Received:
24 July 2016

Revised:
15 September 2016

Accepted:
6 October 2016

<https://doi.org/10.1259/bjr.20160636>

Cite this article as:

Jabeen S, Robbani I, Choh NA, Ashraf O, Shaheen F, Gojwari T, et al. Spectrum of biliary abnormalities in portal cavernoma cholangiopathy (PCC) secondary to idiopathic extrahepatic portal vein obstruction (EHPVO)—a prospective magnetic resonance cholangiopancreatography (MRCP) based study. *Br J Radiol* 2016; **89**: 20160636.

FULL PAPER

Spectrum of biliary abnormalities in portal cavernoma cholangiopathy (PCC) secondary to idiopathic extrahepatic portal vein obstruction (EHPVO)—a prospective magnetic resonance cholangiopancreatography (MRCP) based study

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Objective: To characterize biliary abnormalities seen in portal cavernoma cholangiopathy (PCC) on MR cholangiopancreatography (MRCP) and elucidate certain salient features of the disease by collaborating our findings with those of previous studies.

Methods: We prospectively enrolled 52 patients with portal cavernoma secondary to idiopathic extrahepatic portal vein obstruction, who underwent a standard MRCP protocol. Images were analyzed for abnormalities involving the entire biliary tree. Terms used were those proposed by the Indian National Association for Study of the Liver. Angulation of the common bile duct (CBD) was measured in all patients with cholangiopathy.

Results: Cholangiopathy was seen in 80.7% of patients on MRCP. Extrahepatic ducts were involved in 95% of patients either alone (26%) or in combination with the intrahepatic ducts (69%). Isolated involvement of the intrahepatic ducts was seen in 4.8% of patients. Abnormalities of the extrahepatic ducts included angulation (90%), scalloping (76.2%), extrinsic impression/indentation (45.2%), stricture (14.3%) and smooth dilatation (4.8%). The mean CBD angle was $113.2 \pm 19.8^\circ$. Abnormalities of the intrahepatic ducts included

smooth dilatation (40%), irregularity (28%) and narrowing (9%). Cholelithiasis, choledocholithiasis and hepatolithiasis were seen in 28.6% (12) patients, 14.3% (6) patients and 11.9% (5) patients, respectively. There was a significant association between choledocholithiasis and CBD stricture, with no significant association between choledocholithiasis and cholelithiasis. A significant association was also seen between hepatolithiasis and choledocholithiasis.

Conclusion: The spectrum of biliary abnormalities in PCC has been explored and some salient features of the disease have been elucidated, which allow a confident diagnosis of this entity.

Advances in knowledge: PCC preferentially involves the extrahepatic biliary tree. Changes in the intrahepatic ducts generally occur as sequelae of involvement of the extrahepatic ducts, although isolated involvement of the intrahepatic ducts does occur. Increased angulation of the CBD and scalloping are most commonly seen. Angulation may predispose to choledocholithiasis and thus development of symptomatic cholangiopathy. Choledocholithiasis and hepatolithiasis occur as sequelae of PCC.

INTRODUCTION

Portal cavernoma cholangiopathy (PCC) is the term proposed by the Indian National Association for Study of the Liver in a consensus statement issued in 2014 for biliary abnormalities in patients with portal cavernoma.¹ It is defined as “abnormalities in the extrahepatic biliary system including the cystic duct and gall bladder with or without abnormalities in the first and second generation biliary ducts in a patient with portal cavernoma”.¹ The presence of a portal cavernoma, typical cholangiographic changes on endoscopic or MR cholangiography and the absence of

other causes of biliary changes such as bile duct injury, primary sclerosing cholangitis, cholangiocarcinoma etc. are mandatory to arrive at a diagnosis.¹ Frequency of PCC has been reported to be much greater in patients with idiopathic extrahepatic portal vein obstruction (EHPVO) (80–100%) as compared with cirrhotic liver disease (0–33%) and non-cirrhotic portal fibrosis (9–40%).^{2–4} In addition, pancreatic duct abnormalities were observed in a large proportion of patients with portal vein cavernous transformation.⁵ An early onset of portal vein thrombosis due to umbilical sepsis and long duration of portal

