Van Med J 32 (4):233-239, 2025
DOI: 10.5505/vmj.2025.41736

Radiological Findings and Diagnostic Clues of Portal Biliopathy Secondary to Extrahepatic Portal Vein Thrombosis

Ahmet Akçay

Bezmialem Vakif University, Faculty of Medicine, Department of Radiology, Istanbul, Türkiye

Abstract

Introduction: Portal biliopathy (PB) refers to biliary strictures that develop as a result of chronic extrahepatic portal vein thrombosis (EHPVT), most commonly due to compression by collateral venous structures such as the epicholedochal and paracholedochal plexuses. On imaging, PB can mimic both benign and malignant biliary conditions, which may lead to diagnostic uncertainty and unnecessary invasive procedures. Accurate radiologic recognition is therefore essential for guiding clinical management. This study aims to evaluate the radiologic and biochemical features of PB and to identify imaging findings that may facilitate early and non-invasive diagnosis.

Materials and Methods: This retrospective study included 15 patients clinically and radiologically diagnosed with PB secondary to chronic EHPVT between January 2018 and December 2024.Imaging was assessed by an abdominal radiologist with 5 years of experience. Biliary changes, collateral vessel distribution, and lesion characteristics were evaluated using ultrasound, Doppler ultrasound, Computed Tomography (CT), and Magnetic Resonance Imaging (MRI). Biochemical parameters and clinical records were reviewed.

Results: All patients demonstrated biliary strictures due to peribiliary collateral compression. Collateral types were classified as varicoid in 8 patients (53.3%), fibrotic in 3 (20%), and mixed in 4 (26.7%). Two patients (13.3%) had MFPB with lesions that were T1-weighted hyperintense, T2- weighted hypointense, showed delayed enhancement, and no diffusion restriction. Alkaline phosphatase (ALP), and gamma-glutamyl transferase (GGT) were elevated in 80% of cases; bilirubin was elevated in 46.7%. Endoscopic retrograde cholangiopancreatography (ERCP) was performed in 5 patients (33.3%) for symptomatic management; the remainder were treated conservatively.

Conclusion: PB can be identified with characteristic radiologic findings, particularly in the absence of chronic liver disease. Awareness of paracholedochal compression and mass-forming changes in the setting of portal vein thrombosis is crucial. Accurate radiologic recognition can guide proper clinical management and help avoid unnecessary invasive procedures.

Key words: Biliary stenosis; portal vein thrombosis; portal hypertension; bile duct diseases; portal biliopathy

Introduction

Portal biliopathy (PB) refers to the development of bile duct strictures caused by chronic extrahepatic portal vein thrombosis (EHPVT), without any underlying primary biliary tract pathology (1). In cases of chronic EHPVT, collateral vessels that form at the liver hilum are called portal cavernomas. The term "PB" serves as an umbrella definition and includes more specific conditions such as portal cavernoma biliopathy and mass-forming portal cavernoma biliopathy (MFPB), which are named based on the predominant process leading to bile duct stenosis [2]. The pathogenesis of portal biliopathy involves extrinsic compression of the common bile duct by dilated paracholedochal (Petren's plexus) veins, mural thickening and cholestasis due to intramural expansion of epicholedochal (Saint's plexus) veins, and secondary compression from fibrotic soft tissue in the pericholedochal region (2,3). This fibrotic tissue is believed to develop due to ischemic injury to the bile duct wall, which occurs as a consequence of chronic EHPVT. MFPB leads to extrinsic compression, bile duct narrowing, and in some cases, a mass-like pericholedochal appearance that can mimic cholangiocarcinoma (4,5). PB often remains asymptomatic for years; however, complications such as cholangitis, biliary lithiasis, or secondary biliary cirrhosis may arise, necessitating clinical intervention (6). Radiologic imaging, particularly MRI, plays a critical role in the differentiation of portal biliopathy from both benign conditions such as primary sclerosing cholangitis and IgG4-related sclerosing cholangitis, and malignant entities including cholangiocarcinoma and periampullary tumors. By aiding in this distinction, imaging helps overcome diagnostic challenges and reduces the need for unnecessary invasive procedures (7). This study aims to describe the imaging features and biochemical findings of portal biliopathy. Biliary changes and the

