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Case Report

Portal Biliopathy. Experience in Two Reference Institutions

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1. Abstract

1.1. Background: Portal cavernoma cholangiopathy is defined as cholangiographic anomalies secondary to collateral venous circulation and portal cavernous transformation. Most of the time it is asymptomatic, hence the greater difficulty in achieving prompt diagnosis and treatment.

1.2. Methods: A descriptive study was conducted on patients with portal cavernous cholangiopathy seen at a two referral centers within the time frame of January 2006 to December 2018. Clinical manifestations, cholangiographic alteration pattern, thrombosis extension, treatment, and mortality were analyzed.

1.3. Results: A total of 23 patients with cavernoma cholangiopathy were reported and their median age was 42.5 years. Eight cases (34.8%) presented with cirrhosis of the liver and fifteen cases (65.2%) did not. The most frequent symptoms were abdominal pain and jaundice. CT angiography was the study of choice for characterizing the portal anomalies. ERCP and magnetic resonance cholangiography were used in the evaluation of biliary tract anomalies.

Thrombosis extension showed a preference for the extrahepatic portal vein in the cirrhotic patients. Biliary stricture at the extrahepatic site was predominant in both groups. Six patients (26%) did not require treatment, 3 (13%) required medical treatment, and the rest received endoscopic therapy. Ten patients (21.7%) needed surgical treatment. Nine deaths were reported, and severe acute cholangitis was the main cause. One-year and three-year survival rates were 70.8% and 58.3%, respectively.

1.4. Conclusions: The real frequency of portal cavernoma cholangiopathy is difficult to determine because it is usually asymptomatic. Adequate diagnosis through imaging techniques is decisive for determining the treatment and prognosis of those patients.

2. Keywords: Portal cavernoma cholangiopathy; Portal vein thrombosis; Portal hypertension; Biliary obstruction; Porto-systemic shunt; Endoscopic retrograde cholangiopancreatography

3. Introduction

Portal Cavernoma Cholangiopathy (PCC) has been described by various authors. It was initially known as portal biliopathy and defined as the presence of cholangiographic anomalies secondary to the formation of collateral venous circulation, in the image of a cavernoma. Said anomalies are a consequence of chronic thrombosis of the extrahepatic portal vein associated with portal hypertension. It is observed in 81% to 100% of patients with chronic extrahepatic portal thrombosis, the majority of who do not present with cirrhosis (9%-40%). Thrombosis of the extrahepatic portion of the portal vein is a vascular disorder due to the obstruction of flow at the portal, splenic, or superior mesenteric veins. Cavernous formation surrounding the portal vein is the result of the presence of collateral veins, which are dilated to compensate and direct the blood flow towards the hepatic circulation. PCC develops when those venous plexuses are dilated, thus causing extrinsic compression of the common hepatic duct and gallbladder [1-3].

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Most cases are asymptomatic. Obstructive jaundice is occasionally seen and contributes to the development of choledocholithiasis. Symptomatic disease is reported in 5 to 50% of patients and the most frequent symptoms are: abdominal pain, jaundice, and acute cholangitis. Obstruction of the extrahepatic portal vein is frequent, but the association with obstruction of the biliary tree is not. Patients are usually asymptomatic. It is of vital importance to promptly identify and treat PCC because chronic obstruction can lead to cholangitis or secondary biliary cirrhosis [4-6].

We present herein a retrospective analysis of patients at referral center in Mexico City with clinical and imaging signs of PCC and describe the treatment results. Additionally, a systematic review of literature about case series and management was carried out.

4. Materials and Methods

A retrospective recollection of data was carried out on patients diagnosed with portal cavernoma cholangiopathy that were seen at the *Instituto Nacional de Ciencias Medicas y Nutricion "Salvador Zubiran"* and *Hospital Juarez de Mexico* within the time frame of January 2006 to December 2018.

4.1. Subjects

Patients who had an established diagnosis of cavernous transformation of the portal vein based on clinical, laboratory and imaging findings by ultrasound Doppler, computed tomography splenoportovenography (CTSPV) or Magnetic resonance cholangiopancreatography (MRCP) were retrospectively enrolled for the study. Patients with incomplete clinical record were excluded from the study.