Table 1. MR cholangiopancreaticography imaging parameters

Parameters	Thin-slab T_2 HASTE axial breath-hold	Thin-slab T_2 HASTE coronal breath-hold	Thick-slab T_2 HASTE breath-hold	True FISP axial breath-hold	True FISP coronal breath-hold
TR/TE (ms)	900/92	900/82	4500/754	3.09/1.31	3.09/1.31
Slice thickness (mm)	5	3.5	40	6	6
Number of slices	25	30	1	30	30
Field of view (mm)	285 × 380	318 × 400	349 × 349	276 × 340	318 × 340
Matrix size	168 × 320	179 × 320	307 × 384	182 × 320	210 × 320

HASTE, half-Fourier acquisition with single-shot turbo spin-echo; TE, echo time; TR, repetition time ; True FISP, true fast imaging with steady-state free-precession.

hypertension in patients with idiopathic EHPVO are likely to be contributory.^{2,10} PCC is an increasingly important complication of EHPVO, which impacts the natural history and prognosis of the disease.

Various aetiological factors have been proposed to explain the occurrence of biliary abnormalities in patients with portal cavernoma. Collateral compression, ischaemia or a combination of both are largely in acceptance. Collateral compression occurs secondary to dilatation of the venous plexuses around the common bile duct (CBD) in response to portal vein thrombosis, leading to fine irregularities due to dilatation of the intramural epicholedochal plexus of Saint and/or extrinsic compression due to dilatation of the extramural paracholedochal plexus of Petren.^{6,7,8,9} Besides, it has been proposed that ischaemia occurs as a result of portal vein thrombosis, leading to fibrosis and strictures.^{11–15} Ischaemic theory is supported by the observation that in several patients with EHPVO, the biliary abnormalities are not completely reversed following shunt surgery.^{14,16} An infectious aetiology has also been proposed by various workers.²

There is a varied spectrum of biliary abnormalities in PCC. Many a time, it has posed a diagnostic dilemma by being mistaken for malignancy. In this study, we present the biliary abnormalities seen on MR cholangiopancreaticography (MRCP) in a series of 52 patients with portal cavernoma secondary to idiopathic EHPVO at our centre.

METHODS AND MATERIALS

It was a prospective study carried out in the Department of Radiodiagnosis and Imaging at the Sher-i-Kashmir Institute of Medical Sciences, Srinagar, Kashmir, India, with approval from the Institutional Ethics Committee.

Subjects

Inclusion criteria: patients who had an established diagnosis of idiopathic EHPVO with cavernous transformation of the portal vein based on clinical, laboratory and imaging findings (Doppler and CT splenoportovenography) were prospectively enrolled for the study. They were referred from the departments of medical and surgical gastroenterology. We enrolled 52 patients, who

prospectively underwent MRCP irrespective of age after obtaining informed consent.

Exclusion criteria:

- (1) patients with underlying pancreaticobiliary malignancy
- (2) liver cirrhosis
- (3) patients who were diagnosed cases of oriental cholangiohepatitis
- (4) pregnant females
- (5) patients with pacemaker, metallic implants or severe claustrophobia were excluded from the study.

Clinical information

Clinical records were reviewed for the presence and frequency of symptoms pertaining to cholangiopathy, which include abdominal pain, yellowish discolouration of eyes, prior history of cholangitis and/or acute pancreatitis. None of the patients had undergone cholecystectomy. Relevant laboratory data with emphasis on serum alkaline phosphatase (ALP) and serum

Figure 1. A coronal thick-slab MR cholangiopancreaticography image is showing marked focal narrowing of the common bile duct (CBD) (small arrow) with angulation (long arrow). Lines placed across the duct are showing measurement of the CBD angle. In this case, the angle is $<90^\circ$.

