Imaging of Portal Cavernoma Cholangiopathy

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Portal cavernoma cholangiopathy (PCC) refers to the biliary changes which occur in the setting of extrahepatic portal vein obstruction and secondary portal cavernoma formation. The main radiological findings include the vascular changes in the form of portosystemic collaterals and biliary changes in the form of extrinsic impressions and strictures. Till date, conventional cholangiography has been the gold standard for the diagnosis of PCC. However, it is an invasive procedure and is associated with complications. At present there is a transition towards non-invasive modalities like ultrasound (US), computed tomography (CT) and magnetic resonance imaging (MRI). The recent advances in these modalities provide an excellent delineation of both the vascular and the biliary changes non-invasively in a short time. The findings of PCC using these newer modalities are not so well described in literature. The findings of PCC also overlap with malignant conditions of biliary tract such as cholangiocarcinoma and compression of biliary tract by malignant adenopathies. In this article we describe the vascular and biliary changes associated with PCC on US, CT and MRI. We also describe the imaging findings using each modality along with their advantages and disadvantages. (J CLIN EXP HEPATOL 2014;4:S44–S52)

The entity portal biliopathy, now proposed as portal cavernoma cholangiopathy (PCC), refers to the biliary ductal abnormalities which are secondary to extrahepatic portal vein obstruction (EHPVO) and consequent cavernous transformation of portal vein.¹ PCC has been reported in approximately 70–100% of patients with EHPVO.² However symptoms and abnormal cholestatic tests are less frequent than the cholangiographic abnormalities. This discrepancy may be explained by the forceful contractions of the gallbladder in response to the ingestion of food causing an increase in the intraluminal pressure above that of the pressure in the choledochal varices. This prevents bile stasis and development of cholestatic features.³

Multiple hepatopetal collateral vessels form in response to EHPVO. These collateral veins are known as portal cavernoma. They arise in the periportal and in peripancreatic region along the occluded portal vein and sometimes even around the splenic vein and the superior mesenteric vein. They join the patent intrahepatic portal branches and

thus they provide an alternative route around the thrombosed segment of the main portal vein and its tributaries.⁴ The portal cavernomas thus formed are composed mainly of two venous systems constituted by the paracholedochal veins, which run parallel to the ductal wall, and the epicholedochal venous plexus, located on the surface of the bile duct.⁵ The paracholedochal veins may dilate first and may cause the smooth extrinsic biliary impressions. Epicholedochal veins are in intimate contact with the outer wall of the common bile duct (CBD). Their dilatation may be responsible for the fine and irregular mural changes as the bile duct is thin and pliable. There is also development of collaterals in the gallbladder wall which acts as a collateral pathway via the cystic vein.

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.⁶ 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.⁷

Direct cholangiography was considered as the gold standard for the diagnosis of PCC in the past era (Figure 1). The most common abnormalities in conventional cholangiographic studies include irregular narrowing and stenosis of both the extra and intrahepatic bile ducts, segmental dilatation of biliary ducts with a beaded appearance, ectasia and pruning of the intrahepatic bile ducts⁴ (Table 1). However cholangiography cannot be done in all patients because of invasiveness and complications associated with endoscopic retrograde cholangiopancreatography (ERCP) including the increased risk of hemorrhage due

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Abbreviations: PCC: portal cavernoma cholangiopathy; US: ultrasound; CT: computed tomography; MRI: magnetic resonance imaging; EHPVO: extrahepatic portal vein obstruction; CBD: common bile duct; ERCP: endoscopic retrograde cholangiopancreatography; CEMR: contrast enhanced MR; PC: portal cavernoma; PC: periportal collateral

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Figure 1 (a) Direct splenoportography image showing patent splenic vein (long arrow) with attenuated portal vein (short arrow). Multiple periportal and perisplenic collateral channels (thick arrow) are seen. (b) ERCP image showing short segment smooth stricture involving mid CBD (long arrow) and smooth extrinsic impression involving common hepatic duct (short arrow) (likely due to compression by pericholedochal collateral).

to the peribiliary collaterals. Also PCC is a chronic and slowly progressive pathology where the patient may need repeated imaging during follow-up. Therefore, non-invasive imaging should be preferred and direct cholangiography should be done only when concomitant treatment has to be given and in cases where diagnosis is in doubt on non-invasive studies. Thus there is a slow transition from the invasive modality to non-invasive modalities for the diagnosis of PCC. Hence the findings of PCC on non-invasive imaging have to be well established.

All three modalities including ultrasound (US), computed tomography (CT) and magnetic resonance imaging (MRI) play an important role in the diagnosis of PCC. 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.

