

PORTAL BILIOPATHY, A RARE COMPLICATION IN CASES OF CLD WITH PORTAL HYPERTENSION - A CT SCAN BASED STUDY FROM LIAQUAT NATIONAL HOSPITAL, KARACHI

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ABSTRACT

OBJECTIVE: The aim of this study was to record the incidence of portal biliopathy in cases of chronic liver diseases (CLD) with portal hypertension, that presented to a tertiary care hospital over a period of 2 years.

METHODS: A retrospective, cross-sectional study was performed on data collected from computerized records and CT scan images of cases of CLD. The data was collected from 30th September 2013 to 30th September 2015. The study center was the Department of Radiology, at Liaquat National Hospital, Karachi, Pakistan.

RESULTS: A total of 294 patients presented to Liaquat National Hospital, Department of Radiology with the history of chronic liver disease over a period of 2 years. The median age of presentation was 54.50 years. The sample consisted of 160 males (54.42%) and 134 females (45.57%). 264 patients were found to have portal hypertension with varices. Only 03 female patients were found to have cavernous transformation of portal vein with portal biliopathy. Portal vein thrombosis was seen in 42 patients. Regenerating nodules were seen in 36 patients and hepatomas in 98 patients. **CONCLUSION:** The incidence of portal biliopathy is rare and is very low, but not uncommon in patients of chronic liver disease with portal hypertension and cannot be disregarded. Timely diagnosis and management will be useful to lower morbidity and mortality.

Key words: Portal biliopathy, Complication, CLD, Portal hypertension, CT scan

Introduction

Portal hypertensive biliopathy (PHB) is defined as the set of anatomical and functional alterations of the intra and extra hepatic bile ducts in patients with portal hypertension due to extra hepatic portal vein obstruction (EHPVO). These changes include dilatation and stenosis of the bile ducts, common hepatic duct, gall bladder and intrahepatic ducts and they are due to extrinsic compression of these pathways by paracholecystic and paracholedochal venous plexuses that expand and compress in an attempt to decompress the venous blockage generated by the portal vein thrombosis.¹

It is much less common in cirrhotic portal hypertension,

and it is speculated that the reason is that in cirrhotic patients blocking of the portal circulation occurs at the level of hepatic sinusoid, giving origin to collateral circulation far from the vein complexes around the extra hepatic bile ducts.¹

There are two venous plexuses of bile ducts and gallbladder namely, epicholedochal venous plexus of Saint and paracholedochal veins of Petren. The epicholedochal venous plexus of Saint, which is a fine reticular web on the outer surface of the CBD and hepatic ducts, becomes dilated and causes fine irregularities in the biliary tract. The paracholedochal venous plexus of Petren runs parallel to the CBD and is connected with the gastric, pancreaticoduodenal and portal veins, and to the liver directly; its dilatation

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causes extrinsic compression over CBD. The external pressure and protrusion of these venous collaterals over a thin and pliable CBD and common hepatic duct results in changes in PHB.^{2,3,4}

Portal biliopathy is not the only cause of portal hypertension, there can be other intrahepatic and extrahepatic causes. While the extrahepatic causes include congenital portal vein obstruction, umbilical sepsis, trauma, hypercoagulation state like Budd-Chiari Syndrome and malignant occlusion, intrahepatic causes can be cirrhosis and primary biliary cirrhosis.^{5,6} In general, this disease has been reported in 70%-100% of patients with extra hepatic obstruction of portal vein.⁷

Histological and morphological studies have shown that cavernous transformation is not only dilatation of periportal collaterals but also neogenesis of vessels and formation of connective tissue occurring with increasing duration of thrombosis, which results in porous, tumor-like solid tissue that does not disappear after shunt surgery.³

Typical indications of cavernomatous degeneration of the portal vein are visible by ultrasound: a decrease in its diameter, increased echogenicity of the tissue in the hilum, associated with multiple anechoic tubular structures, corresponding to distended paracholedochal veins, and it is technically difficult to identify the common bile duct. The indentations visible by ultrasound on the common bile duct are secondary to extrinsic compression by the enlarged paracholedochal venous plexus; these are larger and are connected with the gastric vein, pancreaticoduodenal and portal vein, while the irregularities caused by dilation of pericholedochal varices may not be observed by ultrasound, since their size is less than 1 mm. Gallbladder varices, present in between 30%-55% of cases, are visualized as anechoic tubular structures of 1-5 mm in diameter in the external refractive surface of the gallbladder outside the muscular layer.⁸

