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From Neonatal Portal Vein Thrombosis to Complex Portal Cavernoma Cholangiopathy: A Multidisciplinary Therapeutic Challenge

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Abstract **Case Report**

Portal cavernoma cholangiopathy represents a rare biliary complication of chronic extrahepatic portal vein obstruction. Although radiological biliary abnormalities are frequently detected in patients with portal cavernoma, symptomatic disease remains uncommon and usually occurs after years of evolution. We report the case of a 19-year-old female with chronic portal cavernoma secondary to neonatal portal vein thrombosis related to umbilical catheterization, complicated by portal hypertension and recurrent upper gastrointestinal bleeding. She presented with progressive cholestatic jaundice without abdominal pain or infectious syndrome. Imaging demonstrated severe biliary dilatation associated with choledocholithiasis, intrahepatic lithiasis, peri-cholédocien venous collaterals, and massive splenomegaly in the setting of chronic portal cavernoma. Endoscopic retrograde cholangiopancreatography (ERCP) allowed temporary biliary drainage; however, recurrent cholangitis occurred despite stenting, requiring repeat ERCP. Due to persistent biliary compression and advanced portal hypertension, the patient ultimately underwent staged surgical management with cholecystectomy, roux-en-y hepaticojejunostomy, and mesenterico-caval shunt. This case highlights the progressive nature of portal cavernoma cholangiopathy, the role of multimodal imaging in diagnosis, and the importance of multidisciplinary management in advanced symptomatic forms.

Keywords: Portal cavernoma cholangiopathy; Portal vein thrombosis; Biliary obstruction; Portal hypertension.

INTRODUCTION

Chronic portal vein thrombosis with cavernomatous transformation of the portal vein is a major cause of non-cirrhotic extrahepatic portal vein obstruction and may lead to long-term complications related to portal hypertension and hemodynamic alterations within the hepatic hilum. Among these complications, portal cavernoma cholangiopathy refers to the spectrum of morphological and functional biliary abnormalities observed in patients with portal cavernoma [1].

The pathophysiology of this condition is multifactorial. It mainly involves extrinsic compression of the bile ducts by pericholedochal venous collaterals, combined with ischemic injury secondary to impairment of the peribiliary venous plexus during chronic portal vein thrombosis [2]. These mechanisms may result in ductal irregularities, segmental strictures, biliary dilatation, and, in advanced stages, bile stasis promoting secondary stone formation and recurrent cholangitis.

Although biliary abnormalities are detected in 77% to 100% of patients with chronic extrahepatic portal vein obstruction, only 5% to 38% become symptomatic [1,2]. Clinical manifestations generally occur after several years of disease progression and may include obstructive jaundice, chronic cholestasis, biliary pain, or recurrent biliary infections [3].

Diagnosis currently relies on a multimodal imaging approach integrating Doppler ultrasonography, computed tomography angiography, magnetic resonance cholangiography, and endoscopic techniques. Magnetic resonance cholangiopancreatography is considered the reference non-invasive imaging modality, allowing simultaneous assessment of biliary and vascular abnormalities. Therapeutic management remains challenging and poorly standardized, requiring an individualized strategy combining endoscopic drainage, treatment of biliary lithiasis, and, in advanced cases, biliary surgery or portosystemic shunting procedures [4].

We reported the case of a young woman presenting with symptomatic portal cavernoma

cholangiopathy complicating chronic portal vein thrombosis, probably related to neonatal umbilical catheterization, highlighting the progressive nature of this rare condition and the diagnostic and therapeutic challenges it poses.

Case Presentation

A 19-year-old female with a history of chronic portal hypertension secondary to portal cavernoma, diagnosed in 2014, was admitted to our department for progressive cholestatic jaundice of one-month duration. The jaundice was associated with choluria and generalized pruritus, without abdominal pain, fever, vomiting, gastrointestinal bleeding, or bowel habit disturbances. Symptoms evolved in a context of preserved general condition and absence of infectious syndrome.