Table 2. Age and gender of each patient with portal cavernoma cholangiopathy on MR cholangiopancreatography

Serial number	Gender	Age (years)
1	Male	23
2	Male	54
3	Male	17
4	Female	20
5	Female	28
6	Male	27
7	Male	15
8	Female	50
9	Male	40
10	Male	38
11	Female	32
12	Male	45
13	Female	25
14	Female	20
15	Male	40
16	Female	02
17	Female	30
18	Female	25
19	Male	39
20	Female	45
21	Female	25
22	Female	16
23	Male	20
24	Female	21
25	Male	24
26	Female	45
27	Female	55
28	Female	16
29	Male	37
30	Male	26
31	Male	34
32	Male	32
33	Female	30
34	Male	55
35	Female	37
36	Female	62
37	Female	65
38	Male	37
39	Male	19

(Continued)

Table 2. (Continued)

Serial number	Gender	Age (years)
40	Female	25
41	Male	40
42	Male	35

bilirubin levels were obtained. The normal reference range for serum ALP was taken as 44–147 IU ml⁻¹ and for serum bilirubin as 0.3–1.9 mg dl⁻¹.

Imaging technique

MRCP was carried out on a superconductive 1.5-T scanner (Magnetom Avanto; Siemens, Munich, Germany) using a phased-array multicoil. Thin-slab and thick-slab half-Fourier acquisition with single-shot turbo spin-echo (HASTE) sequences were obtained using breath-hold in patients who were co-operative and free breathing with respiratory navigation in those who could not co-operate. Thin-slab sequences were obtained in the axial and coronal planes with a slice thickness of 5 and 3–4 mm, respectively. Thick-slab sequences were obtained in the coronal plane with a slice thickness of 40 mm. True fast imaging with steady-state free-precession sequence was obtained in the axial and coronal planes with a slice thickness of 5 and 3–4 mm, respectively, to assess the portosplenic axis and demonstrate collateral compression around the CBD. The imaging parameters used are tabulated in Table 1.

Image analysis

The images were analyzed on a Siemens workstation using syngo software. The biliary tree was examined for any abnormalities and the following were documented:

- (1) pattern of ductal involvement classified as:
 - extrahepatic: where the changes and dilatation were confined to the extrahepatic biliary tree beyond the hepatic hilum
 - intrahepatic: where the changes were confined to the intrahepatic ducts, *i.e.* the right hepatic duct (RHD) and left hepatic duct (LHD), their confluence and the second and third biliary radicles without any noticeable abnormality or dilatation of the extrahepatic biliary tree
 - both extrahepatic and intrahepatic: where changes were seen in the extrahepatic biliary tree with dilatation and abnormalities of the intrahepatic ducts as well.
- (2) Degree of dilatation: the intrahepatic ducts were considered to be dilated when the diameter of either the right or left intrahepatic duct or both exceeded 4 mm. The degree of dilatation was classified as:
 - mild: 4–6 mm
 - moderate: 6.1–8 mm
 - severe: >8 mm.
- (3) Ductal diameters: the ductal diameters were measured using electronic calipers on thick-slab coronal HASTE images. The RHD and LHD were measured at the site of maximal

Figure 2. A bar graph is showing the genderwise age distribution in our patients. Mean age in males was 33 years; mean age in females was 32 years. There was no significant difference in age distribution between the genders (p -value = 0.483; by Fisher's exact test).

dilatation proximal to the confluence. The common hepatic duct (CHD) and CBD were measured at the site of maximum dilatation proximal and distal to the cystic duct insertion, respectively.

(4) Changes in the intrahepatic ducts: the intrahepatic ducts were considered to be normal when there was no dilatation, irregularity, narrowing or periductal thickening. Abnormal intrahepatic biliary radicles were described as follows:

- smooth dilatation: when there was no evidence of any wall thickening, irregularity or focal narrowing and the intrahepatic ducts showed uniform dilatation along their length
- periductal thickening with irregularity: periductal thickening was identified as the presence of T_2 hyperintense (hyperintense relative to liver parenchyma) thickening around the intrahepatic ducts.

Irregular intrahepatic ducts showed a fine wavy contour without evidence of frank narrowing or stenosis.

- Narrowing/stenosis: was seen as an abrupt change in the calibre of the duct with upstream dilatation.

