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Case Report

Extra-hepatic portal vein obstruction leading to portal cavernoma cholangiopathy without liver dysfunction

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ABSTRACT

In adults, EHPVO mainly occurs following thrombosis, while in children it may be related to congenital malformations and/or neonatal umbilical venous catheterization. EHPVO leads to the cavernous transformation of the portal vein in the absence of recanalization. Portal cholangiopathy (also referred to as portal biliopathy) is common in patients with long-standing chronic PVT. It is due to compression of the large bile ducts by the venous collaterals (portal cavernoma) that form in patients with chronic PVT. Typically, symptoms of portal cholangiopathy include jaundice, biliary colic and pruritus.

Keywords: Extra-hepatic portal vein obstruction, Portal cholangiopathy, Portal cavernoma, Portal vein thrombosis

INTRODUCTION

Portal cavernoma cholangiopathy refers to the biliary tract abnormalities that accompany extrahepatic portal vein obstruction (EHPVO) and subsequent cavernous transformation of the portal vein. EHPVO is a primary vascular disorder of the portal vein in children and adults manifested by longstanding thrombosis of the main portal vein. Nearly all patients with EHPVO have manifestations of portal cavernoma cholangiopathy, such as extrinsic indentation on the bile duct and mild bile duct narrowing, but the majority are asymptomatic. However, progressive portal cavernoma cholangiopathy may lead to severe complications, including secondary biliary cirrhosis.[1]

CASE REPORT

We presente a case report of EHPVO leading to portal cavernoma cholangiopathy. A 35 years old female presented with chief complaint of blood in stools for 1 day. It was associated with dull aching, mild abdominal discomfort localized to upper abdomen. Patient had 3 episodes of malena with foul smelling, tarry coloured stools. There was only one episode of blood in vomitus and patient was immediately brought to our hospital. It was not associated with any yellowish discolouration of eyes or urine. It was not associated with any abdominal distension or any altered sleep pattern. Patient has no past history of jaundice, hepatic encephalopathy or ascites. There was no past history of any chronic illness (diabetes, hypertension, liver disease). Patient was vegetarian by diet, non-alcoholic, non-smoker. There was no specific or relevant history of any drug intake in the past. On examination-patient was calm, conscious and oriented to time place and person; no halitosis, pallor was present, no icterus, no clubbing, no cyanosis, no lymphadenopathy, no edema; no evidence of loss of axillary or pubic hair, no evidence of dupuytren's contracture, flapping tremors were also not present. P/A examination revealed dilated veins over the abdomen, splenomegaly with normal liver span (12 cm), no evidence of ascites. Biochemical profile was as follows: Hb- 5.5 g/dl, RBC- 2.35 millions/cumm, PCV-18.9, MCV-80.4fl, MCH-23.4 pg, MCHC- 29.1 g%, TLC-11000/cumm, DLC- 70/25/02/02/01; LFT- total bilirubin-1.3 mg/dl, direct bilirubin- 0.103 mg/dl, SGOT/AST- 27 U/l, SGPT/ALT- 20 U/l, ALP- 78 U/l, total protein- 7.27 g/dl, serum albumin- 4.23 g/dl, serum globulin- 3.04 g/dl,

PTI- 93.3%, serum lipase- 150 U/l, serum amylase- 104 U/l, HIV- non-reactive, HBsAg- non reactive, HCV- non-reactive, serum ammonia- 47, USG abdomen: liver measures 10.8 cm in size and normal in shape with smooth and regular margins and normal echopattern. Portal vein is 14.3 mm in caliber. Spleen is enlarged measuring 14.5 cm in size and shows normal echopattern. No evidence of free fluid in the peritoneal cavity.

UGI endoscopy was done which showed 2 large and 2 small esophageal varices with RCS. EVL (endoscopic variceal ligation**)** was done and patient improved symptomatically.

MR whole abdomen with contrast findings were suggestive of: mass like cavernoma formation of portal vein with portal cavernoma cholangiopathy alongwith paracholedochal varices; splenomegaly with prominent splenic vein and portsystemic collaterals (Figure 1).

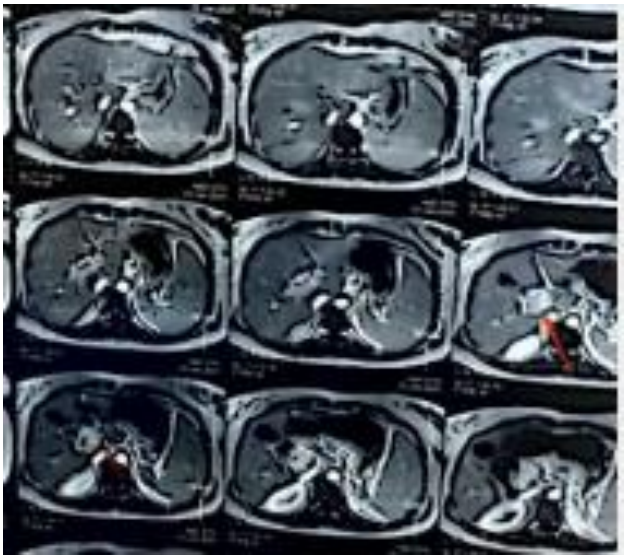

Figure 1: MR abdomen showing portal cavernoma marked with red arrow.