4.2. Clinical Information

Clinical records were reviewed for the presence and frequency of symptoms pertaining to cholangiopathy, which include abdominal pain, jaundice, Cholangitis episodes, variceal bleeding, hemobilia episodes, ascites. Endoscopic or surgical procedures and relevant laboratory data were collected from the patient's clinical charts.

The biliary and portal anomalies were evaluated through imaging studies. Diagnosis of PCC was based on the findings of portal thrombosis, with or without cavernous transformation, and the formation of collateral venous circulation identified either by CTSPV, MRCP or Doppler ultrasound of the liver. In the some cases, the abnormalities of the bile duct were show and described for endoscopic retrograde cholangiopancreatography (ERCP). The images was examined for searching thrombosis extension portal vein and portal cavernoma and any abnormalities of the biliary tree described as 1) pattern of ductal involvement: extra hepatic: the changes and dilatation were confined to the extrahepatic biliary tree beyond the hepatic hilum; intra hepatic: the changes were confined to the intra hepatic ducts; and both extrahepatic and intra hepatic: where changes were seen in the extrahepatic biliary tree with dilatation and abnormalities of the intra hepatic ducts as well. 2) Presence of dilatation. 3) Changes in the intra hepatic and extrahepatic ducts: dilatation, extrinsic impressions/indentations and stricture.

Esophageal or gastric varices were detected at upper endoscopy using the Baveno classification, in which small varices are < 5 mm and large ones are > 5 mm. The Sarin classification was used for gastric varices and describes GOV 1 as the continuation of esophageal varices up to 5 cm beyond the gastro esophageal junction. GOV 2 is varices extending below the esophago gastric junction to the gastric fund us; Isolated Gastric Varices (IGV) are divided into type 1, located in the fund us, and type 2 at any other gastric site [7-8]. The data on biochemical findings at the time of diagnosis, treatment required, and mortality rates were registered.

For the information search, a systematic exploration on MEDLINE, PUBMED and EBSCO was done. The studies were identified using the following terms: "Hypertension portal" OR "portal cavernoma cholangiopathy" OR "portal cavernoma" OR "cholangiopathy" OR "portal biliopathy" OR "extrahepatic portal venous obstruction" AND "treatment". Only studies on humans were considered, and papers written in English were used for the analysis. We included manuscripts reporting endoscopic and surgical treatment of PCC, including case series and case reports, review articles, guidelines or comments to other papers, and reports about PCC treatment or therapy complications.

4.3. Statistical Analysis

Statistical analysis was carried out using the IBM statistical package SPSS® v.22.0 (IBM Chicago, IL). The categorical variables were summarized in terms of frequencies and percentages. Quantitative variables were assessed in terms of mean, median, mode and standard deviation. 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.

5. Results

5.1. Patients

Twenty-three patients were diagnosed with PCC. Age at the time of diagnosis was 45.4±15.5 years. Fourteen patients (60.9%) were men and 9 (39.1%) were women. There were no data of chronic liver disease in 15 cases (65.2%) and 8 (34.8%) patients were diagnosed with cirrhosis of the liver through imaging studies and liver function test alterations. The different etiologies reported were: 12 (52.2%) cases of idiopathic extrahepatic portal vein thrombosis, 3 (13%) cases of viral hepatitis with portal hypertension, 2 (8.7%) cases of essential thrombocytosis, 2 (8.7%) cases of biliary pancreatitis, 2 (8.7%) cases of pancreatic cancer, 1 (4.3%) case of AIH/PBC overlap syndrome, and 1 (4.3%) case of secondary biliary cirrhosis.

5.2. Diagnostic Evaluation and Biochemistry

A CT splenoportovenography was the study of choice for charac-

terizing the portal anomalies and it was performed on 22 patients (95.7%). Cholangiographic anomalies were analyzed with magnetic resonance cholangiography in 16 patients (69.6%) and with ERCP in 15 patients (65.2%). Doppler ultrasound of the liver was utilized as part of the evaluation of portal thrombosis extension in 20 patients (87%) (Figure 1).