*Corresponding Author: Ahmet Akçay Department of Radiology, Faculty of Medicine, Bezmialem Vakif University, Istanbul, Türkiye
Email: ahmetakcaymd@gmail.com **Orcid:** Ahmet Akçay 0000-0001-7016-6004

Received: 27.05.2025, Accepted: 11.07.2025

distribution of portacaval collaterals were assessed on ultrasound, Computed Tomography (CT), and Magnetic Resonance Imaging (MRI). In a subset of patients with mass-forming appearance, imaging findings were also reviewed to differentiate portal biliopathy from malignant biliary lesions. Recognizing these features may help avoid unnecessary invasive procedures and improve clinical decision-making.

Materials and Methods

This retrospective study included 15 patients who were clinically and radiologically diagnosed with PB secondary to EHPVT between January 2018 and December 2024. Inclusion criteria were: (1) radiologically confirmed chronic portal vein thrombosis, and (2) biliary strictures attributed to PB—either due to extrinsic compression by collateral vessels or MFPB—demonstrated on contrast-enhanced MRI/MRCP, contrast-enhanced CT, ultrasound, or Doppler ultrasound. Exclusion criteria included patients with biliary strictures resulting from other causes, such as primary hepatic or biliary diseases (e.g., cirrhosis, primary sclerosing cholangitis), choledocholithiasis, or biliary malignancies. Radiological assessments were independently reviewed by an abdominal radiologist with 5 years of experience. The location of thrombosis within the portal venous system was classified into the portal confluence, splenic vein, superior mesenteric vein (SMV), and portal hilum.The distribution of portoportal collateral vessels was categorized based on contrast-enhanced CT or MRI into the following anatomical regions: pericholedochal, peripancreatic, periportal intrahepatic, perigastric-paraesophageal, and splenorenal. Collaterals causing compression on the common bile duct were further classified according to Shin et al. 's system into paracholedochal, intramural/epicholedochal, or mixed types, based on MRCP and contrast-enhanced CT findings [8]. MFPB was defined as a T2-hypointense lesion surrounding the bile ducts, showing mild delayed enhancement without diffusion restriction on MRI, in line with descriptions in the literature [9]. Apparent diffusion coefficient (ADC) measurements were obtained by manually placing regions of interest (ROIs) on the lesion, and the mean value of the three lowest measurements was calculated for analysis.Clinical records were reviewed to assess underlying thrombotic risk factors, including hypercoagulable states, history of malignancy, or idiopathic etiologies. Additional clinical data—including positron emission tomography-computed tomography (PET-CT), endoscopic retrograde cholangiopancreatography (ERCP), histopathological reports, and biochemical parameters [direct bilirubin, alkaline phosphatase (ALP), and gamma-glutamyl transferase (GGT)]-as well as follow-up records, were documented. Patients with obstructive jaundice or recurrent cholangitis underwent endoscopic retrograde cholangiopancreatography (ERCP). Patients who did not require ERCP were managed conservatively with radiological follow-up and symptomatic treatment.

Ethical approval: The study was approved by the institutional ethics committee (Approval No: E-54022451-050.04-190800).

Statistical analysis: All statistical analyses were performed using IBM SPSS Statistics for Windows, Version 26.0 (IBM Corp., Armonk, NY, USA). Continuous variables were summarized as mean, standard deviation, and range, while categorical variables were presented as counts and percentages. Normality of continuous variables was assessed using the Shapiro-Wilk test. Comparisons between categorical variables were made using the chi-square test. Differences in continuous variables between two independent categorical groups were analyzed using the Mann-Whitney U test due to non-normal distribution in some variables. A p-value < 0.05 was considered statistically significant.