DISCUSSION

The most common cause of PVT is cirrhosis. In a non-cirrhotic liver, PVT is mainly due to inherited or acquired pro-thrombotic states. The clinical picture of PVT varies greatly depending on the acute or chronic presentation. The acute symptoms may include abdominal pain and new-onset ascites. During later stages, the portosystemic shunting develops, leading to esophageal varices, rectal bleeding, and hematemesis along with signs of portal hypertension. Long-standing PVT (or EHPVO) may lead to the development of alternative dilatation of multiple venous channels around the thrombosed PV, thus bypassing the occlusion. This is known as cavernous transformation of the portal vein or portal cavernoma.[2] In our case report patient presented with signs and symptoms of portal hypertension secondary to EHPVO (cause undetermined). EHPVO can present as early as 6 weeks after birth as well as manifest in adulthood. Clinical presentation depends on recent or chronic onset of clinical disease and age of presentation. The most common clinical features are: hematemesis and malena. It is usually not associated with hepatocellular dysfunction. Gastrointestinal bleed is usually recurrent before a patient seeks medical attention. Anemia and splenomegaly are other common features of EHPVO. Ascites can present transiently in 10%-20% of patients and is seen more frequently in adult patients with long-standing disease and declining liver function.[3] On investigations it was revealed that patient was having oesophageal varices (UGI endoscopy) due to portal hypertension with evidence of cavernoma formation leading to portal cholangiopathy. As patient had normal liver function it was proposed that although cholangiopathy has developed but patient is asymptomatic. Most common symptom seen in portal cholangiopathy is jaundice. Jaundice may result from bile duct compression because of dilated venous collaterals. Portal cavernoma cholangiopathy (PCC) is recognized in 90%-100% of the cases; however, only a few patients are symptomatic, usually in the adult age groups which is indicative of advance disease. Pathogenesis of biliary changes in PCC is due to two components: (a) a reversible component, due to biliary compression by engorged collaterals, that reverses after shunt surgery; and (b) a fixed component that does not reverse after shunt surgery and is likely due to ischemic changes in the bile duct, occurring either at the time of portal vein thrombosis or due to prolonged compression by collaterals.[4] The clinical characteristics of patients with PCC are divided into asymptomatic and symptomatic phases. Patients in asymptomatic phase are detected to have the presence of biliary system abnormalities on imaging (MRC or ERC) in the absence of any biliary symptoms. Symptomatic patients on the other hand present with features of chronic cholestasis with or without biliary pain or acute cholangitis related most often to the presence of biliary strictures or stones. Examination of patients with symptomatic PCC usually reveals the presence of jaundice, an enlarged spleen in the majority and hepatomegaly in half to 2/3rd of patients. In addition, most of the patients with EHPVO with PCC have history of variceal bleed as the manifestation of primary disease. Patients with EHPVO usually have a long standing disease lasting for 8-10 years before they present with symptomatic PCC.[5]

We were able to diagnose a case of portal cholangiopathy primarily on the basis of clinical picture as patient was having symptoms (hematemesis, malena) probably due to portal hypertension, with splenomegaly and absence of any evidence of cirrhosis of liver, or ascites. Cavernoma formation had occurred which was impinging upon the biliary system but there was no clinical symptom of biliopathy. Conventionally, medical and endoscopic management is usually recommended for EHPVO. For primary prophylaxis of variceal bleeding, there is insufficient data on whether beta-blocker or endoscopic therapy should be preferred. For control of acute variceal bleed, endoscopic therapy is effective. Surgical shunts are

used for refractory or complicated cases, surgery is primarily indicated when endotherapy fails to control bleeding or delayed sequelae such as portal biliopathy and rectal varices occur. Surgical portosystemic shunting is contraindicated in patients with portopulmonary hypertension or hepatic encephalopathy. Liver transplantation should be considered in these cases.[6]

CONCLUSION

EHPVO is diagnosed when patient has massive splenomegaly with repeated episodes of malena and hematemesis although patient doesn't go into hepatic decompensation. It frequently leads to development of portal cavernoma with associated complications.

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REFERENCES

1. Wani ZA, Bhat RA, Bhadoria AS, Maiwall R. Extrahepatic portal vein obstruction and portal vein thrombosis in special situations: Need for a new classification. Saudi J Gastroenterol. 2015;21(3):129-38.
2. Samant H, Asafo-Agyei KO, Garfield K. Portal vein thrombosis. StatPearls. 2021.
3. Chawla Y, Dilawari JB, Katariya S. Gallbladder varices in portal vein thrombosis. AJR Am J Roentgenol. 1994;162(3):643-5.
4. Khuroo MS, Yattoo GN, Zargar SA, Javid G, Dar MY, Khan BA, Boda MI. Biliary abnormalities associated with extrahepatic portal venous obstruction. Hepatology. 1993;17(5):807-13.
5. Dhiman RK, Behera A, Chawla YK, Dilawari JB, Suri S. Portal hypertensive biliopathy. Gut. 2007;56(7):1001-8.
6. Khanna R, Sarin SK. Non-cirrhotic portal hypertension–Diagnosis and management. J Hepatol. 2014;60:421-41.

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