(5) Changes in the extrahepatic ducts: these were documented using the following terms proposed by the Indian National Association for Study of the Liver:¹⁷

- angulation: degree of angulation of the CBD usually seen at the superior border of the pancreas was measured using electronic calipers. Coronal thick-slab HASTE sequences were used for the purpose. Lines were placed along the long axis of the duct centrally across the lumen above and below the site of angulation and the angle measured (Figure 1). An angle $<145^\circ$ was regarded as significant (in the study by Keizman *et al*,²⁹ in which an angle $\leq 145^\circ$ was an

Figure 3. A bar graph is showing the proportion of patients who are symptomatic to patients who are asymptomatic in those with deranged lab parameters: the ratio of patients who are symptomatic to those who are asymptomatic was 1.7:1 among patients with raised alkaline phosphatase (ALP), 1.27:1 among those with raised bilirubin and 1.5:1 among those with elevation of both parameters.

Figure 4. A pie chart is showing the type of intrahepatic duct abnormalities seen in our patients.

independent risk factor for symptomatic bile duct stone recurrence in a cohort of 232 patients).

- Smooth dilatation: uniformly dilated duct without any evidence of irregularity, narrowing or stenosis. Ductal diameter >7 mm was taken as dilated.
- Scalloping: multiple non-contiguous smooth impressions on the CBD less than one-fourth of the ductal diameter leading to an irregular wavy contour of the duct
- extrinsic impressions/indentations: thumb-like impressions on the bile duct more than one-fourth of the diameter of the duct leading to a nodular contour of the duct
- stricture: narrowing of the ductal lumen with upstream dilatation and normal downstream segment.

(6) Calcular disease: the gall bladder, intrahepatic and extrahepatic ducts were carefully examined for the presence of intraluminal signal voids suggestive of calculi. Signal voids due to calculi were seen as dependent (posteriorly) on axial

Figure 5. An axial T_2 half-Fourier acquisition with single-shot turbo spin-echo image is showing periductal hyperintense thickening (white arrows) with dilated left hepatic ducts (black arrow).

Figure 6. An axial thin-slab T_2 half-Fourier acquisition with single-shot turbo spin-echo image in a patient with extrahepatic portal vein obstruction is showing grossly dilated right lobe intrahepatic ducts with an intraluminal signal void suggestive of calculus (black arrow) with periductal thickening (white arrow). Note can be made of hypointense foci representing Gamna-Gandy bodies in the splenic parenchyma.

images with a thin rim of hyperintense bile anteriorly unlike air, which is non-dependant. In cases where the confusion persisted, few focused CT slices were obtained to confirm the presence of calculi.

(7) Classification and grading: the disease was classified according to the system proposed by Chandra et al² as Type I involving the extrahepatic ducts only, Type II involving the intrahepatic ducts alone and Type III, which involved the extrahepatic as well as intrahepatic ducts. Grading was performed as proposed by Llop et al¹⁸ as:

- Grade I: irregularities or angulations of the biliary tree
- Grade II: indentations or strictures without dilatation
- Grade III: strictures with dilatation.

Figure 7. An axial contrast-enhanced CT image obtained during the portal venous phase is showing intrahepatic extension of the portal cavernoma (long arrow) with dilated right hepatic duct (short arrow) and dilated portal vein radicles (arrowhead) along the dilated duct.

Table 3. Ductal diameters (in millimetres)^a

	Mean $\pm$ SD	Minimum	Maximum	Percentile		
				25	50	75
LHD	6.855 $\pm$ 4.23	2.0	19	3.67	5.75	8.42
RHD	5.029 $\pm$ 3.36	1.0	18	2.45	3.95	6.60
CHD	6.76 $\pm$ 3.18	2.8	15	4.35	5.90	8.92
CBD	5.690 $\pm$ 2.62	2.0	14	4.00	5.20	6.92

CBD, common bile duct; CHD, common hepatic duct; LHD, left hepatic duct; RHD, right hepatic duct; SD, standard deviation.

LHD/RHD diameter: Wilcoxon signed-ranks test; significance at p -value = 0.