IMAGING FINDINGS OF PCC

Vascular Changes

The vascular changes predominantly involve the portal vein. It may be in the form of non-visualization of portal

vein or thrombotic occlusion of portal vein. Similar changes can also be seen extending to the splenic vein or the superior mesenteric vein. There are compensatory periportal collaterals (so called cavernoma) seen extending in peripancreatic and periportal region along the attenuated veins. However intrahepatic branches of portal vein including its main right and left branches are normal. There can be visualization of portosystemic collaterals in advanced stages due to the development of portal hypertension. The vascular changes are well seen with color Doppler or contrast enhanced CT or MRI. Mural, dot-like enhancement of the CBD on portal venous phase represents epicholedochal varices whereas larger collateral veins coursing parallel to the CBD are the paracholedochal varices. In patients with solid tumor-like cavernoma, the individual venous collaterals are not visualized and it is seen as a uniform enhancing hilar mass in the portal venous phase.

Biliary Changes

The biliary changes include biliary stenosis, upstream (pre-stenotic) dilatation, wavy appearance of the bile ducts, angulation or displacement of the CBD, choledocholithiasis and hepaticolithiasis. Loss of the usual tapering of CBD, abnormal increase in the dimension of the ducts proximal to a relatively stenotic segment or a narrow segment of the bile duct, accompanied by upstream ductal dilatation are the most commonly encountered biliary findings. Bile duct dilatation is defined as caliber greater than 4 mm for the secondary intrahepatic bile ducts, greater than 6 mm for the main right or left hepatic ducts or greater than 10 mm for the CBD. Stenotic segments are classified as short or long depending on whether they are <25 mm or >25 mm.⁸ The wavy appearance of the CBD may be due to the impression of periportal collaterals on the wall of the CBD whereas unusual narrowing is usually due to the fibrotic strictures. Whereas the collaterals

Table 1 Direct Cholangiographic Features of Portal Cavernoma Cholangiopathy.

1. Extrahepatic strictures
2. Intrahepatic biliary dilatation
3. Caliber irregularity
4. Ductal ectasia
5. Indentations, displacement and angulations of ducts
6. Clustering and pruning of intrahepatic ducts
7. Filling defects in intra and extrahepatic biliary system

enhance during the portal phase, the thickened CBD wall in the region of the fibrotic stricture enhances in the equilibrium phase. The thickening of wall of biliary system and enhancement of the wall on dynamic scans may confuse the findings with that of cholangiocarcinoma.⁹ The biliary stagnation may lead to formation of secondary biliary or hepatic stones. Even though presence of proximal dilatation and biliary narrowing can be determined by US and CT, MRI is the modality of choice for diagnosing biliary changes.

Ancillary Findings

They are very uncommon and include splenomegaly and signs of cirrhosis such as nodularity of liver surface, atrophic medial segments of right lobe as well as hypertrophic caudate or lateral segments of left lobe, widening of fissures and coarsened echotexture or altered signal intensity of the liver.⁸ Any secondary pathology causing portal vein

obstruction like mass lesion or acute pancreatitis can be seen on imaging.

MULTIMODALITY IMAGING OF PCC

Ultrasound and Color Doppler

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 (Figure 2 a & b). A potential pitfall in gray scale images is to mistake the periportal collaterals in the hepatic hilum as an echogenic pseudotissular, well limited infiltration surrounding

Figure 2 (a) and (b): Gray scale and color Doppler ultrasound image at the level of porta showing non-visualization of portal vein (arrow) with multiple periportal collateral (cavernoma) formation. (c) and (d) Gray scale and color Doppler ultrasound image of gallbladder showing wall thickening (arrow) with multiple collaterals along gallbladder wall (gallbladder varices). (e) Spectral Doppler image of one of the collateral in gallbladder wall (arrow) showing continuous venous flow with phasic variation.