Results

A total of 15 patients were included, with a mean age of 52.2 ± 15.2 years (range: 13–76). The male-to-female ratio was 9:6 (60% male, 40% female). The mean follow-up period was 4.07 ± 3.5 years (range: 1–10 years). The most common portal vein thrombosis localization was the confluence region 12 cases (80%), followed by the splenic vein 1 case (6.7%), SMV 1 case (6.7%), and portal hilum 1 case (6.7%). All patients showed cavernous transformation of the portal vein on imaging, evidenced by multiple serpiginous collaterals in the porta hepatis (portal cavernoma). The distribution of portoportal collateral types was as follows: pericholedochal: 13 patients (86.7%), peripancreatic: 4 patients (26.7%), intrahepatic periportal: 4 patients (26.7%), perigastric-paraesophageal: 3 patients (20%), pericholecystic 5 patients (33.3%) and splenorenal 3 patients (%20). On radiological imaging, biliary abnormalities were present in all patients, ranging from smooth indentations to multi-focal strictures of the common bile duct (CBD) and hepatic ducts with upstream dilation. In all patients, imaging revealed biliary strictures caused by compression from peribiliary collateral vessels. In 8 patients (53.3%), external compression by dilated paracholedochal plexus was identified (figure 1).

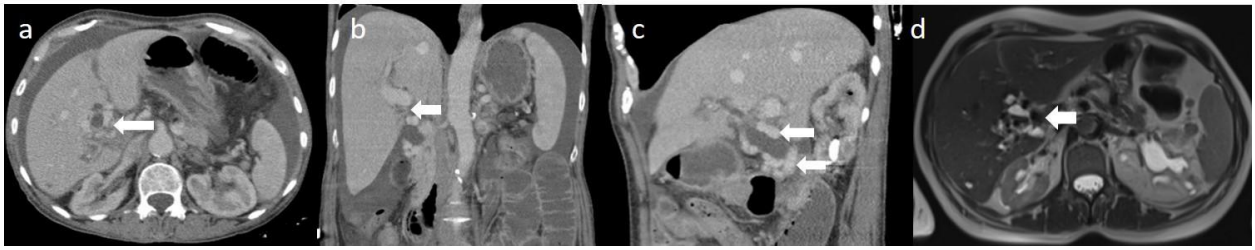

Figure 1: Axial contrast-enhanced Computed Tomography (CT) image (a) shows collateral venous shunts compressing the main bile duct (arrow). Coronal reformatted CT image (b) reveals severe luminal narrowing of the portal vein due to thrombosis (arrow), and sagittal reformatted CT image (c) shows pericholedochal vascular dilations compressing the common bile duct (arrow). Axial T2-weighted image (d) displays hypointense signal voids in collateral vessels compressing the main bile duct (arrow).

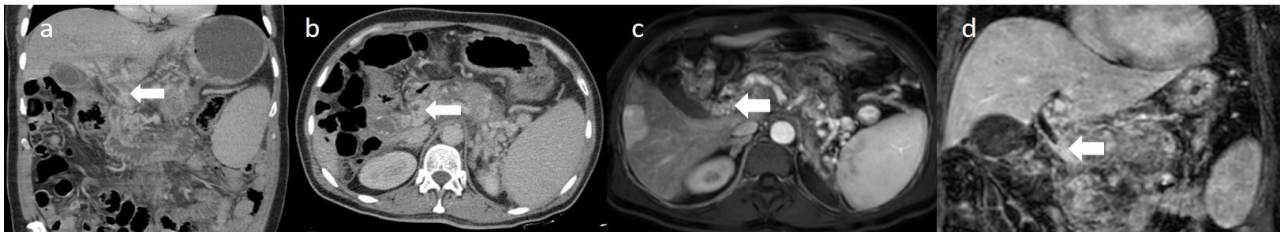

Figure 2: Coronal reformatted (a) and axial (b) Computed Tomography images show common bile duct wall thickening from dilated epicholedochal collaterals (arrow). Contrast-enhanced axial T1-weighted image (c) shows pericholecystic portacaval collaterals (arrow), and coronal T1-weighted image (d) shows distal bile duct compression (arrow) from dilated epicholedochal vessels.

