CASE REPORT J Pub Health Med Res 2014;2(2):67-9

Portal Biliopathy - A Rare Cause of Obstructive Jaundice

Kishan Ashok Bhagwat[1], Vikram M Patil[2], Chandramohan[3], Harman Singh Gill[2], Raghvendra Khanapur[3], Dhruva Rajgopal[3]

[1]Associate Professor, [2] Post Graduates residents, Department of Radiology,
[3]Professor, Department of Surgery
S. S. Institute of Medical Sciences and Research Centre, Davangere-577001, Karnataka, India.

(Received:12/10/2014, Revised: 24/10/2014, Accepted: 8/11/2014)

Abstract :

Portal bilipathy is rare cause of obstructive jaundice – A diagnosis of exclusion. Clinching imaging findings like portal cavernoma, paracholedhocal portal collaterals & epicholedhocal portal collaterals compressing the bile duct, contribute to diagnosis. Multiple causes of portal collaterals could be listed - idiopathic, portal vein thrombosis, portal vein fibrosis etc. No matter what the cause, it's the portal collaterals that are the clinical concern as cause of obstructive jaundice. We report a 60 year old female presenting with obstructive jaundice. Various imaging modalities like Sonography, Contrast enhanced computed tomography and Magnetic resonance imaging were performed sequentially which clinched the diagnosis of portal biliopathy, thus answering the clinical question.

Introduction : Portal biliopathy is one of the rare diseases, seldom presenting with symptoms but often asymptomatic . Portal biliopathy is one of those entities where imaging has the final say in the diagnosis and other investigations contributing precious little. Ruling out differentials like infective cholangitis, sclerosing cholangitis, biliary ascariasis and cholangiocarcinoma is a unavoidable imaging obligation . Presence of Porto-mesentric vein thrombosis, cavernous transformation of the portal vein, splenomegaly and change in caliber of CBD caused by portal collaterals are deterministic important imaging findings that favour portal biliopathy.

Case report: A 60 year old lady presented with progressive yellowish discoloration of skin and urine with prior history of fever. On initial biochemistry analysis features of obstructive jaundice was obtained. Serum bilirubin was elevated (24 g%).

Sonography of abdomen was performed which revealed intrahepatic biliary dilatation. Echogenic contents seen within dilated ducts. CBD was dilated proximally. Distal CBD was not traced due to bowel gas. Mild splenomaegaly was present. Few dilated collaterals were seen in periportal region. Main portal vein was not well visualised.

Triphasic CT scan of abdomen was performed on MDCT scanner with pressure injector showed

1. Splenomegaly with gamma gandy bodies. Leno-renal collaterals noted. (Fig. 1 & 2)
2. Cavernoma/periportal cavernous transformation. Dilated CBD seen,abruptly shouldering into normal caliber just before merging with pancreatic duct at the ampulla of vater. (Fig. 3 & 4)
3. Thickened CBD with epicholedhocal venous channels.Portal vein not visualised beyond its suprapancreatic course with suggestion of collaterals running in the paracholedochal region. Leno renal collaterals well appreciated. (Fig. 5 to 7)

MRCP was performed on 1.5 T MRI and images were reviewed on workstation. MRCP showed multiple cystic IHBR dilatations with dilated CBD tapering smoothly into normal caliber just before merging with pancreatic duct. The pancreatic duct is normal in caliber.

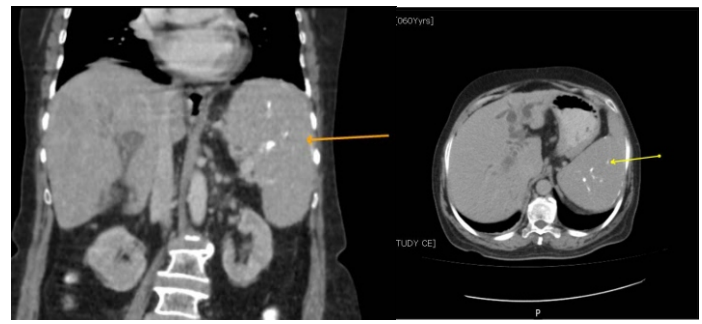

Fig 1 **Fig 2**

Fig - 1 & 2 : Left image - Coronal reformatted portal phase CT imageshowing Splenomegaly (arrow) with intraparenchymal Gamma bodies. Right image - Axial plain CT section showing gamma gandy bodies.