the CBD and this error can be avoided by using color Doppler imaging which clearly identifies the vascular nature of this abnormality. Similar tortuous collaterals can also be seen in or around the wall of the gallbladder, or in the gallbladder fossa in post cholecystectomy patients which are termed as gallbladder varices (Figure 2 c & d). They are seen in 30–55% of patients. They reduce the distensibility of gallbladder thus resulting in reduced fasting gallbladder volume. Very rarely collaterals can also be seen in the wall of the gallbladder and the bile duct, the so called mural varices which present as thickened wall with color flow on Doppler imaging. US distinguishes such varices from inflammatory or neoplastic wall thickening of biliary system, where both of them are seen as thickened wall with enhancement on cross-sectional imaging. Color Doppler images can sometimes be useful in demonstrating a direct communication between the portoportal collaterals and the intrahepatic portal vein branches. All these above mentioned collaterals are seen as a network of serpentine vessels and demonstrate low velocity continuous waveform on pulsed Doppler¹⁰ (Figure 2 e). It is also used to detect surgical shunts in post-operative patients of EHPVO. US is also capable of detecting narrowing or stenosis of the CBD with associated biliary duct dilatation proximal to the narrowing¹¹ and associated choledocholithiasis. However the exact level of narrowing and number of levels of narrowing are difficult to determine with US. Even though impression of collaterals on biliary system can be seen atleast on color Doppler imaging, it is difficult to differentiate between extrinsic compression and ischemic type of narrowing of CBD on ultrasound images. The CBD can also get obscured by high level echoes in the porta hepatis and by the multiple collateral channels. The signs of cirrhosis and splenomegaly are well detected with gray scale sonography with visualization of portosystemic collaterals on color Doppler imaging. However the main drawback of ultrasound is that it is operator dependant and the need for good window for visualization of structures which is usually hindered by overlying bowel gas and obesity. Although US with color Doppler can show most of the changes of portal biliopathy, cross-sectional imaging in the form of CT or MRI is usually required to confirm the disease and to rule out any biliary malignancy and search for possible cause of portal vein obstruction.

In a study of 46 patients with EHPVO,¹² the color Doppler imaging findings were correlated with ERCP and splenoportography in 3 patients. In all these cases, color Doppler easily distinguished the anechoic structures at the porta hepatis as periportal collaterals, the hepatic artery and dilated bile ducts. It revealed the indentations made on the CBD by the collaterals besides showing the presence of biliary calculi. Gallbladder varices were seen in all cases. In a retrospective study of 24 patients⁹ with cavernous transformation of the portal vein and ERCP correlation in 6 patients, biliary abnormalities were seen in 13

(54%) patients only. The CBD findings included thickening with luminal narrowing and narrowing with extrinsic compression.

Computed Tomography

CT demonstrates both vascular and biliary findings of PCC. The protocols which are recommended are single-phase studies (images obtained during the portovenous phase only), dual-phase scans performed during hepatic arterial phase and portovenous phase or triple phase scans performed during hepatic arterial phase, portal venous phase and equilibrium phase where portal venous phase is the phase which provides maximum amount of information. Postprocessing tools such as isotropic multiplanar reformation, maximum intensity projection, volume rendering techniques and surface shaded display are used extensively in demonstrating vascular and biliary changes. Maximum intensity projection, volume rendering techniques and surface shaded display demonstrate the network of collateral veins and its anatomic relationship with the bile ducts. They are also utilized in the demonstration of portosystemic collaterals. Minimum intensity projection images can provide cholangiographic like images similar to conventional studies and detect areas of narrowing and stenoses (Figure 3). 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.⁸ 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. The collaterals in perisplenic, perigastric, esophageal, retroperitoneal, gastrohepatic ligament are easily made out which are difficult to diagnose with sonography. CT can depict secondary biliary ductal dilatation caused by the portal collaterals. However the level, extent, number and cause of narrowing cannot be accurately depicted with CT like sonography. Any presence of choledocholithiasis can sometimes be made out. The main purpose of CT remains in ruling out other causes of biliary dilatation such as cholangiocarcinoma or extrinsic compression by metastatic lymphadenopathy (Figure 4). 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 are well demonstrated with CT. In a retrospective study of 11 patients on MRI ($n = 7$) and CT ($n = 4$), it was found that high resolution CT or MRI were comparable for providing anatomic detail of presence and severity of bile duct dilatation, portal vein

Figure 3 Extent of CBD changes on CT (a) MIP image shows CBD and intrahepatic biliary dilatation with biliary changes in the CBD, CHD and IHBRs (arrow). (b) Thick MIP image shows smooth narrowing of CBD and CHD (arrow). (c) Thick MIP image shows smooth narrowing confined to CBD (arrow).

Figure 4 CT imaging of mass-like portal cavernoma (a) Axial CECT image at level of porta showing portal cavernoma (PC) formation with non-visualized portal vein (long arrow). Multiple collaterals are also seen in gastrohepatic region and in perisplenic region (short arrow) with splenomegaly. (b) Axial CECT image showing biliary radical dilatation in right lobe of liver (arrow) due to portal biliopathy.

obstruction, portal cavernoma and portosystemic collaterals.⁸ However all the information about the vascular and biliary findings of PCC on CT comes at the expense of radiation. As the disease may require repetitive imaging, CT is not the preferred technique because of the risks of radiation exposure.