Multi-detector CT (MDCT), using narrower collimation and lower acquisition time, gives high quality images for visualizing the collateral circulation, the product of portal vein obstruction. MDCT angiography techniques and post-processing, clearly demarcate the signs of cavernous transformation of portal vein, the compressive effect of collateral circulation around the bile duct, and gallbladder varices.⁹

(ERCP) features include biliary segments with narrowing of variable length and degree, indentations and irregularity of the contours of the bile duct and the presence of angles, ectasia and calculi. An absence of branching can be seen in the intrahepatic bile ducts and in some dilated ducts.³

Magnetic Resonance (MRCP) is currently the non-invasive diagnostic modality of choice, allowing an adequate characterization of the intra-and extra hepatic bile duct and with a capacity similar to ERCP for visualization of changes in the bile duct.¹⁰

Materials and Methods

A retrospective, cross-sectional study was performed on data collected from patients with a history of chronic liver disease. The study center was the Department of Radiology, at Liaquat National Hospital, Karachi. The data was collected for a period of 2 years; from 30th September 2013 to 30th September 2015. Approval of the Ethical Review Committee of the institution was obtained before the data collection. As this was a retrospective study, only computer records were accessed. No formal consent was required from the patients, nor was there any contact with them. Full confidentiality of patients was maintained by using CT numbers as the reference. Patients who were clinically and biochemically suspected of chronic liver disease that underwent CT scanning for a definitive diagnosis were included. All examinations were performed on an MDCT scanner (Toshiba Activion 16 slice CT scanner). Contrast enhanced CT scans (collimation. 4 x 2.5 mm; reconstruction section thickness, 4 mm; reconstruction intervals, 4 mm) were obtained 50 seconds after intravenous administration of 100 ml of iopamidol 370 mg I/ml (Iopamiro), injected at a rate of 2.8 ml/sec using a mechanical power injector. Opacification of the digestive tract was achieved with oral administration of 1000 ml of Urografin suspension (370 mg I/ml) administered 4 hours before the scan. CT scans were retrospectively reviewed on PACS workstations and a constellation of findings were recorded; including features and complications of chronic liver disease i.e. Altered/shrunken and irregular margins of liver

with altered left to right lobe ratio, caudate lobe enlargement, portal vein dilatation, thrombosis of portal vein and its branches, cavernous transformation of portal vein, portal biliopathy, collateral vessels (near GB fossa, porta hepatis, perisplenic, paragastric, periesophageal and peripancreatic regions), arteria- lized lesions/hepatomas, regenerating nodules, ascites and splenomegaly

Results

A total of 294 patients presented to Liaquat National Hospital, Department of Radiology with the history of chronic liver disease over a period of 2 years. The median age of presentation was 54.50 years. The sample consisted of 160 males (54.42%) and 134 females (45.57%). 264 patients were found to have portal hypertension with or without associated varices. Only 03 female patients were found to have cavernous transformation of portal vein with associated portal biliopathy. Portal vein thrombosis was seen in 42 patients. Regenerating nodules were seen in 36 patients and hepatomas in 98 patients. (Table1).

CLD with Portal Hypertension	264 Patients	89.79 %
Cavernous transformation of PV	03 Patients	1.02 %
Portal Biliopathy	03 Patients	1.02 %
Portal vein thrombosis	42 Patients	14.28 %
Regenerating Nodules in liver	36 Patients	12.24 %
Hepatomas in liver	98 Patients	33.33 %

Table 1: Features of chronic liver disease in our study of 294 patients.

