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BILIOPATHIE PORTALE A RARE COMPLICATION OF DREPANOCYTOSE : PORTAL BILIOPATHY A CASE REPORT.

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ABSTRACT

Portal biliopathy is a rare complication of chronic portal vein thrombosis, exceptionally reported in sickle cell disease. We describe a 16-year-old girl with sickle cell thalassemia who developed portal cavernoma, collateral circulation, hepatomegaly, gallstones, splenosis, and biochemical cholestasis after splenectomy and extensive portal vein thrombosis. The condition results from extrinsic compression of the bile ducts by venous collaterals and ischemic changes of the peribiliary plexus, mechanisms exacerbated by the hypercoagulability of sickle cell disease. Imaging, particularly MRCP, is central to diagnosis, while management ranges from surveillance to endoscopic or surgical intervention depending on clinical severity. Awareness of this entity is essential to avoid confusion with malignant biliary obstruction and to guide appropriate multidisciplinary care.

KEYWORDS

Portal biliopathy, sickle cell disease, portal vein thrombosis, MRCP, case report

MAIN ARTICLE

Introduction

Sickle cell disease is a chronic hemoglobinopathy characterized by recurrent episodes of vaso-occlusion and intravascular hemolysis. These mechanisms lead to multisystem complications, including hepatic vascular damage. Among these, portal biliopathy, although rare, is clinically significant. It is defined as an abnormality of the bile ducts secondary to extrinsic compression by hypertrophied collateral veins and varices at the hepatic hilum, resulting in a portal cavernoma [1,2].

Results

Case Report :

We report the case of a 16-year-old female patient followed since the age of 3 years for sickle cell thalassemia, initially diagnosed in the context of arthralgia, asthenia, and severe anemia, for which she required a transfusion program.

She later developed splenic infarction on splenomegaly, treated by splenectomy. Subsequently, she presented with extensive portal vein thrombosis involving the intrahepatic portal branches and the superior mesenteric vein, complicated by collateral venous circulation forming a portal cavernoma. Gallstones and laboratory evidence of biliary cholestasis were also observed.

Abdominal CT (portal venous phase) :

The examination showed extensive thrombosis of the portal vein extending to the bilateral intrahepatic branches and the superior mesenteric vein, with the development of collateral circulation arising from the epigastric and paracholedochal venous plexuses, resulting in a portal cavernoma. There was hepatomegaly without dysmorphia, with heterogeneous contrast uptake more pronounced at the periphery. Intrahepatic bile ducts were visualized. Extensive collateral circulation was present in the perihepatic, perisplenic, peripancreatic, and left gastric regions. Multiple splenosis nodules were seen at the splenectomy site. (figure 1)

Discussion

In older children, the main causes of portal vein thrombosis include pylephlebitis, colectomy in the context of inflammatory bowel disease, splenectomy, splenic embolization, sickle cell disease, and treatment with asparaginase [3,4].

In the context of sickle cell disease, portal biliopathy develops through two main mechanisms. The first is extrinsic compression of the bile ducts by dilated venous collaterals, particularly the paracholedochal and epicholedochal plexuses, secondary to chronic portal venous obstruction. This is explained by the mechanical hypothesis whereby increased paracholedochal venous pressure above 15 mmHg compresses the bile duct, which normally has a pressure of 4–10 mmHg [5]. The second mechanism is ischemic injury due to impaired vascularization of the bile ducts, further aggravated by microvascular thrombosis typical of sickle cell disease, leading to stenosis, fibrosis, or strictures [6].

Although most patients remain asymptomatic, some may develop chronic cholestasis, abdominal pain, recurrent cholangitis, or gallstone disease. Symptoms are usually correlated with the extent of radiological abnormalities and the duration of obstruction [1,7].

Diagnosis relies on imaging. Doppler ultrasound is considered the first-line modality for detecting portal hypertension, varices, and bile duct dilatation [8]. CT scan is excellent for demonstrating venous collaterals and hepatic parenchymal changes, with periportal varices appearing as enhancing tubular structures after contrast injection [3]. MRI and MRCP are the modalities of choice for evaluating the biliary tree, showing segmental stenoses, dilatations, extrinsic compression, and wall thickening. They provide excellent soft-tissue resolution, avoid radiation exposure, and help differentiate ischemic changes from fibrosis [5,9]. Angiography is reserved for complex cases, particularly to guide therapeutic interventions such as transjugular intrahepatic portosystemic shunts [10].

Radiological classification distinguishes three types of portal biliopathy [2]: varicose, with multiple irregular stenoses that may mimic cholangiocarcinoma; fibrotic, with dominant strictures whose enhancement kinetics help differentiate them from malignant lesions; and mixed or pseudotumoral forms.

The natural history ranges from chronic cholestasis to hepatic atrophy and gallstone formation [1,4].

Management depends on symptoms and severity. Asymptomatic patients require clinical and radiological follow-up. Symptomatic cases may require endoscopic biliary stenting, surgical

portosystemic shunts, anticoagulation to prevent progression of thrombosis, and liver transplantation in advanced cases [6,7].