CHD/CBD diameter: Wilcoxon signed-ranks test; significance at p -value = 0.015.

^aDifference among ductal diameters: Friedman and Kendalls W test; significance at p -value = 0.

Statistical analysis

Statistical analysis was carried out using the IBM statistical package for social sciences SPSS® v. 20 (IBM Corp., New York, NY; formerly SPSS Inc., Chicago, IL). Categorical variables were summarized in terms of frequencies and percentages. Quantitative variables, which included ductal diameters and CBD angle, were assessed in terms of mean, median, mode, standard deviation, maximum, minimum and percentile values. Histograms with frequency distribution curves and box plots were used for the quantitative variables. Association between two categorical variables was assessed using the Fisher's exact test. Quantitative variables were compared using the Wilcoxon signed-ranks test. A p -value <0.05 was taken as significant.

RESULTS

42 (80.76%) out of the 52 enrolled patients showed evidence of biliary abnormalities on MRCP. More than half of the patients (57.1%) belonged to the 21–40-year age group with a mean age of 32.6 ± 13.78 years. The age and gender of individual subjects are tabulated in Table 2. There was an equal proportion of males and females with no significant difference in the distribution of age groups between the two genders. The genderwise age distribution is depicted in Figure 2.

Clinical records of 15 (36%) patients revealed the presence of symptoms and/or complications pertaining to cholangiopathy, which included abdominal pain mostly localized to the epigastrium or right hypochondrium in 86.6% (13) patients and

Figure 8. Box plot summary of ductal diameters is showing higher mean diameters in the left hepatic duct (LHD) and common hepatic duct (CHD) as compared with the right hepatic duct (RHD) and common bile duct (CBD), respectively. Patients with large ductal diameters represented by the outliers had CBD strictures, choledocholithiasis or hepatolithiasis.

yellowish discolouration of eyes in 73% (11) patients. There was a history of cholangitis in 6 (40%) patients and admission for acute pancreatitis in 1 patient. In 27 (64%) patients, there were no symptoms that could be attributed to cholangiopathy. The ratio of patients who were symptomatic to those who were asymptomatic was 1:1.8.

The mean ALP level was $189 \pm 14 \text{ IU ml}^{-1}$. 30 (71.4%) patients had modest elevations in ALP, which were less than twice the upper limit of normal (normal reference values $44\text{--}157 \text{ IU ml}^{-1}$). Only six patients had ALP values more than two times the upper limit of normal. The mean serum bilirubin level was

$2.1 \pm 0.78 \text{ mg dl}^{-1}$. 25 (59.5%) patients had mild elevations in the serum bilirubin level between 2 and 3 mg dl^{-1} . 20 (47.6%) patients had both elevated ALP and serum bilirubin levels. The proportion of patients who were symptomatic and those who were asymptomatic among those with deranged lab parameters is shown in Figure 3.

Extrahepatic biliary tree showed abnormalities in 95.2% (40) of patients either alone in 26.2% (11) patients or along with the intrahepatic ducts in 69% (29) patients. Only 2 (4.8%) patients showed isolate involvement of the intrahepatic ducts with a normal extrahepatic biliary tree. According to the classification

Figure 9. (a–d) Frequency distribution curves of ductal diameters [a—left hepatic duct (LHD), b—right hepatic duct (RHD), c—common hepatic duct (CHD) and d—common bile duct (CBD)] are revealing a wide range of distribution due to varying ductal involvement. The severity of involvement varies from patient to patient as reflected in the given figure. For the LHD, 57% of the values were between 4 and 10 mm. 7 (16%) patients had LHD diameters >10 mm. 11 (26%) patients had LHD diameters <4 mm. 42.8% of patients showed RHD diameters in the range of 4–10 mm, with three patients having RHD diameters measuring >10 mm. CHD diameters were >5 mm in 67% of patients, whereas only 50% of patients had CBD diameters above 5 mm. Significant difference was noted between the LHD/RHD diameters and CHD/CBD diameters using the Wilcoxon signed-ranks test.

