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Staged Management of Portal Biliopathy - Case Report

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Abstract: *Portal biliopathy (PBP) occurs as a consequence of portal hypertension. Patients with extrahepatic portal hypertension (EHPVO) are more prone to develop PBP due to the longstanding nature of disease which occurs in childhood. Here we present a 18 years old male with EHPVO presenting with jaundice. He was managed in a staged manner with proximal splenorenal shunt (PSRS) as first stage procedure and biliary drainage as a second procedure for the biliary obstruction .*

Keywords*: portal biliopathy ; extrahepatic portal vein obstruction; portal hypertension; Biliary obstruction; secondary biliary cirrhosis; proximal splenorenal shunt*

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I. Introduction

Portal biliopathy denotes the changes in the biliary tree both intra and extrahepatic and gallbladder due to portal hypertension. The changes associated with PBP are common in portal hypertension as a result of EHPVO, which accounts for up to 40% of cases of portal hypertension in India [1]. Although all patients with EHPVO develop portal biliopathy, symptomatic portal biliopathy is seen in 5-33% of cases [2,3].Management of symptomatic cases is essential as they progress to complications like biliary obstruction and secondary biliary cirrhosis if left untreated.

II. Case Report

18 years old boy presented to us with jaundice for 2 months duration. He also had recent episode of hemetemesis and melena. On examination he had stunted growth and was pale and icteric. Spleen was enlarged and was palpable 6cm below left costal margin. Blood investigation showed his haemoglobin-5.5 gms %, total count - 6500 Cells/Cu.mm and platelet count -1.2 lakhs /Cu.mm . His total bilirubin was 11.5 mgs/dL and direct bilirubin was 6 mgs /dL. Bone marrow aspiration showed hypercellular marrow. Fibroscan done showed score of 5.2Kpa.

Upper GI endoscopy revealed Grade 2 oesophageal varices 2 columns and portal hypertensive gastropathy. Doppler ultrasound showed cavernous transformation of main portal vein and splenic vein diameter of 13.3mm . MRI abdomen with MRCP showed normal sized liver with bilateral IHBR dilatation. Splenomegaly with splenorenal collaterals was also noted. There was a stricture at the common hepatic duct.

ERCP showed no flow of contrast beyond common hepatic duct. During the evaluation he developed cholangitis and a percutaneous transhepatic biliary drainage (PTBD) of the left system was done. After the patient recovered from cholangitis proximal splenorenal shunt was done .

Since the patient had persistent jaundice after PSRS, six weeks following shunt surgery, Roux-en-Y hepaticojejunostomy was done end to side fashion. The patient improved symptomatically and PTBD output reduced which was removed after confirmation with check cholangiogram which showed free flow of contrast through the hepaticojejunostomy.

III. Discussion

Portal biliopathy is reported in 80–100% cases of EHPVO[4]. The epi-choledochal and the para-choledochal plexus which runs parallel to biliary tract are responsible for the obstructive symptoms . The inflammatory process gives rise to fibrosis of bile duct, which leads to stricture formation and eventually to secondary biliary cirrhosis.

Although Portal biliopathy has been observed mostly in EHPVO patients, such changes have been reported in a milder form in patients with cirrhosis, non-cirrhotic portal fibrosis, and congenital hepatic fibrosis. It is not clear for the high frequency of PBP in EHPVO patients .

Abnormal LFTs can be present without overt jaundice or cholangitis. The subset of patients with asymptomatic PBP and abnormal LFTs is an area of concern of whether abnormal LFTs alone should represent an indication for surgery to protect the liver from the effects of prolonged subclinical biliary obstruction.

Magnetic resonance cholangiography has replaced ERCP as investigation of choice to diagnose several biliary tract abnormalities. [5]. Endoscopic management has been reported to be successful [6]. But the drawback is endoscopic management does not address the underlying etiology and is not successful in long term.

Portosystemic shunt surgery will decompress the choledochal collaterals and relieve the jaundice in most of the patients. Proximal splenorenal shunt with splenectomy is the preferred procedure.

One third of patients will have persistent biliary obstruction even after shunt surgery[7]. This is because the obstruction was not due to extraneous compression by the choledochal collaterals but is because of the stricture of the bile duct. In such cases biliary drainage will bypass the obstructed segment.

Few reports of drainage of biliary system by segment 3 bypass is available as definitive procedure without PSRS. This procedure can be done if the abnormalities in the biliary tree is confined to extrahepatic biliary system alone[8].

IV. Conclusion

Portal biliopathy is more common in patients with EHPVO. Regular follow-up in patients with EHPVO can identify these patients early and can be helpful in early treatment and better outcomes. Shunt procedure followed by biliary decompression is the procedure of choice for patients with portal biliopathy.

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