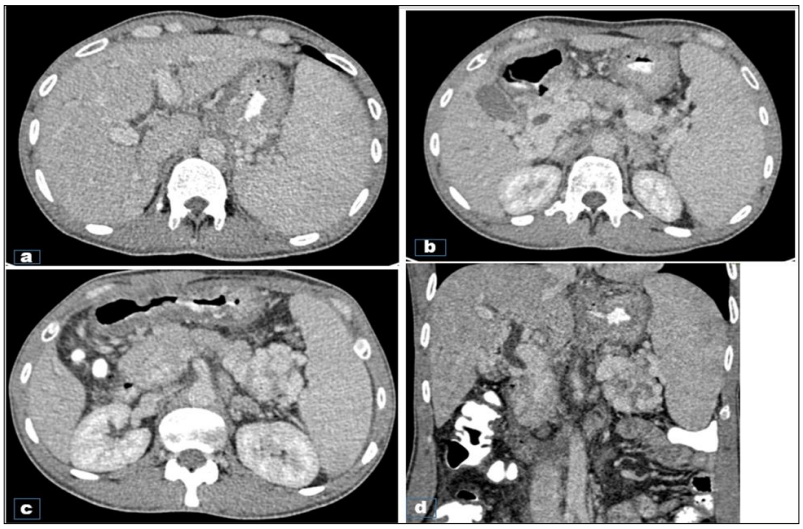

Fig 1: a) Axial section in venous phase showing dilated Intrahepatic biliary duct near biliary duct confluence. b) Axial section in venous phase showing portal cavernoma causing compression on the distal portion of common bile duct resulting in dilatation of CBD. c) Axial section in venous phase showing peripancreatic cavernoma and splenorenal collateral with diffuse thickening of gastric wall. d) Coronal section showing portal cavernoma causing compression on the distal portion of common bile duct resulting in dilatation of CBD with splenorenal collateral.

Methodology

Data retrieved from med synapse PACS.

Discussion

Portal biliopathy is an abnormality seen in the biliary system secondary to cavernous transformation of the PVT. It is also known as portal cholangiopathy or a hilar cavernoma. The varices in the hepatic hilum may displace and compress the bile ducts and may result in dilatation of the upstream bile ducts. The pathogenesis of portal biliopathy is thought to be ischemia at the time of PVT and local ischemia due to prolonged local wall compression by the peribiliary collaterals.

Patients often have asymptomatic portal biliopathy, but they may present with associated cholelithiasis, choledocholithiasis, biliary strictures, hemobilia, and cholangitis. Cross-sectional imaging, particularly with MRI, and MRCP can be used to diagnose portal biliopathy. A combination of cavernous transformation of the PV and collaterals scalloping the bile ducts is seen. The correct diagnosis of portal biliopathy should be based on cross-sectional imaging performed with IV contrast medium rather than on MRCP alone to rule out diffuse masslike thickening of the common bile duct (CBD) due to fibrous tissue deposition, which may be mistaken -Mass-forming Portal Cavernoma Cholangiopathy [1].

The mechanisms suggested for PCC are two: (a) extrinsic mechanical compression of biliary system by the collaterals (b) fibrosis of biliary system due to ischemic injury of biliary system caused by thrombosis of the venous system [2]. The collaterals initially produce smooth indentation or scalloping of the bile ducts, which further on leads to sequence of events like progressive narrowing, stenosis and kinking of biliary ducts leading on to proximal biliary dilatation. This can progress to secondary biliary cirrhosis in later stage of the disease.

Direct cholangiography was considered as the gold standard for the diagnosis of PCC in the past & is not done because of invasiveness and complications associated with endoscopic retrograde cholangiopancreatography (ERCP) including the increased risk of hemorrhage due to the peribiliary collaterals & need repeated imaging during follow-up.

All three modalities including ultrasound (US), computed tomography (CT) and magnetic resonance imaging (MRI) play an important role in the diagnosis of PCC. Ultrasound is the initial screening modality and may be useful in the follow-up of patients with PCC. It shows absence of portal vein with presence of cavernoma formation which is seen as multiple tubular anechoic structures surrounding the porta. It may vary from non-visualization of portal vein to portal vein that is completely thrombosed. The color Doppler sonography demonstrates the periportal collaterals as a network of vessels at the porta with hepatopetal flow along with absence of flow in portal vein (fig 1)

Introduction of computed tomography with helical scanning and multidetector (MDCT) technologies and recent advances in magnetic resonance imaging with faster acquisition techniques without breathing artefacts permit excellent anatomical depiction of both the biliary tree and portal system in a non-invasive manner. In this article we will review the spectrum of imaging features of PCC on US, CT and MRI.

