Case Report

Extra Hepatic Portal Vein Obstruction leading to variceal bleed due to Portal Hypertension

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Abstract

Portal Hypertension can be due to many causes other than cirrhosis. We report a case of extra hepatic portal vein obstruction leading to portal hypertension and varices, managed successfully by creating a Porto Caval shunt.

Introduction

Portal hypertension is defined by a hepatic venous pressure gradient (HVPG) >5mmHg. It may be due to increased resistance to flow because of extra hepatic portal vein obstruction, a pre-hepatic cause or others like (hepatic and post-hepatic) and increased portal blood flow. Portal hypertension results in the development of collaterals to bypass the increased resistance to flow within the portal bed and to return blood to the systemic circulation. Esophageal varices, the most important collaterals, form and bleed only if HVPG >12 mmHg.[1]

Gastrointestinal bleeding is the most severe complication of portal hypertension and esophageal and gastric varices are by far the most common sources of bleeding in these patients. After an initial variceal hemorrhage,

the frequency of recurrent bleeding ranges from 30% to 40% within the subsequent 6 weeks.[2]

Therefore, the general consensus is that all patients who survive an episode of variceal bleeding must receive some effective form of treatment to prevent rebleeding. The currently available therapeutic options include pharmacotherapy, endoscopic therapy, radiologic transjugular intrahepatic portosystemic shunt (TIPS), surgical shunt, and liver transplantation.

Hence, the primary objective of drug, radiologic, and surgical therapy for portal hypertension is the reduction of HVPG, the target value being 12 mmHg.[1] TIPS and surgical shunts achieve this target by creating a low-resistance channel between the hypertensive portal system and the inferior vena cava system. However, the liver is deprived of the portal flow and portosystemic encephalopathy and liver failure can ensue.

Though surgical shunting is associated with higher degree of encephalopathy but decreases rebleeding considerably. On the other hand TIPS also significantly decrease re-bleeding incidence but is associated with other complications, especially stent dysfunction and is not as effective as surgery in lowering the chances of re-bleeding.[3]

We report a case of recurrent variceal bleeding due to extra hepatic portal vein obstruction managed successfully by surgically creating a low resistance channel, a Porto Caval shunt using a Poly Tetra Fluoro Ethylene H type graft (PTFEH).

Case Report

A 13-year-old boy presented with 3 episodes of massive hematemesis together with epistaxis in the last week. Patient was complaining of hematemesis 3-4 times every year for the last six years, for which he was transfused 35 units of blood but was not investigated for the cause of hematemesis.

On examination, he was vitally stable, with splenomegaly 3" below the costal margin. Collaterals were present around the umbilicus. Upper GI endoscopy revealed grade III esophageal and gastric varices. Ultra sound doppler showed a blunted portal vein at porta hepatis measuring 9mm in size, thin collateral vessels originating from the blunted portal vein with portal pressure 32mmHg.

Sub capsular wedge biopsy done intra-operatively showed preserved lobular architecture which could not explain the cause of portal hypertension.

Blood picture, coagulation profile, liver function tests and all the other investigations were within normal limits.

The patient was diagnosed as a case of extra hepatic portal vein obstruction leading to portal hypertension and was operated for the same. Operative findings were consistent with portal vein thrombosis. There was a bunch of dilated veins simulating a haemangioma, which made mobilization of the portal vein difficult and hazardous. Therefore, we did a Porto-caval anastamosis using PTFEH graft (Figure). A good flow was observed and patient made an uneventful recovery with portal pressure declining to normal, from 32mm of saline to 10mmHg.

Figure. Showing the anastomosis between Portal Vein and Inferior Vena Cava usinga PTFEH graft

The patient was followed for one year post-operatively. There were no episodes of hematemesis or epistaxis.

Discussion

Portal hypertension is characterized by a pathologic increase in portal venous pressure that leads to the formation of an extensive network of porto systemic collaterals that divert a large fraction of portal blood to the systemic circulation, bypassing the liver.

The occurrence of extra hepatic portal vein thrombosis (EPVT), as seen in the presented case, can be influenced by both local and systemic etiological factors. Local factors comprise of disorders leading to decreased portal flow such as liver cirrhosis and hepatobiliary malignancies.[4,5] Systemic risk factors for EPVT consist mainly of acquired and inherited abnormalities leading to hypercoagulability.[6]

The clinical outcome of EPVT may be associated with these concomitant medical conditions or with manifestations of portal hypertension. Due to the rarity of the condition little is known about the determinants of survival and causes of death of patients with EPVT.[7]

Ultrasound (US) techniques such as duplex US or spectral Doppler imaging and CDI or power Doppler imaging are the modalities of choice in the evaluation of

imaging are the modalities of choice in the evaluation of the liver and portal hypertension (PH). These techniques are noninvasive, rapid, and highly sensitive and specific. Ultrasound in our case revealed portal vein obstruction as well but still the endoscopy was not performed to rule out the presence of bleeding varices.

Angiographic techniques such as SP, transhepatic portography, transumbilical catheterization, transjugular catheterization, wedge hepatic venography, and arterial portography are invasive. However, they are much more specific examinations for evaluation of PH and they are indicated when definitive surgery or radiologic intervention is contemplated.

Endoscopic diagnosis can be difficult when the view is obscured by blood. Nevertheless, a diagnosis of variceal hemorrhage is acceptable when a venous spurt is seen or there is fresh blood in the lower esophagus in the presence of varices. In about half of the cases there is no active bleeding; variceal haemorrhage is indicated by the presence of a "white nipple sign" (plug of platelet fibrin on a varix) or when varices are the only lesion identified.[8,9]

Available treatments of portal hypertension have been limited to systemic agents such as the beta-blockers; mechanical therapies such as endoscopic sclerotherapy / band ligation, transjugular intrahepatic portosystemic shunt (TIPS), surgical shunting, and liver transplantation.

Surgical therapy for this disease aims to prevent or control esophageal variceal hemorrhage. Liver transplantation is regarded as an effective procedure to cope with complications caused by portal hypertension. This procedure is expensive and only a few patients can afford it. Differences in etiology, major clinical symptoms, liver function and systemic status are assumed that there is no fixed approach to follow. Hence individualized therapeutic protocol is feasible for the patient on the basis of current knowledge about the mechanism of portal hypertension.[10,11] As our patient was fulfilling the criteria of child Pugh classification as child "A"[12] we opted for Porto caval shunting using the PTFEH graft over sclerotherapy or banding, where we wanted the patient to get a permanent cure of his disease rather than just preventing re-bleed.

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