Her past medical history included hospitalization on the second day of life for undocumented upper gastrointestinal bleeding requiring neonatal intensive care, during which umbilical vein catheterization had reportedly been performed. No history of neonatal sepsis, omphalitis, abdominal trauma, or abdominal surgery was noted. Since childhood, the patient had been followed for portal hypertension related to chronic portal vein thrombosis with cavernomatous transformation. She had experienced two previous episodes of upper gastrointestinal bleeding secondary to esophageal varices, managed endoscopically with variceal ligationand non-selective beta-blocker therapy. However, beta-blockers had been discontinued two years prior to admission.

On admission, the patient was hemodynamically stable with normal respiratory function, afebrile, and in preserved general condition (WHO performance status 0). Physical examination revealed marked cutaneomucosal jaundice associated with diffuse scratching lesions. Signs of portal hypertension were present, including collateral venous circulation over the abdominal flanks and massive splenomegaly extending beyond the umbilicus. No ascites, abdominal tenderness, palpable abdominal mass, peripheral edema, or signs of chronic liver failure were identified.

Laboratory investigations demonstrated a predominantly cholestatic liver profile with elevated alkaline phosphatase (301 IU/L, approximately 2× upper limit of normal) and gamma-glutamyl transferase (72 IU/L, 2.1× upper limit of normal), associated with moderate hepatocellular injury (AST 83 IU/L and ALT 57 IU/L). Severe hyperbilirubinemia was noted, with total bilirubin reaching 32.4 mg/dL predominantly conjugated, consistent with significant obstructive cholestasis (Table 1).

Liver synthetic function remained relatively preserved, with a prothrombin time of 74% and Factor V activity of 68%, without evidence of hepatic failure. Complete blood count revealed pancytopenia, including anemia (hemoglobin 8.9 g/dL), leukopenia (1,900/mm³), neutropenia (1,200/mm³), and thrombocytopenia (50,000/mm³), suggestive of hypersplenism related to severe portal hypertension. Inflammatory markers were not significantly elevated (Table 1).

Table 1: Baseline laboratory investigations and reference ranges

Parameter	Result	Normal values
Total bilirubin	32.4 mg/dL	0.2–1.2 mg/dL
AST (Aspartate aminotransferase)	83 IU/L	10–40 IU/L
ALT (Alanine aminotransferase)	57 IU/L	7–45 IU/L
Alkaline phosphatase (ALP)	301 IU/L	40–130 IU/L
Gamma-glutamyl transferase (GGT)	72 IU/L	8–35 IU/L
Prothrombin time	74%	70–100%
Factor V activity	68%	70–120%
Hemoglobin	8.9 g/dL	12–16 g/dL
White blood cell count	1,900/mm³	4,000–10,000/mm³
Neutrophil count	1,200/mm³	1,500–7,500/mm³
Platelet count	50,000/mm³	150,000–400,000/mm³

Abdominal ultrasonography was performed as the initial imaging modality and demonstrated a normal-sized liver with preserved echotexture, associated with marked dilatation of both intrahepatic bile ducts and the common bile duct measuring 27.8 mm. Multiple intraductal microlithiasis were identified, along with gallbladder sludge and microlithiasis. Doppler examination failed to visualize the portal vein, which was replaced by multiple periportal venous collaterals consistent with cavernomatous transformation. Massive splenomegaly and porto-systemic collateral circulation were also noted, supporting the diagnosis of chronic portal hypertension secondary to extrahepatic portal vein obstruction.

To further characterize the vascular abnormalities and biliary involvement, computed tomography angiography was performed. CT confirmed chronic portal vein thrombosis with cavernomatous transformation and extensive porto-systemic collateral circulation involving the splenic and peri-hilar regions. Marked splenic vein dilatation and massive

splenomegaly were present. Significant dilatation of both intrahepatic and extrahepatic bile ducts was identified without clearly detectable intrinsic obstructive lesions, raising suspicion of portal cavernoma cholangiopathy with extrinsic biliary compression (Figure 1).