CT clearly depicts the cavernous transformation of the portal vein, presence of the intra and extrahepatic portions of the parabiliary and peribiliary plexuses, and gallbladder varices. The extent of thrombosis and patency of portal vein with its extension into splenic vein and superior mesenteric vein and also portal vein branches can be depicted by CT. The exact location of periportal collaterals with exact localization of portosystemic collaterals can be made out [3]. CT is also useful in providing additional information about the cause of portal vein obstruction and excluding neoplastic causes such as tumoral thrombosis. Also ancillary vascular findings related with portal hypertension such as splenorenal shunts, gastric or esophageal varices and morphological stigma associated with cirrhosis of liver.fig.

MRI at present has replaced direct cholangiography as the imaging investigation of choice for PCC with direct cholangiography being reserved for interventional purposes & are helpful in depicting the vascular abnormalities of the portal vein and their relationship with the biliary tract ie portal vein thrombosis along with distribution of collaterals. MRCP abnormalities of the biliary system in PCC include a wavy appearance of the bile ducts, gallbladder and bile duct wall thickening, focal biliary stenosis, proximal dilatation, CBD angulation and proximal choledocholithiasis. MRCP can exclude similarly CT Mimickers of PCC- primary sclerosing cholangitis, recurrent pyogenic cholangitis, HIV cholangiopathy and neoplastic conditions like cholangiocarcinoma.

Comparison of imaging stydies [5]

	Ultrasound & color Doppler	Computed tomography	MRCP with MR portography
	Initial screening modality and used for follow-up. However, is operator dependant	Demonstration of vascular and biliary changes at the expense of radiation	Gold standard for PCC replacing direct cholangiography
Vascular changes	Demonstration of portal cavernoma and GB varices with color and spectral Doppler. Exact delineation of collaterals not possible	Exact delineation of extent of vascular thrombosis and portosystemic collaterals	Exact delineation of extent of vascular thrombosis and portosystemic collaterals similar to CT
Biliary changes	Assesses biliary dilatation. However, CBD changes usually difficult to delineate due to bowel gas. Detects associated lithiasis	Assesses biliary dilatation as well CBD changes. However, less accurate than MRCP	Provides complete assessment of biliary and CBD changes similar to direct cholangiography. Also helps differentiate varicoid and fibrotic type. Detects associated lithiasis
Ancillary changes	Demonstrates associated findings of cirrhosis	Demonstrates associated findings of cirrhosis and helps in ruling out mimickers of PCC	Demonstrates associated findings of cirrhosis and helps in ruling out mimickers of PCC

Portal hypertension sec to portal cavernoma can likely causes hemodynamic and mucosal changes in the entire gastrointestinal (GI) tract more so in stomach & duodenum. portal hypertension in the GI tract: 1. Varices. • Esophageal varices. • Gastric varices. • Ectopic varices& Rectal varices. 2. Portal hypertensive gastropathy, enteropathy[7], and colopathy. 3. Gastric antral vascular ectasia. PHG is recognized endoscopically as a mosaic-like pattern called snakeskin mucosa with or without red spots. Most patients with PHG are asymptomatic, but a significant number of patients exhibit symptoms related to chronic GI bleeding and chronic blood loss/iron deficiency anemia. A smaller proportion of patients exhibit evidence of active GI bleeding with an incidence of 3% to 60% of patients.

computed tomography[5] scan as enhancement on the inner layer of the gastric walls, which may reflect gastric

congestion. PHG needs to be differentiated from GAVE syndrome. Distinction between them is extremely important as treatment differs. PHG responds to a reduction in portal pressures while GAVE syndrome does not. GAVE syndrome typically occurs in noncirrhotic patients and is limited to the antrum and pylorus, whereas classic PHG exclusively involves the fundus and body.

Chandra [6] *et al* proposed a classification system for PCC based on the location of narrowing in direct cholangiography. Type I indicates involvement of extrahepatic ducts, type II indicates involvement of intrahepatic bile ducts, type III a indicates extrahepatic bile duct and unilateral intrahepatic bile duct involvement and type III b indicates extrahepatic bile duct and bilateral intrahepatic bile duct involvement.

Conclusion

PHG and PHC can cause acute and/or chronic GI bleeding. Their pathogenesis is still not completely understood, but currently available evidence suggests that their development is related to portal hypertension. Diagnosis for both is endoscopic. The differential diagnosis is sometimes difficult and in these situations biopsy and histologic examination may be helpful.

Conflicts of interest All authors have none to declare.

Funding. Nil

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