Table 1: Patient Characteristics and Portal Biliopathy Findings

Parameter	Subcategory	(n, %)
Mean age (years)	—	52.2 ± 15.2 (13–76)
Gender Distribution (n, %)	Male	9 (60%)
	Female	6 (40%)
Mean follow-up duration (years)	—	4.07 ± 3.5 (1–10)
Porto-portal Collateral Types (n, %)	Pericholedochal	13 (86.7%)
	Pericholecystic	5 (33.3%)
	Peripancreatic	4 (26.7%)
	Intrahepatic periportal	4 (26.7%)
	Perigastric-paraesophageal	3 (20%)
	Splenorenal	3 (20%)
Portal Biliopathy Pathogenesis (n, %)	External compression (paracholedochal plexus)	8 (53.3%)
	Mural thickening (epicholedochal plexus)	3 (20%)
	Mixed (paracholedochal & epicholedochal)	4 (26.7%)
	Mass-forming portal biliopathy	2 (13%)

In 3 patients (20%), mural thickening and stenosis were due to epicholedochal plexus involvement (figure 2*)*. In 4 patients (26.7%), a mixed type with contributions from both plexuses was observed (Table 1). Two patients (13%) manifested a MFPB pattern: MRI revealed an ill-defined, solid-appearing periportal mass encasing the CBD, which was T1-weighted hyperintense, T2-weighted hypointense with

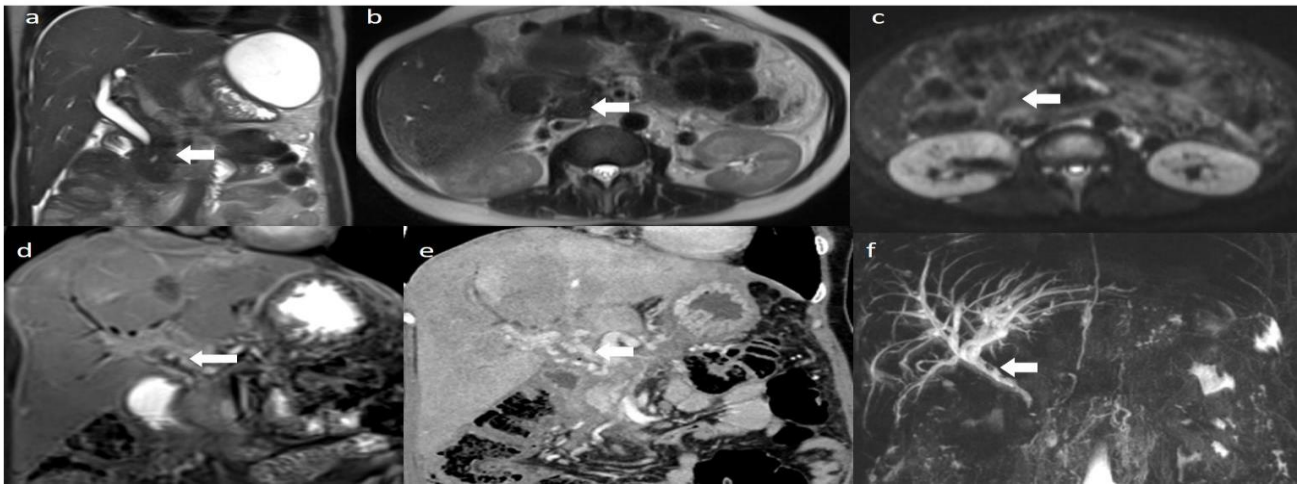

Figure 3: Coronal (a) and axial (b) T2-weighted images show hypointense soft tissue around the common bile duct (arrow), indicating mass-forming PB. Diffusion Weighted Imaging (c) reveals no significant diffusion restriction in the lesion (arrow). Contrast-enhanced T1-weighted image (d) and coronal reformatted Computed Tomography image (e) images show dilated collateral veins (arrow) along the common bile duct and central intrahepatic duct dilation. Follow-up Magnetic Resonance Cholangiopancreatography image (f) shows gallstones (arrow) in the common bile duct.