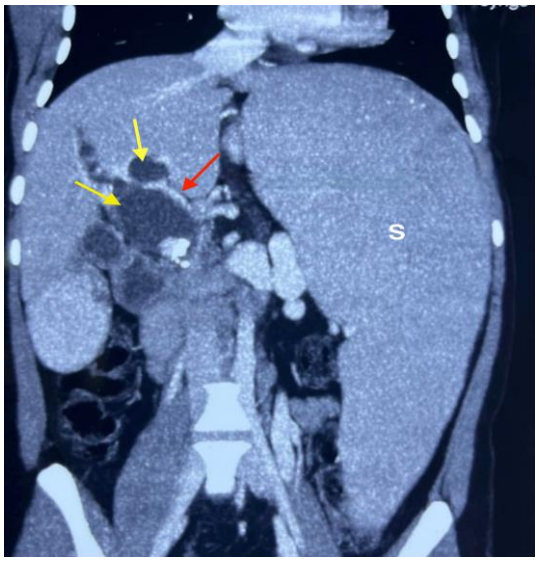

Figure 1: Coronal contrast-enhanced CT image demonstrating cavernomatous transformation of the portal vein with multiple hilar venous collaterals (red arrow), causing biliary compression (yellow arrow), associated with marked splenomegaly (S)

Magnetic resonance cholangiopancreatography (MRCP) combined with MR portography provided a more comprehensive evaluation of biliary and vascular abnormalities. MRCP demonstrated severe dilatation of the biliary tree with enlargement of the common bile duct up to 32 mm secondary to distal stricture of distal CBD. Associated intrahepatic lithiasis and gallbladder microlithiasis were identified. MR portography confirmed complete portal vein occlusion replaced by a serpiginous peri-cholédocien venous network extending along the extrahepatic bile duct. Multiple porto-systemic collateral pathways and massive splenomegaly containing Gamna-Gandy bodies were also observed. These findings were highly suggestive of advanced symptomatic portal cavernoma cholangiopathy with secondary biliary obstruction (Figure 2).

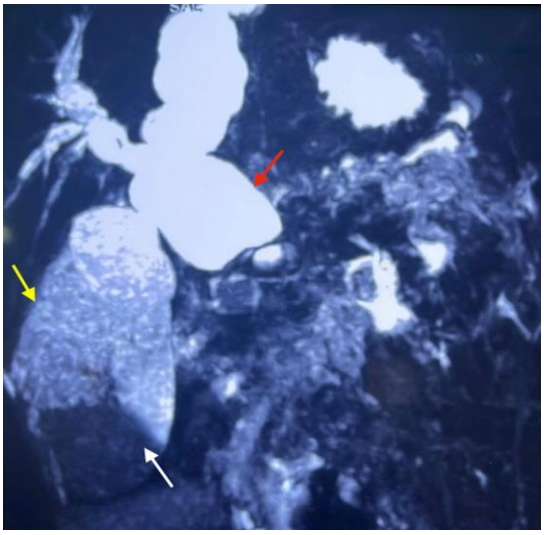

Figure 2: MR cholangiography demonstrating marked common bile duct dilatation (red arrow) due to portal cavernoma-associated biliary compression, accompanied by gallbladder distension (yellow arrow) and multiple gallstones (white arrow)

Endoscopic ultrasound (EUS) was performed to better assess the distal bile duct and peri-biliary vascular structures before therapeutic intervention. It confirmed marked dilatation of the common bile duct containing mobile sludge and demonstrated extensive peri-pancreatic venous collaterals surrounding the biliary tract (Figure 3). These findings were particularly

important for evaluating the risk of endoscopic sphincterotomy and procedure-related hemorrhage.

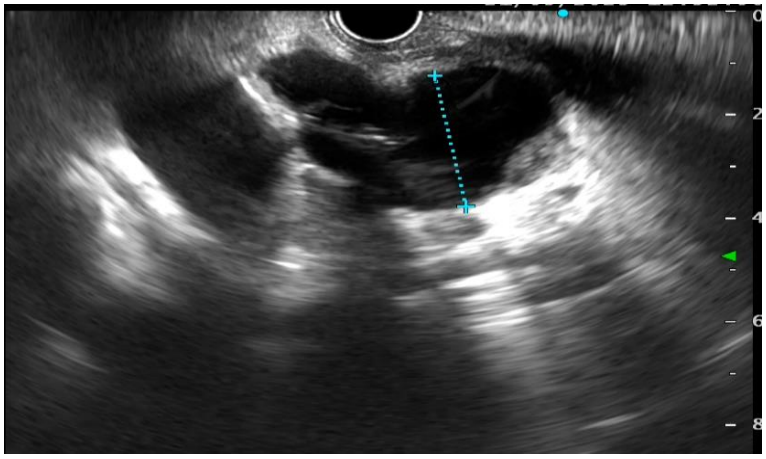

Figure 3: Endoscopic ultrasound showing marked common bile duct dilatation with intraluminal sludge