Figure 10. A bar graph is showing the distribution of extrahepatic duct abnormalities with angulation of the common bile duct being the most common finding.

system proposed by Chandra et al based on the pattern of ductal involvement, Type I was seen in 26.2% patients, Type II in 4.8% patients and Type III in 69% of patients.

In 26.2% of patients, no dilatation of the intrahepatic ducts was seen. Mild dilatation (4–6 mm) was seen in 23.8% patients, moderate dilatation (6.1–8 mm) in 21.4% patients and severe dilatation (>8 mm) in 28.6% patients.

Abnormalities seen in the intrahepatic ducts are summarized in Figure 4 with representative images (Figures 5–7).

Ductal diameters are summarized in Table 3 along with the box plot and frequency distribution curves in Figures 8 and 9, respectively. The LHD showed a larger mean diameter than the RHD (6.85 ± 4.23 vs 5.02 ± 3.36 mm), with a statistically significant difference using the Wilcoxon signed-ranks test (p -value <0.001). Also, the CHD showed a greater degree of dilatation than the CBD (6.76 ± 3.18 vs 5.69 ± 2.62 mm), which was statistically significant (p -value <0.05).

Abnormalities seen in the extrahepatic ducts are summarized in Figure 10. The most frequently observed abnormality was an increased angulation of the CBD seen in 90% of patients. The CBD angle seen in these patients is summarized in Table 4 and in the frequency distribution curve in Figure 11. The mean CBD angle was $113.2 \pm 19.8^\circ$ with a range of 60–150°. Only two

patients had angles $\geq 145^\circ$. Extrahepatic duct abnormalities are shown in Figure 1 and 12–20.

Calcular disease in the form of cholelithiasis, choledocholithiasis and hepatolithiasis was seen in 28.6% (12) patients, 14.3% (6) patients and 11.9% (5) patients, respectively (Figures 6 and 17). The biliary abnormalities and the presence/absence of cholelithiasis in patients with choledocholithiasis and hepatolithiasis are summarized in Tables 5 and 6. There was no significant association between choledocholithiasis and cholelithiasis (p -value = 1.000; Fisher's exact test). Statistically significant association was seen between choledocholithiasis and CBD stricture (p -value = 0.029), hepatolithiasis and CBD stricture (p -value = 0.015) as well as between hepatolithiasis and choledocholithiasis (p -value = 0.015) using the Fisher's exact test.

According to the grading system proposed by Llop et al, Grade I was seen in 11.9% (5) patients, Grade II in 14.2% (6) patients and Grade III in 73.8% (31) patients.

DISCUSSION

PCC is a recognized complication of EHPVO.²⁰ With advances in the management of variceal bleeding and improved survival, increasing number of cases are being identified. MR cholangiography is the imaging modality of choice in diagnosing these cases.¹

Table 4. Common bile duct (CBD) angle

	Mean $\pm$ SD	Minimum	Maximum	Percentile		
				25	50	75
CBD angle	$113.21 \pm 19.8^\circ$	60°	150°	103.75°	115.00°	125°

SD, standard deviation.

Figure 11. Frequency distribution curve of common bile duct (CBD) angle in our patients: mean CBD angle in the study group was 113.2° with a range of 60–150°. 67% of the values lie between 80 and 120°. Only two patients had angles equal to or <145°.

Biliary changes have been documented on cholangiography (endoscopic retrograde cholangiopancreatography (ERCP)/MRCP) in 78–100% of patients with idiopathic EHPVO.^{2,11,18–23}

We identified biliary abnormalities in 81% of our patients. Most of the patients were asymptomatic (64%) with mild elevations in ALP and serum bilirubin levels. 36% of patients had symptoms, the commonest being abdominal pain. Six patients had episodes of recurrent cholangitis, which indicates that there is a risk of

development of complications thus influencing morbidity and mortality. Most of the studies have reported symptomatic cholangiopathy in 5–38% of patients with EHPVO.^{4,16,20,24,25}

Although there is a varied spectrum of biliary abnormalities in patients with PCC, certain features increase the likelihood of diagnosis. PCC has a predilection for the extrahepatic ducts which were involved in nearly all the patients (95.2%). Isolated involvement of the intrahepatic ducts as is often seen in oriental

Figure 12. A thin-slab T_2 half-Fourier acquisition with single-shot turbo spin-echo coronal MR cholangiopancreatography image in the same patient as in Figure 1 is showing acute angulation of the common bile duct (CBD) (long white arrow) with a large collateral impinging on the CBD (small white arrow). The black arrow is showing focal dilatation of the common hepatic duct.