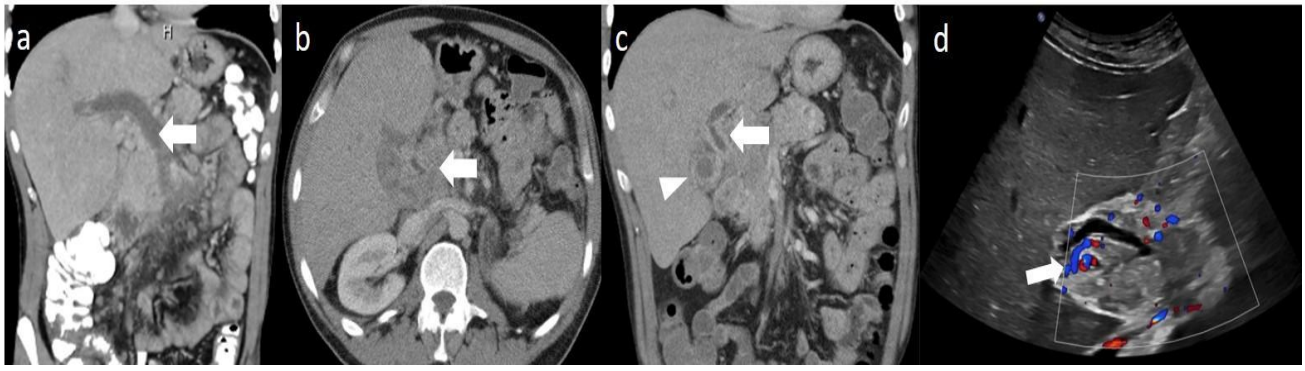

Figure 4: Coronal reformatted abdominal Computed Tomography (CT) image (a) shows portal vein thrombosis (arrow). Axial CT (b) reveals soft tissue thickening from mass-forming PB (arrow) compressing the common bile duct. Coronal reformatted CT image (c) displays pericholedochal (arrow) and pericholecystic (arrow head) collateral veins. Colored Doppler Ultrasonography image (d) shows color Doppler signals (arrow) indicating vascularity in the pericholedochal soft tissue.

mild delayed contrast enhancement (figure 3,4). Both cases also showed mixed-type (paracholedochal and epicholedochal) venous collaterals. These lesions radiologically mimicked a tumor. In the presence of EHPVT, multiple portoportal collateral vessels were observed in the peribiliary region. No diffusion restriction was noted, with the lesions appearing hypointense on both diffusion weighted imaging (DWI) and ADC maps, findings that favored a diagnosis of MFPB. The measured ADC values in these patients were 1100 mm^2/s and 1190 mm^2/s, respectively. In one patient diagnosed with MFPB, PET-CT revealed no increased fluorodeoxyglucose (FDG) uptake in the lesion and no pathological lymphadenopathy, and subsequent endoscopic ultrasound (EUS)-guided biopsy showed no histopathological evidence of malignancy, instead demonstrating fibrotic soft tissue associated with vascular structures. Among the 15 patients, essential thrombocytosis was identified in 4 cases (26.7%), Janus kinase 2 V617F (JAK2 V617F) mutation-related thrombophilia in 1 case (6.7%), and chronic pancreatitis as the presumed etiology in 2 cases (13.3%). Extra-pancreaticobiliary malignancies (breast, colon, or cervical cancer) were present in 3 patients (20%), while the remaining 5 patients (33.3%) were classified as idiopathic. Serum ALP and GGT levels were elevated in 12 patients (80%), with mean values of 280 ± 85 U/L (normal range: 40–150 U/L) and 210 ± 75 U/L (normal range: <73 U/L), respectively. No statistically significant difference in ALP and GGT levels was observed between patients with epicholedochal and paracholedochal plexus involvement ($p > 0.05$). Elevated total bilirubin levels were noted in 7 patients (46.7%), with a mean of 4.55 mg/dL (range: 0.42–12.69 mg/dL). Similarly, there was no significant difference in the presence or level of hyperbilirubinemia among the collateral subtypes