Upper endoscopy was performed on every patient and large esophageal varices were identified in 13 patients (56.5%), severe portal hypertensive gastropathy in 10 (43.5%) cases, mild portal hypertensive gastropathy in 4 (17.4%) patients, small esophageal varices in 4 (17.4%) cases, and a GOV 1 varix in 1 patient (4.3%).

Laboratory test results were evaluated at the time of diagnosis showing normal CBC parameters. Jaundice was present in 8 cases (34.7%) and the rest of the liver function tests showed a cholestatic pattern with mild transaminase elevation (Table 1).

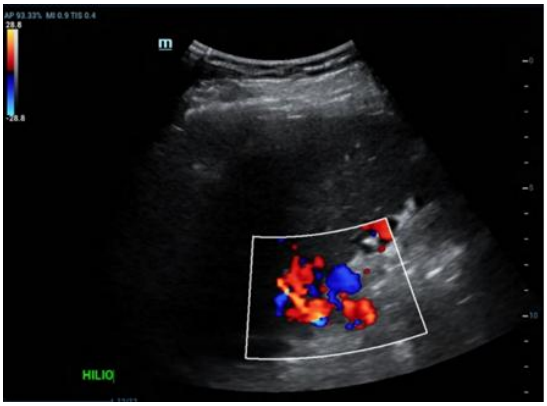

Figure 1: Doppler ultrasound showing multiple periportal veins at the hepatic hilum corresponding with a portal cavernoma.

Table 1: Clinical and biochemical at diagnosis in patients with PCC with and without cirrhosis.

Parameter	Patients with cirrhosis	Patients without cirrhosis	p
	n = 8 /n (%)	n = 15 /n (%)	
Clinical signs			
Jaundice	7 (87.5)	5 (33.3)	0.012*
Cholangitis	3 (37.5)	4 (26.7)	0.61
Cholangitis episodes (>3)	3 (37.5)	4 (26.7)	0.88
Variceal bleeding	7 (87.5)	7 (46.6)	0.018
Hemobilia	3 (37.5)	2 (13.3)	0.182
Abdominal pain	5 (62.5)	11 (73.3)	0.61
Ascitis	7 (87.5)	5 (33.3)	0.012
Hemoglobine (mg/dl)	11.1 + 1.7	13.1+ 3.5	0.159
WBC (mm^3)	5.3+ 1.3	6.1+ 2.4	0.543
Platelets	148+ 105	246+ 180	0.178
Total billirubin (mg/dl)	10.1+ 10.9	7.48+ 13.9	0.664
AST (UI/l)	57.5+ 24.3	44.5+ 51.1	0.42
ALT (UI/l)	35.6+ 8.0	35.9+ 33.9	0.984
AP (UI/l)	125.3+ 111.9	186.8+ 213.1	0.53
GGT (UI/l)	213+ 152	330+ 310	0.331
INR	1.27+ 0.12	1.51+ 0.21	0.354

Data are expressed as means ± standard deviation /frequencies (percentages),* p <0.05.

5.3. Radiological Findings

(Table 2) shows the radiological findings in patients with PCC, with and without cirrhosis. Each patient was documented with cavernous transformation at the portal vein (Figure 2). Thrombosis extension showed an exclusive preference for the extrahepatic portal vein in patients with cirrhosis, with a statistically significant difference in relation to patients with no cirrhosis. Stricture was only present in the extrahepatic biliary tree in both groups (Figure 3). As for collateral circulation formation, it was predominant in the group of patients with no cirrhosis.

Table 2: Radiological characteristics in patients with PCC with and without cirrhosis.