Therapeutic ERCP demonstrated severe dilatation of the extrahepatic bile duct with evidence of extrinsic compression at the suprapancreatic portion of the common bile duct, most likely related to portal venous collaterals. Multiple mobile biliary calculi and biliary sludge were identified within the dilated biliary tree. Given the extensive peri-biliary collateral circulation and the associated hemorrhagic risk, sphincterotomy was intentionally avoided. Endoscopic biliary drainage was achieved through placement of a plastic stent (10 Fr/9 cm), resulting in satisfactory bile flow and subsequent clinical improvement (Figure 4).

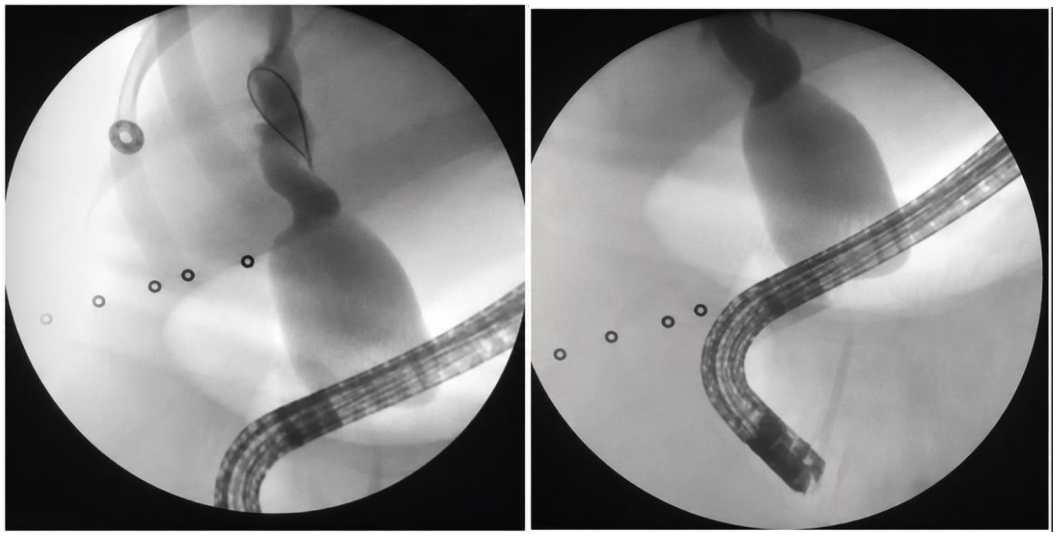

Figure 4: ERCP showing marked common bile duct dilatation with biliary drainage by plastic stent placement.

Clinical and biochemical improvement was observed following endoscopic drainage, with progressive reduction of total bilirubin levels from 9.8 mg/dL at 24 hours to 5.8 mg/dL after seven days. Upper gastrointestinal endoscopy performed during hospitalization identified small-to-medium grade II esophageal varices without red wale signs, not requiring ligation. Non-selective beta-blocker therapy was subsequently reintroduced.

Three months later, the patient developed acute cholangitis with recurrent jaundice and inflammatory syndrome. Repeat ERCP revealed obstruction of the previously inserted biliary stent by thick biliary sludge and microlithiasis. Cholangiography demonstrated saccular dilatation of the common bile duct measuring approximately 40 mm, containing abundant biliary debris and multiple calculi, associated with persistent intrahepatic ductal dilatation. Following removal of the obstructed stent, a new plastic prosthesis (10 Fr/12 cm) was inserted, allowing satisfactory biliary drainage.