($p > 0.05$). Of the 15 patients, 5 (33.3%) underwent ERCP, primarily for biliary drainage and symptomatic relief of cholestasis. Among these, 2 patients required biliary stent placement due to persistent strictures despite initial balloon dilation, while 3 patients underwent successful balloon dilation and sphincterotomy without the need for stenting. The remaining 10 patients (66.7%) were managed conservatively with radiological follow-up and symptomatic treatment. None of the conservatively managed patients developed progressive biliary obstruction during the follow-up period.

Discussion

Among 15 patients with chronic extrahepatic portal vein thrombosis, biliary strictures resulted exclusively from peribiliary collateral compression—paracholedochal in eight, epicholedochal in three, and mixed in four cases. Two patients exhibited mass-forming portal biliopathy, characterized on MRI by T1 hyperintensity, T2 hypointensity, mild delayed contrast enhancement, and absence of diffusion restriction on DWI and ADC maps, facilitating differentiation from malignant biliary strictures. Although 80 % of patients demonstrated elevated cholestatic enzymes, these laboratory values did not distinguish collateral subtype. One-third of the cohort underwent endoscopic intervention, while the remainder were managed conservatively without progression of obstruction. These findings underscore the critical importance of radiologic imaging in the noninvasive diagnosis and management of portal biliopathy. Similar to the findings reported by Walser et al., in all of our subjects, portal vein thrombosis occurred without underlying chronic liver disease, with hypercoagulability and malignancy being the main contributing factors (4). This distinction is crucial, as PB is more likely to develop in cases of portal vein thrombosis without chronic liver disease. In patients with chronic liver disease, portal venous flow is typically redirected through the gastroesophageal varices or the coronary vein, which gradually enlarges due to reduced hepatopetal portal perfusion. However, in the absence of chronic liver disease, collateral circulation primarily drains through the anterior and posterior superior pancreaticoduodenal veins [5]. Consistent with this hemodynamic pattern, our study demonstrated that the most common locations for collateral vessel development were the pericholedochal, pericholecystic, and peripancreatic regions. In contrast, collateral formation in the perigastric area was observed in only a small number of patients. These, along with paracholedochal veins, cause compression on the common bile duct, resulting in biliary stenosis and the typical obstruction observed in PB. This distinct vascular pathway may play a significant role in the increased occurrence of PB in these patients. In PB, beyond pericholedochal plexus compression, MFPB involves soft tissue proliferation around the common bile duct, leading to strictures. The underlying mechanisms of pericholedochal soft tissue thickening remain a subject of debate in the literature. One hypothesis proposes that it arises from ischemia and inflammation of the bile duct wall secondary to chronic EHPVT, while another suggests it may be due to chronic hypertrophy of the epicholedochal plexus (4,6). Importantly, MFPB can mimic malignant biliary strictures. The differential diagnosis includes cholangiocarcinoma, primary biliary lymphoma, and IgG4-related sclerosing cholangitis. A key distinguishing feature of MFPB is the presence of extrahepatic portal vein thrombosis (EHPVT) and pericholedochal venous dilatation, without evidence of chronic liver disease or malignancy-related invasion. MFPB can be differentiated from malignant strictures on MRI through its characteristic imaging features: hyperintensity on T1-weighted images, hypointensity on T2-weighted images, and mild delayed enhancement following contrast administration, as also demonstrated by our cases, which showed no diffusion restriction (hypointense on DWI and ADC maps). Additionally, the lack of increased FDG uptake on PET-CT serves as a useful diagnostic clue (7). Similarly, IgG4-related cholangiopathy can present with bile duct wall thickening and extrahepatic peribiliary mass-like lesions that cause cholestasis, making it a critical differential diagnosis in suspected PB cases. However, typical features of IgG4-related disease—such as multisystem involvement, elevated serum IgG4 levels, increased FDG uptake on PET-CT, and diffusion restriction on DWI—help distinguish it from MFPB. In the absence of these findings, diagnostic uncertainty may persist. In such cases, close clinical monitoring and a favorable response to steroid therapy can support the diagnosis of IgG4-related cholangiopathy (8–10). Portal biliopathy is traditionally categorized into varicoid, fibrotic, and mixed types based on collateral distribution and bile-duct morphology (11). The predominance of paracholedochal (varicoid) compression in our cohort underscores the hemodynamic rerouting that occurs in non-cirrhotic EHPVT, as previously described by Walser et al. (4), and suggests why decompressive shunting may yield favorable outcomes in these patients. Mixed patterns—reflecting both epicholedochal thickening and paracholedochal compression—mirror the combined remodeling process outlined by Shin et al. (11) and have been associated with more refractory biliary stenosis (8),