Magnetic Resonance Imaging

MRI at present has replaced direct cholangiography as the imaging investigation of choice for PCC with direct cholangiography being reserved for interventional purposes. The sequences to be acquired include routine T1WI and T2WI with magnetic resonance cholangiopancreatography (MRCP) and dynamic contrast enhanced MR (CEMR) images, also known as MR portography. MRCP images are obtained by acquiring heavily T2-weighted images in coronal plane centered in the biliary tree with reconstruction of maximum intensity projection images. They are useful in providing cholangiographic images which are comparable to direct cholangiographic images. MR portography images are obtained by acquiring axial volumetric three dimensional dynamic gradient echo images following gadolinium contrast administration with images obtained at arterial, portal venous and equilibrium phases. They are helpful in depicting the vascular abnormalities of the portal vein and their relationship with the biliary tract (Figure 5). They depict extension of portal vein thrombosis along with distribution of collaterals as described in CT.

The MR portography images also provide anatomical relation of biliary strictures with the periportal collaterals. This gives additional information about extraluminal pathology which is not given by direct cholangiographic studies. MR portography also delineates the entire splenoportal axis which helps to evaluate the feasibility of shunt surgery.¹³ Another advantage of dynamic MR is to distinguish between biliary ductal wall fibrosis and malignant wall thickening when both of them present with wall thickening. The malignant wall thickening shows early enhancement whereas fibrosis shows delayed enhancement. T1WI and T2WI show the collaterals and the portal vein, when patent, as flow voids. T2WI provide excellent information about biliary dilatation. These sequences also depict the features of cirrhosis if present. 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. The wavy appearance is most likely due to the extrinsic compression of CBD by periportal collaterals whereas biliary ductal and gallbladder wall thickening is due to either inflammatory fibrosis or intramural varices. Focal stenosis may be due to compression or ischemic narrowing (Figure 6). Lithiasis is due to bile stagnation proximal to the narrowing with secondary stone formation. There can be single or multiple and short or long smooth tapering bile duct strictures, segmental or global bile duct dilation, and filling defects.

Figure 5 MR imaging of portal cavernoma (a) Axial T2 WI and (b) Axial T1 WI image showing absence of portal vein flow void with multiple periportal collateral (PC) formation (arrow). (c) Coronal CEMRV image showing similar findings of portal cavernoma formation (arrow).

Figure 6 MR imaging of portal cavernoma (a) Axial CEMR image showing multiple periportal collaterals (cavernoma formation) (long arrow) with proximal biliary dilatation (short arrow) (due to portal cavernoma cholangiopathy). Multiple collaterals are seen in retroperitoneum (thick arrow) with splenomegaly. (b) MRCP image showing bilobar biliary dilatation (short arrows) with obstruction at level of common hepatic duct (long arrow) (due to portal cavernoma cholangiopathy).

In the clinical setting of long-standing portal hypertension and in the presence of para and epicholedochal varices, diagnosis can be made with confidence. Condat et al in their prospective study of 25 patients showed that findings at MRCP were similar to direct cholangiographic images in 4 patients in whom both the studies were done.¹⁴ In a retrospective study of 16 patients,¹⁵ biliary findings on MRCP included biliary stenosis in 93.7%, upstream dilatation in 68.7%, wavy appearance of the biliary tree in 87.5% and angulation of the CBD in 75% patients. None of the patients had choledocholithiasis. The frequencies of the ancillary vascular findings detected on contrast studies were gallbladder varices in 100% and choledochal varices in 93.7% patients. In a retrospective study of 12 patients,¹⁶ it was found that there was no discrepancy in the general contour or the level of the stricture and dilatation of the bile ducts between MRCP and direct cholangiography in five patients who had undergone comparative studies.

Chandra et al¹⁷ 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. This classification system can also be applied to MRCP images.¹⁸ Recent reports of MRCP imaging in PCC show that either type I or type III are the most frequent, with the extrahepatic bile ducts being the most common location of narrowing.¹⁵ It is important to note that MRCP can overestimate bile duct stenosis compared to conventional cholangiographic studies as was seen in a retrospective study of 10 patients of EHPVO with direct cholangiographic comparison in 3 patients.¹⁹

Llop et al²⁰ classified cholangiographic findings of 67 patients with portal vein thrombosis using MRCP. They classified PCC according to the severity of biliary dilatation where 6 patients had grade I (irregularities or angulations of the biliary tree), 12 patients had grade II (stenosis without dilation) and 34 patients had grade III (strictures with dilation) changes. The authors concluded that portal vein thrombosis was symptomatic in the most severe form of PCC.