Characteristics	Patients with cirrhosis	Patients without cirrhosis	p
	n = 8 /n (%)	n = 15 /n (%)	
Ultrasound			
Cholelithiasis	2 (25)	5 (33.3)	0.661
Choledocolithiasis	2 (25)	3 (20)	0.673
Thrombosis extension			
PV	6 (75)	8 (53.5)	0.000*
PV / SV	-	1 (6.7)	-
PV / SMV	2 (25)	1 (6.7)	0.000*
PV / SV / SMV	-	1 (6.7)	-
Biliary stenosis			
Extrahepatic	4 (50)	7 (46.7)	0.004*
Intrahepatic	1 (12.5)	1 (6.7)	0.000*
EHBT plus IHBT right	-	1 (6.7)	-
EHBT plus IHBT left	1 (12.5)	-	-
EHBT plus IHBT bilateral	-	5 (33.3)	-
Collateral circulation			
Peri-pancreatic	1 (12.5)	3 (20)	0.000*
Peri-main biliary duct	4 (40)	8 (53.3)	0.000*
Hepatic hilum	2 (25)	5 (33.3)	0.000*
Splenic hilum	2 (25)	5 (33.3)	0.000*
Peri-gástric	3 (37.5)	6 (40)	0.000*
Gastric fundus	-	2 (13.3)	-
Peri-splenic	-	4 (26.7)	-
Lower esophagus	2 (25)	6 (40)	0.000*
Gallbladder	-	1 (6.7)	-

* p <0.05.

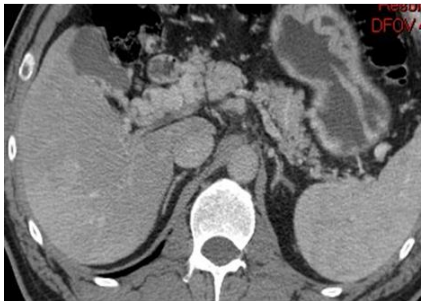

Figure 2: Cavernous degeneration of the porta. A mild dilation at the intrahepatic biliary tract is observed. Extensive perigastric and peripancreatic collateral porto-sistemic circulation. Splenomegaly.

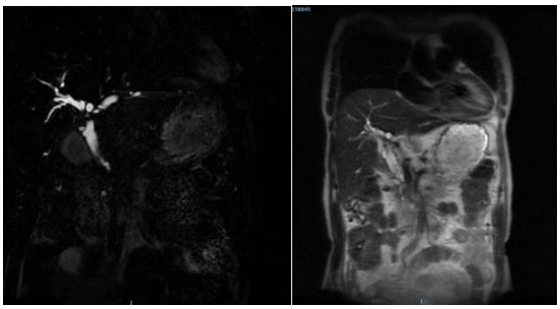

Figure 3: Magnetic cholagioresonance showing intra and extrahepatic biliary dilation with a signal absence at the distal common biliary duct.

5.4. Therapeutic Strategies

Treatment was focused on portal hypertension control and the management of biliary tract obstruction. Seven patients (29.2%) required only medical treatment with non-selective beta blockers. In pro-thrombotic pathologies, oral anticoagulation was used. Most of the complications caused by biliary tract obstruction were treated endoscopically, with biliary stent placement in 10 patients (41.7%) and sphincterotomy in one patient (4.2%) (Figure 4). Surgical treatment was required in 10 patients (41.7%). It was combined with medical treatment in 3 cases (12.5%) and with endoscopic treatment in 5 patients (20.8%). (Table 3) describes the treatments employed.

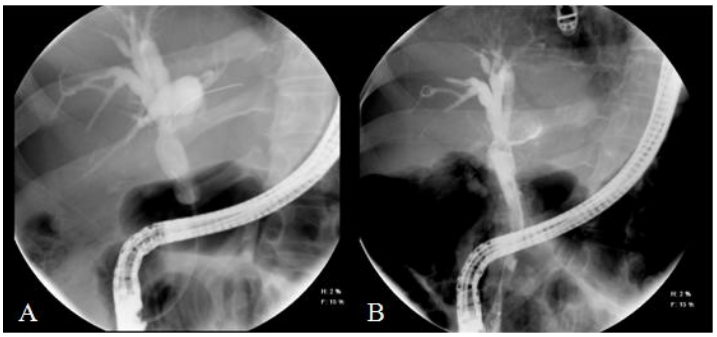

Figure 4: Endoscopic cholangiography of a patient with portal cavernous cholangiopathy. Dilation of the intra and extrahepatic biliary tract with distal main biliary duct stenos is observed (A). Biliary endoprotesis insertion (B).