indicating a need for adjunctive endoscopic interventions. Although fibrotic-predominant cases are thought to be less amenable to vascular decompression and may require stenting (12), our limited sample size precludes definitive conclusions. Nonetheless, these subtype-specific insights highlight the value of detailed imaging classification for tailoring therapeutic strategies. Similarly, no significant differences in serum ALP, GGT, or bilirubin levels were found among the collateral subtypes. Although no statistically significant differences were detected, this may be attributed to the limited sample size and consequently low statistical power. Further studies with larger cohorts are needed to clarify both the prognostic implications of collateral types and their potential relationship with the severity of biochemical cholestasis. Although cavernous transformation develops in nearly all patients with EHPVT, symptomatic PB is observed in only 5-30% of cases (2). In our study, ERCP was performed in 5 of 15 patients (33.3%) to manage biliary obstruction and related symptoms, in line with previously reported intervention rates for symptomatic portal biliopathy. In these patients, cholestasis may lead to complications such as cholelithiasis, cholangitis, or hepatic abscess. If not treated, the condition can progress to secondary biliary cirrhosis over time (13). Asymptomatic patients generally do not require active treatment. In symptomatic cases, both endoscopic and surgical options are available, although the most effective treatment strategy remains unclear (14). Endoscopic interventions—such as sphincterotomy, balloon dilatation, and stent placement—are commonly used as first-line therapies. When endoscopic treatment is unsuccessful or not feasible, surgical approaches like portosystemic shunt creation or hepaticojejunostomy may be necessary (15).

Study limitations: This study has several limitations. Histopathological confirmation was not available in all cases; however, portal biliopathy is a condition that can often be reliably diagnosed through clinical and radiological findings. Additionally, the retrospective design and limited sample size may affect the generalizability of the results.

Conclusion

In conclusion, our study highlights important radiologic clues for identifying PB, a rare condition that can mimic malignant biliary strictures. This resemblance is particularly common in MFPB. Accurate diagnosis requires recognizing specific imaging findings on contrast-enhanced CT and MRI. Combining these radiologic findings with clinical and laboratory data helps ensure correct diagnosis and appropriate treatment. Early recognition through imaging can prevent unnecessary invasive procedures and their potential complications, such as cholangitis or pancreatitis. Ultimately, our findings emphasize the key role of radiologic imaging in managing patients with PB effectively.

Ethical approval: The study was approved by the institutional ethics committee (Approval No: E-54022451-050.04-190800).

Conflict of Interest: The authors declare no conflict of interest.

Funding statement: No external funding was received for this study.