Address Correspondence to :
Dr. Kishan Ashok Bhagwat
Associate Professor of Radiology
SSIMS & RC, Davangere, Karnataka
Mob. : 7259805999
E-mail : bhagwatkishan@gmail.com

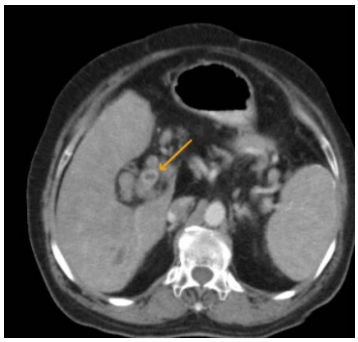

Fig. 3

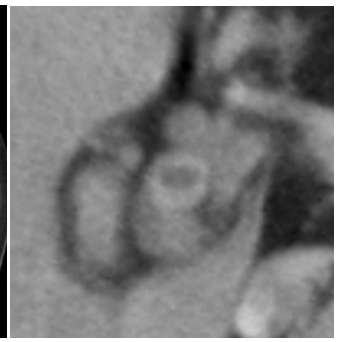

Fig. 4

Fig. 3 & 4 : Axial CT image in portal venous phase(zoomed image on the right) showing enhancing CBD wall(arrow) due to epicholedochal veins.

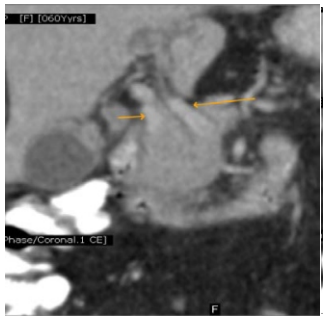

Fig. 5

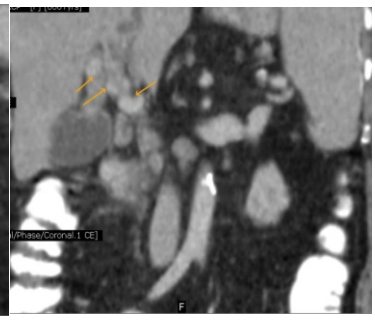

Fig. 6

Fig. 5 & 6 : Coronal reformatted images in the portal phase showing paracholedochal venous(arrows) channels causing compression of CBD giving it a varicoid appearance seen well on MRCP image below.

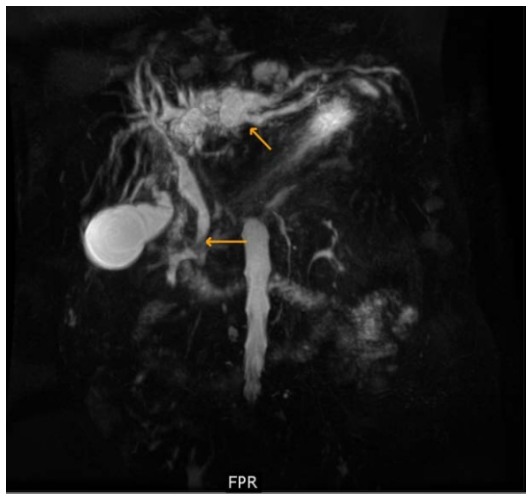

Fig. 7

Fig. 7 : MRCP showing IHBR dilatations(upper arrow) with varicoid common bile duct(loer arrow) due to paracholedochal venous compression.

Constellation of imaging features inferred as Compensated porta hypertension with portosystemic collaterals with Portal biliopathy - Fibrotic type with portal hypertension with coexisting cholangitis.

Discussion

Portal biliopathy though a rare entity , often remains undiagnosed as it remains silent in most of the cases. Patients with portal biliopathy and portal vein thrombosis presenting with symptoms account less than 50 %[1]. In the early days it was referred has pseudocholangiocarcinoma/ pseudosclerosing cholangitis but later the term portal biliopathy was coined.