Table 3: Treatment of patients with portal cavernous cholangiopathy with and without cirrhosis

Treatment	Patients with cirrhosis	Patients without cirrhosis
	n = 8 /n (%)	n = 15 /n (%)
Medical treatment		
Anticoagulation	-	5 (31.3)
NSBB	2 (25)	1 (6.3)
Anticoagulation/NSBB	-	2 (12.5)
Surgical treatment		
Splenectomy	4 (50)	1 (6.3)
Biliodigestive derivation	-	1 (6.3)
Warren´s Surgery	-	1 (6.3)
Mesocavalderivation	-	1 (6.3)
Sugiura	-	2 (12.5)
Endoscopic treatment		
Biliary endoprotesis	6 (75)	4 (25)
Sphincterotomy	1 (12.5)	-

In the two groups, complications were a consequence of portal hypertension, and more frequently so, in patients without cirrhosis (p 0.001). The main documented complication was variceal bleeding, in 10 patients (41.7%).

Nine deaths were reported (37.5%) and 3 patients (12.5%) were lost to medical follow-up. Severe cholangitis was the main cause of death, presenting in 3 cases (37.5%), followed by gastrointestinal bleeding in 2 cases (25%): The other 4 patients died from complications resulting from their primary diseases. The one-year survival rate was 70.8% and the 3-year survival rate was 58.3%.

The treatment of symptomatic PCC should be determined on a case by case basis. The main objective of treatment should be focused on the management of portal hypertension and relief of biliary obstruction. The various series cases published in current literature show that there is no consensus regarding optimal treatment and that this individualize for case.

6. Discussion

Abnormalities of the biliary tract in patients with portal vein thrombosis and portal hypertension were described in three articles published in the 1990s, establishing the term "portal biliopathy" suggested by Sarin et al. in 1992. Due to the different nomenclature described by various authors and the different criteria used to define the biliary alterations, as well as the deficient standardization of the clinical importance, natural history, and prognosis of the disease, the Indian National Association for Study of the Liver (INASL) published the first Portal Cavernoma Cholangiopathy Consensus Statement in 2012. The consensus defines the anomalies in the biliary tract, including the cystic duct and gallbladder, based on the following criteria: 1) the presence of a portal cavernoma; 2) cholangiographic changes in ERCP or magnetic resonance cholangiography, and 3) the absence of other causes, such as bile duct lesion, primary sclerosing cholangitis, or cholangiocarcinoma [9, 10].

The etiology of anomalies in the extrahepatic portion of the portal vein varies according to age group and geographic area. In developing countries, intra-abdominal infections represent most of the cases, particularly in children. In the United States and Western Europe, PCC is more frequent in adults, considering the cirrhotic and non-cirrhotic etiologies, with the pro thrombotic states being the most frequent in the non-cirrhotic group of patients. Some cases are considered idiopathic. The median age in our patients was 42 years, which we believe is the reason why cirrhosis of the liver was the most frequent etiology. In patients with no cirrhosis, the main cause was prothrombotic disease. A total of 8.7% of the cases were considered idiopathic, data that correspond to the results of different published series [11-13].

The principal pathogenic theory is that of obstruction of the portal vein, with later collateral vein formation, varices, and cavernoma. The venous flow is diverted through the periductal system by the Saint's plexus, causing mild irregularities in the biliary tract and the Petren's plexus, consequently producing extrinsic compression at the bile duct. An alteration at the left hepatic duct is observed more often, which could be due to the prominence of the collateral veins at the union of the umbilical vein and the left branch of the portal vein. No predominance of a unilateral alteration was apparent in our patients. To the contrary, most of the cases presented with bilateral involvement. A second theory is that of ischemic origin due to prolonged compression caused by the presence of collateral veins and diminished portal circulation flow [14-18].