Author contributions: Conceptualization (A.A.); Study Design (A.A.); Data Collection (A.A.); Data Analysis and Interpretation (A.A.); Writing – Original Draft (A.A.); Writing – Review & Editing (A.A.)

References

1. Buyruk AM, Erdoğan Ç, Tekin F, Turan İ, Özütemiz Ö, Ersöz G. The use of fully covered self-expandable metal stents in the endoscopic treatment of portal cavernoma cholangiopathy. *BMC Gastroenterol.* 2023;23(1):414. .
2. Nikpanah M, Sailer A, Khoshpouri P, Panda A. Mass-forming Portal Cavernoma Cholangiopathy. *Radiographics.* 2023;43(9):e230074.
3. Hulshoff JB, Cuperus FJC, de Haas RJ. Mass-Forming Portal Biliopathy Presenting as Extreme Wall-Thickening of the Common Bile Duct. *Diagnostics (Basel).* 2020;10(9):623.
4. Walser EM, Runyan BR, Heckman MG, Bjarnason H, Andrews JC, Schleck CD, et al. Extrahepatic portal biliopathy: proposed etiology on the basis of anatomic and clinical features. Radiology 2011; 258(1): 146–153.
5. Sarma MS, Ravindranath A. Portal Cavernoma Cholangiopathy in Children and the Management Dilemmas. *J Clin Transl Hepatol.* 2020;8(1):61-68.
6. Abud A, Maddur H, Salem R. Management of Symptomatic Portal Cavernoma Cholangiopathy With Transplenic Portal Vein Recanalization and Transjugular Intrahepatic Portosystemic Shunt. *Hepatology.* 2021;73(1):456-459.
7. Moomjian LN, Winks SG. Portal cavernoma cholangiopathy: diagnosis, imaging, and intervention. Abdom Radiol (NY) 2017; 42(1): 57–68.
8. Madhusudhan KS, Das P, Gunjan D, Srivastava DN, Garg PK. IgG4-related sclerosing cholangitis: a clinical and imaging

review. AJR Am J Roentgenol 2019; 213(6): 1221–1231.
9. Vibert E, Azoulay D, Aloia T, Pascal G, Veilhan LA, Adam R, et al. Therapeutic strategies in symptomatic portal biliopathy. Ann Surg 2007; 246(1): 97–104.
10. Kim SY, Moon SH, Cho YA, Lee SM, Kim JH. Portal biliopathy misdiagnosed as hilar cholangiocarcinoma. Korean J Gastroenterol. 2022 Jul 25;80(1):38-42.
11. Shin SM, Kim S, Lee JW, Lee JH, Park SH, Yu E, et al. Biliary abnormalities associated with portal biliopathy: evaluation on MR cholangiography. AJR Am J Roentgenol 2007; 188: 341–347.
12. Chevallier P, Denys A, Novellas S, Schmidt S, Schnyder P, Bruneton JN. Magnetic resonance cholangiography features of biliary abnormalities due to cavernous transformation of the portal vein. Clin Imaging 2006; 30(3): 190–194.
13. Perlemuter G, Bejanin H, Fritsch J, Delaitre B, Buffet C, Amouyal G, et al. Biliary obstruction caused by portal cavernoma: a study of 8 cases. J Hepatol 1996; 25(1): 58–63.
14. Kotinda APST, Poujol-Robert A, Payance A, Leenhardt R, Camus Duboc M, Dray X et al. Endoscopic ultrasound evaluation of portal cavernoma cholangiopathy and endoscopic management of choledochal variceal rupture during ERCP. *Endoscopy*. 2024;56(S 01):E39-E40.
15. Giri S, Kale A, Singh A, Shukla A. Long-Term Outcomes of Endoscopic Management of Patients with Symptomatic Portal Cavernoma Cholangiopathy with No Shuntable Veins for Surgery or Failed Surgery. *J Clin Exp Hepatol*. 2022;12(4):1031-1039.