Figure 13. A coronal thick-slab T_2 half-Fourier acquisition with single-shot turbo spin-echo image is showing common bile duct angulation (white arrow) and shallow common hepatic duct impressions (white arrowhead) with cystic duct scalloping (black arrow).

Figure 14. A coronal thick-slab T_2 half-Fourier acquisition with single-shot turbo spin-echo image is showing shallow impressions (short arrows) and indentation (long arrow) along the common duct wall.

cholangiohepatitis is rare (4.8% in our study). Extrahepatic duct involvement has been consistently reported in cholangiographic studies carried out in patients with EHPVO.^{11,19–21,24–28} Chandra et al² reported Type II disease in only 2 patients, whereas Type I and Type III were seen in 11 and 29 patients, respectively, on ERCP.

Figure 15. A thin-slab T_2 half-Fourier acquisition with single-shot turbo spin-echo image obtained in the coronal plane is showing large flow voids (short arrows) surrounding the common duct (long arrow) causing its kinking, narrowing and displacement.

Figure 16. An axial true fast imaging with steady-state free-precession image in the same patient as in Figure 15 is showing large hyperintense paracholedochal collateral vessels surrounding the common bile duct (arrow).

An increased angulation of the CBD with scalloping is typically seen in these patients. We obtained a mean angle of 113° in our patients with PCC. Walser et al³ compared the CBD angle in patients with EHPVO with and without biliopathy. They reported a significantly increased angulation of the CBD in patients with biliopathy (mean 128° ; p -value = 0.008). This was attributed to the dilatation of the anterior and superior pancreaticoduodenal veins in response to portal vein thrombosis, which compresses and displaces the bile duct superior to the pancreas. Studies by Keizman et al²⁹ and Warren³⁰ have shown that increased angulation of the CBD predisposes to the development of choledocholithiasis. Thus, the CBD angle may be an important predictor for the development of choledocholithiasis and hence symptomatic cholangiopathy in patients with EHPVO. In our study, six patients with choledocholithiasis had a mean CBD angle of 100.8° against a mean CBD angle of 113° in the entire series.

Figure 17. A coronal thick-slab T_2 half-Fourier acquisition with single-shot turbo spin-echo image is showing angulation and focal narrowing of the common bile duct (black arrow) with intraluminal signal voids suggestive of calculi (white arrows) proximal to the site of narrowing with upstream dilatation.

Figure 18. An axial thin-slab T_2 half-Fourier acquisition with single-shot turbo spin-echo image is showing a thick-walled gall bladder (black arrow) with paracholedochal collaterals (short white arrows) seen as signal voids around the common bile duct (long white arrow). Note can be made of the splenic hilar collaterals (grey arrow) with splenomegaly.

The intrahepatic ducts are not entirely spared in PCC. Abnormalities observed included upstream dilatation due to diseased extrahepatic ducts, narrowing and periductal thickening. The thickening could be secondary to small intramural varices or fibrosis due to ischaemia. Intrahepatic ductal abnormalities have been described in earlier studies in the form of dilatation, focal narrowing and irregularity.^{11,20,21,25–28}

The CHD and LHD showed a greater degree of dilatation than the CBD and RHD, respectively. We observed a preferential

Figure 19. A coronal thin-slab T_2 half-Fourier acquisition with single-shot turbo spin-echo image is showing a dilated common hepatic duct (white arrow) with abrupt change in calibre at the superior border of the pancreas (black arrow). There is no evidence of paracholedochal collaterals suggestive of an ischaemic aetiology. Note can be made of splenomegaly.