Given the recurrence of cholangitis despite adequate endoscopic management, the persistence of biliary compression, severe portal hypertension, and massive splenomegaly, a multidisciplinary discussion favored definitive surgical management. The patient was considered to have advanced symptomatic portal

cavernoma cholangiopathy refractory to endoscopic treatment. A staged surgical approach was therefore undertaken. She initially underwent a mesocaval shunt in order to reduce portal hypertension and peri-biliary venous congestion, followed subsequently by cholecystectomy combined with Roux-en-Y hepaticojejunostomy.

Early postoperative outcome was favorable, with progressive resolution of jaundice and no immediate postoperative complications. The patient demonstrated satisfactory clinical recovery without recurrence of cholangitis or gastrointestinal bleeding during early follow-up. Continued multidisciplinary surveillance was recommended to monitor portal hypertension and long-term biliary status.

DISCUSSION

Portal cavernoma cholangiopathy (PCC) encompasses the spectrum of biliary tract abnormalities observed in patients with chronic extrahepatic portal vein obstruction presenting with cavernous transformation of the portal vein. Since the consensus published by Dhiman *et al.,* in 2014, the term "portal cavernoma cholangiopathy" has been preferred over older nomenclature such as portal biliopathy or portal hypertensive biliopathy, as it more accurately reflects the underlying vascular pathophysiology and biliary involvement observed in non-cirrhotic patients with chronic portal vein thrombosis [1]. Although radiological biliary abnormalities are frequently detected in patients with chronic portal obstruction, symptomatic presentations remain relatively rare and typically manifest only after several years of disease progression.

The pathophysiology of PCC is complex and multifactorial. The primary mechanism involves extrinsic compression of the bile ducts by the venous collateral networks that develop around the biliary tree following portal obstruction. Two main venous plexuses are implicated: the epicholedochal plexus of Saint and the paracholedochal plexus of Petren, both of which become markedly dilated due to portal hypertension and the formation of the portal cavernoma. This compressive component is further compounded by ischemic injury secondary to chronic alteration of the peribiliary microvasculature. This hypoperfusion can lead to biliary fibrosis, fixed strictures, and progressive cholangiopathy. Over time, chronic biliary stasis promotes the formation of biliary sludge, common bile duct (CBD) stones, recurrent cholangitis, and secondary intrahepatic lithiasis [2].

Our case report illustrates several prominent features of advanced, symptomatic PCC. The patient likely developed chronic portal vein thrombosis during the neonatal period, secondary to umbilical vein catheterization. Umbilical vein catheterization is a well-established risk factor for extrahepatic portal vein thrombosis in children and can remain asymptomatic for many years before presenting as complications of portal hypertension or biliary manifestations. In our patient, the prolonged latency period between the presumed neonatal thrombotic event and the onset of symptomatic cholangiopathy highlights the indolent, slowly progressive nature of this condition. The concomitant protein C and S deficiency may have further contributed to thrombus persistence and the subsequent progression toward severe chronic portal obstruction.

Our patient presented with a severe and complex form of PCC characterized by major intra- and extrahepatic ductal dilatation, massive common bile duct ectasia, intrahepatic lithiasis, recurrent bouts of acute cholangitis, and a prominent pericholedochal collateral network. Biliary stone formation in PCC is secondary to chronic cholestase, bile stasis, and ischemia of the biliary epithelium. According to recent series, common bile duct stones are found in approximately 33.8% of symptomatic patients. Furthermore, data shows that the overall prevalence of biliary abnormalities reaches 67% when patients are systematically evaluated using high-resolution imaging modalities [3-5]. The combination of biliary obstruction and a massive peribiliary collateral network significantly increases therapeutic complexity and elevates the procedural risks of interventions; notably, it exposes patients to a reported iatrogenic hemobilia rate of nearly 30% during endoscopic maneuvers such as balloon sweeping or stent exchange [6].