The pathology though not completely understood, hypercoagulopathy leading to PVT is seen to be main causative factor for portal biliopathy rather than PVT caused by cirrhosis or other factors.[1]. Extrinsic compression of the portal vein by paracholedocal and epicholedochal collateral venous channels leads to portal biliopathy[2,3],or ischemic common bile stricture due to venous collaterals are factors to be considered.[4,5]

In the present case we see this patient presenting with obstructive jaundice clinically and bilirubin levels(24gms%) favouring the clinical diagnosis. On MRCP we could appreciate the abrupt change in distal CBD caliber due to focal stricture often seen with fibroid type of portal biliopathy. Portal bilio-pathy is classified into fibrotic, varicoid and mixed types.[1]. Though choledocholithiasis in seen in few cases with portal biliopathy[6], extensive biliary sludge wasn't mentioned in the literatures we searched. Portal vein thrombosis with cavernous transformation with superior mesenteric vein thrombosis is seen in the majority of the cases in a study conducted by Eric M. Walser , MD et al[7]. In our case though we don't see thrombus in the superior mesenteric vein, focal SMV wall calcification is seen. Splenomegaly though found in many cases as well as our case , is not the characteristic finding to add weight to the diagnosis of portal biliopathy.

Gamma gandy bodies form as a sequence of chronic portal vein hypertension and appear as high attenuating focal spots in the spleen.[8], which we could appreciate in our case and is not mentioned as a consequent finding with portal biliopathy in many literatures.

Though the presence of leno-renal collaterals and absence of GB varices is less often seen in patients with portal biliopathy, it is not rare to find these features which we see in our case.

Treatment includes portosystemic shunting or stent placement. Extrinsic compression by the collaterals has good prognosis on treatment. Ischemic stricture of the CBD is least likely to respond to stent procedure.[1]

Imaging findings of portal vein thrombosis with

paracholedochal and epocholedhocal collateral venous channels causing ischemic distal CBD stricture with gamma gandy bodies is very well appreciated in our case.

CONCLUSION:

Portal biliopathy is not a complete alien as a cause of surgical jaundice. Since it presents as a rare cause of obstructive jaundice, it is not wrong for the radiologists to be cautious and consider the clinical background before asserting the diagnosis as portal biliopathy. Not to overdiagnose, but making sure never to underdiagnose is a challenge to the radiologists in this scenario. Thus periportal collaterals irrespective of etiology, have to be dealt with caution, as these could the only cause of obstructive jaundice when other common causes are ruled out.

REFERENCES:

1. Dhiman RK, Puri P, Chawla Y, et al. Biliary changes in extrahepatic portal venous obstruction: compression by collaterals or ischemic? *Gastrointest Endosc* 1999; 50:646–52.
2. Petren T. Die extrahepatischen gallenwegsvenen and ihre: pathologischanatomische bedeutum. Verh Anat Ges 1932 ; 41 : 139 – 43 .
3. Saint JH . The epicholedochal venous plexus and its importance as a means of identifying the common duct during operations on the extrahepatic biliary tract . Br J Surg 1961 ; 48 : 489 – 98 .
4. Khuroo MS, Yattoo GN, Zargar SA, et al. Biliary abnormalities associated with extrahepatic poral vein obstruction. *Hepatology* 1993; 17:807–13.
5. Webb LJ, Sherlock S. The aetiology, presentation and natural history of extra-hepatic portal venous obstruction . Q J Med 1979 ; 48 (192): 627 – 39 .
6. Chandra R , Kapoor D, Tharakan A , Chaudhary A , Sarin SK . Portal biliopathy . J Gastroenterol Hepatol 2001 ; 16 (10): 1086 – 92 .
7. Eric M. Walser et al .Extrahepatic Portal Biliopathy: Proposed Etiology on the Basis of Anatomic and Clinical Features. Radiology 2011;258:123-34

How to Cite this article :
Kishan A B, Vikram M P, Chandramohan, H S Gill, R Khanapur, D Rajgopal Portal Biliopathy - A Rare Cause of Obstructive Jaundice. J Pub Health Med Res 2014;2(2):67-9

Funding: Declared none
Conflict of interest: Declared none