Discussion

Portal biliopathy (PB) is the term used to define the biliary obstruction and morphologic changes occurring in the biliary tree and gallbladder in patients with portal hypertension. Extrahepatic obstruction of the portal vein (EHOPV) accounts for approximately 40% of portal hypertension cases worldwide, but it is almost always the culprit when PB is present.^{11,12,13} Chronic extrahepatic PV obstruction is common in the developing world and rare in Western nations.⁷ Symptomatic PB is rare, with estimates ranging between

5% and 38% of patients with PVT.¹⁰ Chandra et al have proposed a morphological classification based on topography of the cholangiography findings as shown in (Tab. 2).¹¹ The most common etiologies of

Type	Findings
I	Involvement of extrahepatic bile duct
II	Involvement of intrahepatic bile ducts only
III a	Involvement of extrahepatic bile duct and unilateral intrahepatic bile duct
III b	Involvement of extrahepatic bile duct and bilateral intrahepatic ducts

Table 2: Chandra et al have proposed a morphological classification based on the topography of the cholangiography findings

extrahepatic portal vein obstruction are shown in (Tab. 3).⁷ Development of collaterals is common and often involves the epicholedochal and pericholedochal venous plexuses.¹⁰

Children (N=275) ^a	Adults (N=356) ^b
1. Idiopathic (55%)	1. Idiopathic (45%)
2. Abdominal infections (36%)	2. Abdominal infections (20%)
3. Umbilical catheterization (6%)	3. Trauma/other (15%)
4. Trauma/other (3%)	4. Prothrombotic disorders (9%)
5. Pancreatitis (1%)	5. Pancreatitis (1%)

^aPooled results from 7 studies published from 1962 to 1994.

^bPooled results from 6 studies published from 1979 to 1997.

Table 3: Most Common Etiologies of Extrahepatic Portal Vein Obstruction

These are unusual presentations for cases of portal hypertension. Only few cases reported earlier had such grave presentation. For instance, Sumanthi et al presented a case of portal hypertension with obstructive jaundice secondary to portal biliopathy.⁵ Similarly, Chawla et al reported a case of portal biliopathy with obstructive jaundice and hematemesis.⁶ Choudhuri et al published the first case report of relief of CBD obstruction secondary to portal cavernoma after a proximal lienorenal shunt, in a patient with symptomatic portal biliopathy with extrahepatic portal venous obstruction (EHPVO). Other authors have published similar results.¹⁴

Our study was aimed towards the incidence of portal biliopathy and the presence of cavernous transformation of portal vein in CLD patients suffering from

EHPVO because long standing obstruction of the portal vein leads to replacement of the portal vein by large collaterals along the CBD - the so-called cavernomatous transformation of the portal vein. These large collaterals compress the pliable CBD, leading to the changes seen on CT and ERCP. Also, with increased duration of portal vein thrombosis, there is vascular neogenesis and formation of tumor-like connective tissue, which can encase the CBD or cause angulation of the bile ducts leading to portal biliopathy.³

Approximately one quarter of patients with chronic liver disease develop complications ranging from: portal hypertension, caudate lobe enlargement, portal vein dilatation, thrombosis of portal vein and its branches, cavernous transformation of portal vein, portal biliopathy, collateral vessels (near GB fossa, porta hepatis, perisplenic, paragastric, periesophageal and peripancreatic regions), arterialized lesions/hepatomas, regenerating nodules, ascites and splenomegaly. However in our study, 42 out of a sample of 294 patients were found to have thrombosis of portal vein related to CLD resulting in cavernomatous transformation of portal vein in 03 patients with subsequent development of portal biliopathy (Tab. 1). This statistic of a striking 14.28% cannot be ignored because it supports the compression theory in EHPVO.³

All these three patients were known case of chronic liver disease with portal hypertension and portal vein thrombosis. CT scan was done in these patients which not only showed hepatosplenomegaly, portal vein thrombosis with its cavernomatous transformation, multiple collateral vessels along superior mesenteric and portal veins, perisplenic, paragastric, peripancreatic and along anterior abdominal wall, but also revealed an ill-defined soft tissue thickening and enhancing mass measuring 2.7 x 2.7 cm in the region of mid CBD extending from porta hepatis upto pancreatic head encasing the CBD and right and left hepatic ducts resulting in minimal dilatation of IHDs. It was seen abutting the head of pancreas and duodenum with loss of fat planes. Therefore, on the basis of imaging, diagnosis of portal biliopathy was made (Fig. 1, 2 and 3).