Portal vein thrombosis is not infrequent, but few cases present with clinical manifestations. Different case series have described portal

vein thrombosis, reporting cholangiographic changes in 78 to 100% of the cases. Not many series have been published, and so real frequency is unknown, making our study one of special interest because it describes the clinical presentation at the time of diagnosis and the abnormalities of the biliary tree. The clinical course is usually asymptomatic and clinical manifestations are present in 5 to 50% of patients. Llop E et al [19]. reported a case series that included 67 patients. They found no significant differences in the clinical presentation in relation to the different degrees of cholangiographic patterns and the extension of venous thrombosis in patients with and without cirrhosis. A predominance of thrombosis at the extrahepatic portion of the portal vein was observed, and interestingly, there was greater thrombosis extension and collateral circulation in the patients without cirrhosis [20, 21].

Diagnostic criteria are established by ERCP or magnetic resonance cholangiography. ERCP is a therapeutic, as well as a risky, procedure. In our patients, diagnosis was made mainly through ERCP or magnetic resonance cholangiography. CT angiography and Doppler ultrasound of the liver were complementary studies utilized for better characterization of the venous collateral circulation, anatomic stricture correlation, bile duct dilation, and liver function status, as well as for the purpose of conducting a therapeutic approach [22-25].

The treatment of symptomatic patients is focused on the management of complications resulting from portal hypertension or cholangitis due to biliary obstruction. Endoscopic drainage should be considered a first therapeutic option in patients with cholangitis secondary to stricture or choledocholithiasis. Biliary stone extraction and biliary stent placement have been shown to be safe in patients with PCC. In the present case series, 70% of patients that required endoscopic treatment had previously undergone surgery to reduce portal hypertension. Even so, anatomic biliary tree alterations persisted, with torpid progression and the development of infections due to the prolonged obstruction at the biliary tree. Endoscopic sphincterotomy is currently the first choice in symptomatic cases. It has not been associated with higher bleeding rates and the use of the Dormia basket and extraction balloon is safe in most cases. We also documented a case of hemorrhage after endoscopic treatment, with fatal results. Therefore, biliary stent placement is recommended only in patients with cholangitis or obstructive jaundice. Stricture has been resolved in some cases through repetitive stent replacement in 3 to 5-year periods. However, it was not resolved in three of our patients that underwent multiple stent replacements [26-30].

Vilbert et al. published a case series with 64 patients that analyzed the therapeutic strategies in patients with PCC, emphasizing the importance of identifying bile duct obstruction to prevent complications. Surgical treatment was evaluated, and retroperitoneal splenorenal anastomosis was recognized as initial treatment. In cases of symptom persistence, biliodigestive diversion was recommended over repetitive endoscopic procedures. In our series, surgical treatment was initially performed, with a later need for biliary endoprosthesis placement. None of the patients had a second surgical intervention. Endoscopic treatment was not required in three of our patients. They only needed medical treatment for complications due to portal hypertension and oral anticoagulation to treat a pro-thrombotic pathology [31, 32].

Exclusive medical treatment was considered in 30% of our patients because they had prothrombotic pathologies. Beta-blockers were administered for portal hypertension control. Two cases presented with variceal bleeding that required endoscopic management.

Patients with symptomatic portal cavernoma cholangiopathy are usually diagnosed in the fifth decade of life, due to late clinical manifestations, which was similarly observed in our study. It is of vital importance to assess patients with portal vein thrombosis and make an early diagnosis of any anatomic alterations of the biliary tree to prevent liver function deterioration.

Is important to point out that there are limitations to our study. First, because this study was a retrospective review, selection bias was unavoidable. Moreover, the small number of patients limits the applicability of the treatments we

propose. Finally, the histopathological evidence to corroborate the diagnosis of cirrhosis was not available for all the patients we included.

7. Conclusion

In conclusion the portal cavernoma cholangiopathy is considered an infrequent complication and it is the most severe manifestation of portal vein thrombosis. It is hard to determine its real frequency due to the large number of patients that are asymptomatic. The most frequent symptoms are secondary to complications derived from portal hypertension and episodes of cholangitis. Most patients need numerous treatments to reduce portal hypertension and biliary tree anomalies and the rate of complete resolution is low. Early identification and diagnosis through imaging techniques are decisive for optimum treatment and outcome in patients with PCC.

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