Figure 20. An axial true fast imaging with steady-state free-precession image is showing a large pancreaticoduodenal collateral vessel (white arrow) partially encircling the common bile duct (black arrow).

involvement of the supraduodenal bile duct and the LHD over the RHD. Relatively poor vascularization of the supraduodenal bile duct predisposing it to ischaemia and more extensive collateral circulation around the LHD where the umbilical vein joins the left branch of the portal vein have been proposed as plausible explanations.^{31–33}

Calcular disease was also seen in our patients in the form of cholelithiasis, choledocholithiasis and hepatolithiasis and has been described in other studies as well. Cholelithiasis in the setting of PCC may be due to chronic cholestasis, changes in the lithogenicity of the bile or other factors such as reduced portal flow and associated liver atrophy, as proposed by Harmanci and Bayraktar.³⁴ Reduced contractile function of the gall bladder has also been proposed to contribute to the development of gallstones due to the presence of gall bladder wall collaterals. Patients with asymptomatic PCC may not have a critical level of extrahepatic biliary obstruction resulting in abnormal emptying of the gall bladder and bile lithogenicity.³⁵ The significant association between choledocholithiasis and CBD stricture with no significant association with cholelithiasis suggests that these patients develop primary CBD stones secondary to stricture as

Table 5. Occurrence of cholelithiasis and type of common bile duct (CBD) abnormalities in six patients with choledocholithiasis^a

Patients	Cholelithiasis	CBD abnormalities
1	Present	Scalloping
2	Absent	Scalloping
3	Present	Stricture
4	Absent	Stricture
5	Absent	Stricture
6	Absent	Smooth dilation

^a p -value for cholelithiasis/choledocholithiasis is 1.000; p -value choledocholithiasis/CBD stricture is 0.029 (Fisher's exact test).

Table 6. Occurrence of cholelithiasis, choledocholithiasis and type of intrahepatic and common bile duct (CBD) abnormalities in five patients with hepatolithiasis^a

Patients	Cholelithiasis	Choledocholithiasis	Intrahepatic duct abnormality	CBD abnormality
1	Absent	Absent	Irregularity with periductal thickening	Scalloping
2	Present	Present	Irregularity with periductal thickening	Scalloping
3	Absent	Absent	Irregularity with periductal thickening	Stricture
4	Absent	Present	Narrowing	Stricture
5	Absent	Present	Narrowing	Stricture

^a*p*-value for hepatolithiasis/choledocholithiasis is 0.015; *p*-value for hepatolithiasis/CBD stricture is 0.015 (Fisher's exact test).

sequelae of portal cavernoma rather than in association with cholelithiasis. In our study, hepatolithiasis was associated with extrahepatic ductal abnormalities in all the patients. This suggests that intraductal calculi in PCC occur secondary to bile stasis as a result of extrahepatic biliary obstruction unlike oriental cholangiohepatitis, where intraductal calculi often occur in the presence of a normal extrahepatic biliary tree. There have been reports of intrahepatic calculi in patients with PCC in various studies. Animal studies have shown decreased bile acid synthesis and bile flow following interruption of portal flow with an increased tendency towards formation of stones.^{36,37} Pigment stones have been seen to develop in patients with ductal stenosis without stones in the gall bladder.^{22,37}

Our study had various limitations. Firstly, we did not have a large sample of patients. Calcular disease was seen in a small proportion of patients and larger volume studies are required to clearly establish the aetiopathogenesis of the disease. Secondly,

a long-term follow-up was not available. Thirdly, a comparable sized group with EHPVO and without PCC was not available so that the significance of each finding could be clearly established. The exact reason as to why a proportion of patients with EHPVO develop more severe PCC requires further studies.

CONCLUSION

In conclusion, PCC has a predilection for the extrahepatic ducts, especially the supraduodenal bile duct. Increased angulation of the CBD with scalloping is the most common finding. Angulation of the common duct predisposes to development of choledocholithiasis. Choledocholithiasis and hepatolithiasis develop owing to pathological extrahepatic ducts as sequelae of PCC.

ACKNOWLEDGMENTS

We acknowledge support provided by the Departments of Surgical and Medical Gastroenterology.

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