Portal biliopathy is a rare but serious complication of chronic liver disease which if undetected, results in high mortality. Management of portal biliopathy depends upon clinical manifestations. Asymptomatic

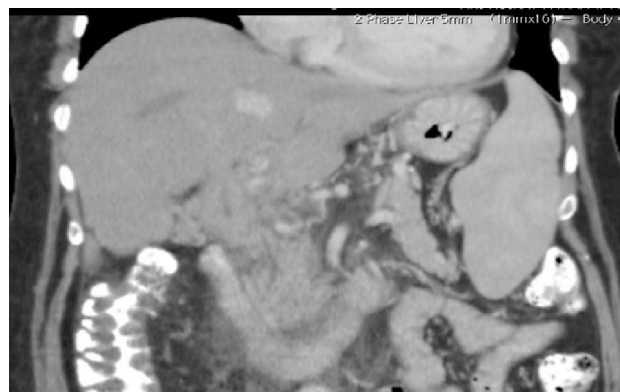


Figure 1: CT scan Coronal Image: An ill-defined soft tissue thickening and enhancing mass measuring 2.7 x 2.7 cm in the region of mid CBD extending from porta hepatis upto pancreatic head encasing the CBD and right and left hepatic ducts. There is minimal dilatation of IHDs. Findings most likely represent secondary inflammation of walls of CBD due to Portal Biliopathy.

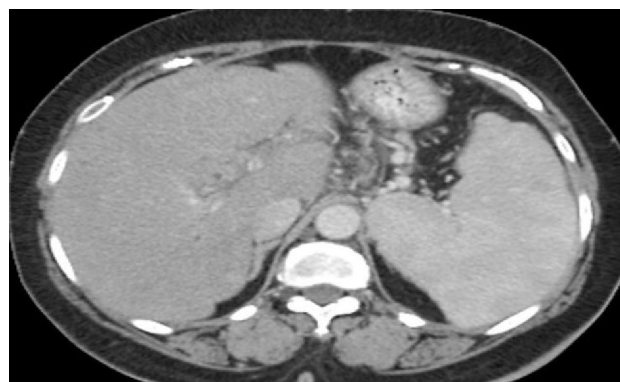


Figure 2: CT scan Axial Image: Hepatosplenomegaly and Cavernous transformation of portal vein with subsequent collaterals are also noted.



Figure 3: CT scan Axial Image: The mass is also seen abutting the head of pancreas and duodenum with loss of fat planes.

patients generally do not require treatment. CBD calculi can be removed by ERCP and if there is stricture or narrowing in the duct, stent can be placed. Stones causing obstruction can be removed by endos-

copric sphincterotomy. Care should be taken, as there are large venous collaterals around ampullary region, which can bleed profusely. In patients with symptomatic biliary obstruction not amenable to endoscopic therapy, a Porto systemic shunt is indicated to decompress the portal system. If changes of portal biliopathy do not regress even then, hepaticojejunostomy can be performed to alleviate biliary obstruction.^{15,16}

Although our study leads to significant findings, it was not aimed towards determining the best intervention and management of the condition; nor was there any follow up with the patients to ascertain the sequelae of this affliction. Also, being a single institution experience, it is somewhat confined to a less than diverse group of patients. A multi-center study would be even more thorough in determining the prevalence of portal biliopathy in patients suffering from chronic liver disease with portal hypertension.

Conclusion

The incidence of portal biliopathy is low, but not uncommon in patients of chronic liver disease with portal hypertension and cannot be disregarded. Timely diagnosis and management will be useful to lower morbidity and mortality. Cavernous transformation of the portal vein due to extrahepatic portal vein obstruction is not infrequent but biliary obstruction in association with this disorder is distinctly uncommon. Proper case management is very important as prolonged biliary duct obstruction can lead to the development of ascending cholangitis or later on secondary biliary cirrhosis. Only 03 cases of portal biliopathy were detected over a period of 02 years in our study representing its relative rarity, however a larger multicenter study would be more representative.

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