Conclusion

Portal biliopathy is a rare but clinically relevant complication of sickle cell disease. Diagnosis relies mainly on imaging, with MRCP being the most useful modality. Management requires a multidisciplinary approach. Understanding the pathophysiology and the optimal use of imaging techniques is essential to improve patient outcomes.

FIGURES:

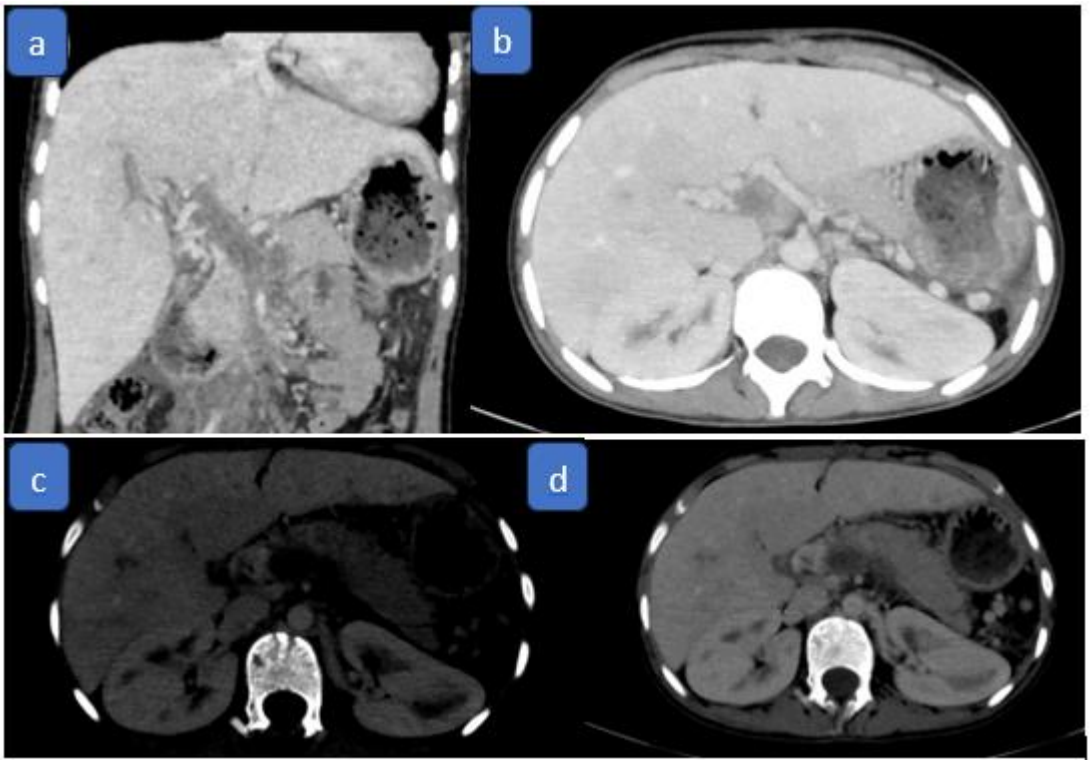

Figure 1: Coronal and axial CT scans during portal phase showing extensive portal vein thrombosis with development of a portal cavernoma. Axial CT in arterial phase shows heterogeneous hepatomegaly with visible bile ducts. Splenectomy site with splenosis nodules is also noted.

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REFERENCES

1. Dhiman RK, Chawla Y, Vasishta RK, et al. Portal hypertensive biliopathy. Gut. 2007;56(7):1001-1008.
https://doi.org/10.1136/gut.2006.103606

2. Walser EM, et al. Portal Biliopathy: Imaging and Clinical Spectrum. Radiology. 2011;258(2):461-469.
https://doi.org/10.1148/radiol.10090923

3. Shin SM, Kim S, Lee JW, et al. Biliary abnormalities associated with portal biliopathy: CT and MR findings. AJR Am J Roentgenol. 2006;187(6):W646-W652.

4. Kumar A, Saraswat VA. Portal cavernoma cholangiopathy: current understanding and management. J Clin Exp Hepatol. 2014;4(Suppl 1):S67-S76.
https://doi.org/10.1016/j.jceh.2013.08.011

5. Goyal N, et al. MRCP in portal biliopathy: Spectrum of findings and correlation with clinical presentation. Eur J Radiol. 2008;66(3):505-512.

6. Sarin SK, et al. Portal biliopathy: definition, clinical features, and management. Semin Liver Dis. 2002;22(1):17-25.
https://doi.org/10.1055/s-2002-23206

7. Hulshoff A, et al. Mass-forming portal biliopathy: a diagnostic challenge. Diagnostics (Basel). 2020;10(9):673.
https://doi.org/10.3390/diagnostics10090623

8. Nayak NC, Vasdev N, Saigal S, et al. Portal biliopathy in children: case series and review. J Pediatr Surg. 2013;48(9):E9-E14.

9. Martel M, et al. Portal biliopathy in non-cirrhotic portal hypertension: report of a case and literature review. Clin Imaging. 2015;39(6):1125-1128.

10. Garcia-Tsao G, Sanyal AJ, Grace ND, Carey W. Prevention and management of gastroesophageal varices and variceal hemorrhage in cirrhosis. Hepatology. 2007;46(3):922-938.
https://doi.org/10.1002/hep.21907