Editorial

Imaging in portal cavernoma cholangiopathy: current understanding and future perspectives

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Comment on: Jabeen S, Robbani I, Choh NA, *et al.* Spectrum of biliary abnormalities in portal cavernoma cholangiopathy (PCC) secondary to idiopathic extrahepatic portal vein obstruction (EHPVO)-a prospective magnetic resonance cholangiopancreaticography (MRCP) based study. Br J Radiol 2016;89:20160636.

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Portal cavernoma cholangiopathy (PCC) as the name suggests refers to the spectrum of biliary changes that occur secondary to cavernomatous transformation of the portal vein in patients with extrahepatic portal vein obstruction (EHPVO) (1-5). PCC has previously been widely referred to as 'portal biliopathy' in medical literature but the current terminology is perhaps more readily comprehensible.

EHPVO is a disorder which is often not well understood presumably owing to its relatively low prevalence in the West as compared to the East. EHPVO is a distinct primary disorder, typically of the paediatric and young-adult population, characterized by chronic thrombosis of the extrahepatic portal vein. It may or may not involve the intrahepatic portal vein branches, the splenic and/or the superior mesenteric veins. EHPVO excludes other secondary causes of portal vein thrombosis such as liver cirrhosis and hepatic neoplasms (e.g., hepatocellular carcinoma) which may be associated with portal vein thrombosis (3). In the developing world, EHPVO constitutes one of the leading causes of non-cirrhotic portal hypertension, and variceal bleeding is typically the commonest presenting symptom. Opposed to liver cirrhosis the disorder has a relatively innocuous course, and with controlled variceal bleeding >10-year life-expectancy is as high as 100% (3-5).

PCC or biliary changes in patients with EHPVO are primarily sequelae of extrinsic compression due to engorgement of the two venous plexi of the bile ducts namely the paracholedochal plexus of *Petren* and the epicholedochal plexus of *Saint*, which serve as a collateral pathway to maintain antegrade flow to the liver (2,3). Other theories such as ischemic insult, inflammation and/or fibrosis of the biliary tree due to prolonged biliary compression have also been put forth. It is important to underscore that on imaging nearly all patients (81–100%) with EHPVO manifest biliary changes termed as 'portal cavernoma cholangiopathy' but only a small percentage (5–30%) manifest clinical symptoms. Symptomatic PCC although relatively uncommon can seriously impair the patient's quality of life secondary to chronic cholestasis, cholangitis, stone disease, and/or biliary cirrhosis (4,5). Asymptomatic PCC does not need any intervention, but those with symptomatic, advanced or severe PCC might need either endoscopic (e.g., biliary stenting, biliary sphincterotomy), radiological (e.g., percutaneous transhepatic biliary drainage) or surgical intervention (e.g., hepaticojejunostomy or choledochoduodenostomy) depending upon the complexity of symptoms. Indubitably, timely diagnosis and appropriate management can avert complications and improve the patient's quality of life.

Jabeen *et al.* (6) have indeed done a commendable job to appraise and collate the spectrum of biliary abnormalities affecting the intra-and extrahepatic biliary tree in patients with PCC. The study elucidates various salient MR features of PCC in their study population of 52 patients. Contrary to what we would generally expect, 'scalloping' or 'wavy' contour of the extrahepatic bile duct (secondary to indentations by the dilated peribiliary venous plexi) was

not the commonest finding but the second most common abnormality detected in 76%. This was preceded by increased angulation of the common duct identified in as many as 90% of the patients. Whether it is 'kinking', 'wavy' contour or 'smooth impression', all are considered to be sequelae of extrinsic compression by the dilated paracholedochal venous channels, thus reaffirming our current understanding of the etiopathogenesis of PCC. Whilst the smaller peri-biliary veins are believed to cause 'scalloping', the larger anterior and posterior superior pancreaticoduodenal veins encircling the bile duct are presumed to produce kinking (7). The potential role of fibrosis in kinking however cannot be entirely negated. Both scalloping/indentation and kinking have earlier been reported as the imaging hallmarks of PCC on ERCP as well as MRI (7-9).

Other important manifestation of PCC is the development of common duct stricture presumably secondary to ischemia and/or fibrosis; this was identified in 14.1% although the incidence could be higher (25–50%) as has been shown in earlier studies (9). This is the subset of PCC which one has to be careful before inadvertently labelling as cholangiocarcinoma. Similar finding during the era of direct cholangiography earned it the nomenclature 'pseudo cholangiocarcinoma sign'. Evaluation of MRCP in conjunction with contrast-enhanced cross-sectional imaging can avert this erroneous diagnosis. Also, this variant of PCC, previously also called as fibrotic type by Shin *et al.* (9), becomes relevant as the current study shows a significant association between choledocholithiasis and common bile duct stricture which may predispose the patient to developing symptoms such as jaundice and cholangitis. Likewise, the authors also found a significant association between hepatolithiasis and choledocholithiasis but not between choledocholithiasis and cholelithiasis. These findings become pertinent from the point of view of clinical management and prognosis. However, as the authors have rightly pointed out larger study population and a longer-term follow-up would be required to further consolidate their findings.

It has been previously reported that those with portal plus mesenteric vein thrombosis are much more prone to develop PCC than those with portal vein involvement alone (7). Similarly, it should be a subject of future studies to see if the extent of splanchnic venous axis involvement has any relationship with the differential involvement of the intra-and extrahepatic biliary tree in PCC. Jabeen *et al.* (6) reported isolated extrahepatic biliary tree involvement in 26.2% (Chandra *et al.*, type I), isolated intrahepatic bile duct involvement in 4.8% (type II) and both intra-and extrahepatic involvement (type III) in 69% of patients. It would have been interesting if the authors had correlated these biliary changes with the extent of cavernomatous transformation identified on CT spleno portal venography (which was performed to confirm EHPVO). Whether the cavernomatous transformation remains confined to the extrahepatic portal vein in those with type I changes, and whether or not the intrahepatic portal vein branches are involved in those with type II and III changes would be interesting to know.

The current study has quite thoroughly evaluated the intrahepatic bile duct changes on MRI and their findings corroborate with those in the earlier literature. Interestingly, the current study too demonstrates preferential involvement of the left hepatic duct which has been believed to be related to the formation of prominent collaterals where the umbilical vein joins the left branch of the portal vein. Correlative analysis of MRCP and contrast enhanced studies in a larger study population could potentially be helpful in endorsing this proposition.

With increasing temporal and spatial resolution of MRI, another potential area of research would be to assess for gallbladder wall changes that may be detectable on MRCP (e.g., scalloping) as the development of gallbladder wall varices via the cystic vein have been previously found to be a characteristic feature of PCC (10), although this may be quite challenging.

Finally, one ought to be aware of the solid, 'tumour-like' fibrotic variant of PCC that may be occasionally encountered in clinical practice (3,8). This was probably not encountered in the current study owing to its rarity, and relatively small patient population. Although rare this 'pseudotumoral' entity needs a special mention because if inadvertently biopsied can bleed the patient to death (7,8). Abundant connective tissue proliferation around individual periductal veins can occasionally simulate a solid hilar mass and pose a diagnostic dilemma. However, awareness of this entity and review of the dynamic contrast (especially portal venous phase) images can help clinch the correct diagnosis and save the patient from catastrophic effects of a potential biopsy (3).

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