Three patterns of biliary involvement on the basis of the pathogenesis, etiology and type of narrowing have also been described in PCC on MRCP.¹⁶ They are varicoid type, fibrotic type and mixed type. In the varicoid type the bile ducts show an irregular and wavy contour. This was proposed to be secondary to multiple smooth extrinsic indentations of dilated collateral veins on the biliary tract. This type may mimic a cholangiocarcinoma spreading along the bile duct giving rise to the pseudo-cholangiocarcinoma sign. In the fibrotic type there is presence of one or more segments (commonly one) of narrowing with proximal dilatation. This is secondary to the ischemic strictures. MR shows biliary wall thickening with delayed progressive enhancement in the late phase of dynamic contrast enhanced images. This is in contradiction to varicoid type which shows enhancement of CBD in portal phase due to the enhancement of pericholedochal collaterals. The CBD is the most common segment involved in this type whereas entire biliary system can be involved in the former type. In the mixed type, there are combined features of both varicoid and fibrotic type with biliary system showing both irregular and wavy contours with multiple areas of narrowing and dilatation. However the wall thickening at the level of the narrowed segment does not show delayed enhancement. Biliary calculi can be formed due to stasis

Table 2 Comparison of Different Imaging Modalities for Diagnosing Portal Cavernoma Cholangiopathy (PCC).

	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

CBD—common bile duct, CT—computed tomography, GB—gallbladder, MRCP—magnetic resonance cholangiopancreatography.

and are seen as signal voids within the ductal system on routine T1 and T2-weighted images. This should not be confused with pneumobilia which is also seen as signal voids within the biliary ducts especially in patients with prior ERCP. Another interesting finding seen in patients with PCC is transient zonal attenuation difference. This corresponds to increased enhancement of the peripheral hepatic parenchyma with relatively less enhancement of the central perihilar area seen in the arterial phase.²¹ The pathogenesis beyond this is that when blockage takes place at the level of the portal trunk, portal blood flow remains adequate for the central zones of the liver whereas not for the peripheral zone of liver. There is activation of arterial response which produces increased flow to the periphery of the liver in the arterial phase.

Walser et al¹ studied the CBD angle using MRCP images in 44 patients of EHPVO. They determined the CBD angle by placing lines along the axis of the extrahepatic bile duct using electronic protractor at the level of pancreatic head and above it, by using coronal thick-slab MRCP images. They found that there was kinking or tethering of the CBD at the superior portion of the pancreatic head seen more commonly in patients with portal biliopathy. There was a significant ($P = .008$) more acute angulation of the CBD at the superior border of the pancreatic head in the portal biliopathy patients (median—110°) than in the non portal biliopathy patients (median—128°). They postulated that the reason for this acute angulation of CBD at the superior edge of pancreatic head is the dilated anterior and posterior pancreaticoduodenal veins which develop in response to portal vein obstruction. These compress and displace the distal bile duct, causing angulation and proximal biliary dilatation.

Mimickers of PCC

Multiple non neoplastic and neoplastic conditions can cause biliary dilatation with strictures and mimic PCC. They include conditions like primary sclerosing cholangitis, recurrent pyogenic cholangitis, HIV cholangiopathy and neoplastic conditions like cholangiocarcinoma. The predominant findings in primary sclerosing cholangitis include multifocal strictures involving intra and extrahepatic ducts with beading and pruning of ducts along with segmental ectasia and ductal wall thickening. The vascular findings of portal cavernoma are usually absent. Cholangiocarcinoma presenting as biliary stricture and biliary wall thickening is usually confused with strictures of PCC. However, it can be differentiated by the pattern of enhancement, lymphnodal enlargement and absence of vascular findings. The mass-like enhancing portal cavernoma can sometimes be difficult to differentiate from mass forming cholangiocarcinoma.

CONCLUSION

Portal cavernoma cholangiopathy (PCC) is an entity associated with extrahepatic portal vein obstruction which is characterized by specific vascular, biliary and ancillary findings. Even though direct cholangiography has been the gold standard in the past, there is increased use of non-invasive modalities with a high degree of diagnostic accuracy. We recommend that US should be the initial imaging modality for suspected cases of PCC. MRCP with MR portography is the modality of choice for mapping of the biliary and vascular abnormalities (Table 2).

CONFLICTS OF INTEREST

All authors have none to declare.

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