Imaging plays a pivotal role in the diagnosis and characterization of PCC. In our patient, a multimodal approach combining Doppler ultrasonography, CT angiography, magnetic resonance cholangiopancreatography (MRCP), endoscopic ultrasound (EUS), and endoscopic retrograde cholangiopancreatography (ERCP) allowed for a comprehensive assessment of both vascular and biliary anomalies. Our patient's biliary involvement was classified as Grade III according to the Llop *et al.,* classification (defined by the presence of a stricture associated with upstream dilatation), a grade known to carry a high positive predictive value (41%) for the development of clinical symptoms [7]. Morphologically, the patient’s profile corresponded to a mixed type according to the Shin *et al.,* classification, combining compressive remodeling with ischemic-appearing lesions [8].

MRCP remains the non-invasive gold standard of choice, as it provides a simultaneous, high-resolution evaluation of both the biliary tree and the portal system while avoiding the risks associated with invasive direct cholangiography [9]. In our case, MRCP clearly demonstrated complete occlusion of the main portal vein trunk, which was replaced by a serpiginous pericholedochal venous network, associated with massive biliary dilatation and distal choledocholithiasis.

EUS permitted a more precise evaluation of the peripancreatic collaterals responsible for the biliary compression, while ERCP confirmed the site of biliary obstruction and facilitated therapeutic drainage [10].

The management of symptomatic PCC remains highly challenging, and no standardized therapeutic algorithm is currently universally accepted. Endoscopic therapy typically serves as the first-line intervention in patients presenting with acute cholestasis, cholangitis, or mechanical biliary obstruction. ERCP with biliary drainage and plastic or fully couverte metallic stent placement provides effective temporary decompression of the biliary tract; however, recurrence rates remain high because endoscopic management does not address the underlying portal hypertension, leading to a long-term re-intervention rate of approximately 50% [11]. In our patient, initial endoscopic drainage achieved transient clinical and biochemical improvement, but a recurrence of acute cholangitis occurred three months later due to stent occlusion by sludge and microcalculi, necessitating a repeat endoscopic intervention.

In advanced stages of PCC, portal decompression via surgical or radiological portosystemic shunts represents the only etiological treatment capable of altering the natural history of the disease. Currently, interventional radiology via portal vein recanalization combined with transjugular intrahepatic portosystemic shunt (PVR-TIPS) has emerged as a robust, modern alternative, boasting a median technical success rate of 91% [12]. Several studies have demonstrated that portosystemic decompression leads to a dramatic regression of compressive biliary abnormalities and a significant reduction in infectious biliary episodes.

Nevertheless, when fixed strictures, significant lithiasis, or irreversible biliary lesions have developed due to the ischemic component, complementary biliary surgery may still be required following successful portal decompression. In our case, the severity of the portal hypertension, massive splenomegaly, recurrent cholangitis, and persistent cholangiopathy led to a logical, sequential surgical strategy: an initial mesenteric-caval shunt for vascular decompression, followed by a cholecystectomy combined with a Roux-en-Y hepaticojejunostomy [13].

This case report illustrates the progressive nature of portal cavernoma cholangiopathy and underscores the importance of early diagnosis of biliary manifestations in patients with chronic portal vein thrombosis. It highlights the critical role of multimodal imaging and emphasizes the necessity of a multidisciplinary approach involving hepatologists, advanced endoscopists, interventional radiologists, and hepatobiliary surgeons. Finally, our case highlights the limitations of isolated endoscopic treatment in advanced forms of PCC and supports the benefit of a combined approach utilizing portosystemic decompression alongside definitive biliary surgery in patients presenting with complex, mixed, and recurrent biliary involvement.

CONCLUSION

Portal cavernoma cholangiopathy (PCC) is a progressive complication of chronic extrahepatic portal vein thrombosis that requires early detection via high-resolution multimodal imaging (MRCP and EUS). While isolated endoscopic therapy is effective for immediate biliary decompression during acute cholangitis, it yields high long-term recurrence rates as it does not address the underlying portal hypertension. Ultimately, this case demonstrates that for advanced, mixed-type PCC, a sequential multidisciplinary approach combining surgical portosystemic decompression with definitive biliary reconstruction (Roux-en-Y hepaticojejunostomy) is the most effective strategy to achieve sustained clinical resolution and prevent secondary biliary cirrhosis.

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