Original Article

PROFILE OF EXTRAHEPATIC PORTAL VENOUS OBSTRUCTION (EHPVO) IN A TERTIARY CARE HOSPITAL IN PAKISTAN

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ABSTRACT

Objective: To study the clinical and laboratory profile of patients with EHPVO in a tertiary care hospital of Pakistan and to differentiate EHPVO from cirrhosis of liver and to see the effect on liver function tests.

Methodology: This is a prospective observational study conducted at Department of Gastroenterology, Pakistan Institute of Medical Sciences, Islamabad.Twenty five patients of 12–55 years of age with the features of portal hypertension were included in this study. After careful history and physical examination patients were subjected for laboratory investigations including liver function test, renal function test, blood CP, PT, APTT, HbsAg and anti HCV, other specialized procedures including endoscopy, liver biopsy and ultra sound was also done in all patients.

Results: Portal vein thrombosis was the predominant cause of EHPVO, accounting for 88% of cases. All patients were presented with upper GI bleeding, splenomegaly was observed in 88% of patients. None of the patients had clinical, biochemical or liver biopsy evidence of chronic liver disease.

Conclusion: The diagnosis of extra hepatic portal venous obstruction and differentiation from cirrhosis can be easily made by characteristic clinical features, normal liver function tests and doppler ultrasound. Portal vein thrombosis (PVT) is the predominant cause of EHPVO in Pakistani patients, as seen at this tertiary care hospital in Pakistan.

KEY WORDS: EHPVO, Cirrhosis, Thrombosis.

Conflict of interest: *None.*

Pak J Med Sci October - December 2007 (Part-I) Vol. 23 No. 5 677-680

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* Received for Publication: February 19, 2007

* Accepted: July 2, 2007

INTRODUCTION

Extrahepatic Portal Vein Obstruction (EHPVO) is an important cause of noncirrhotic portal hypertension, especially in Third World countries. The etiology and clinical presentation are different in children and adults.[1] Mechanical obstruction to portal venous flow occurs as a result of partial or complete obstruction of the main portal vein or splenic vein. In the absence of cirrhosis of liver it is called extra hepatic portal venous obstruc-tion, a form of presinusoidal hypertension. About 20% of patients presenting with portal hypertension have pre hepatic pathology.[2] The aetiology of extra hepatic portal vein obstruction in most cases is unknown.[3]

The causes of prehepatic portal hypertension are usually due to portal vein thrombosis following umbilical sepsis, intraluminal trauma during exchange transfusion and pylephlebitis following intra abdominal sepsis in childhood age group.[4]

In adults and older age group extra hepatic portal vein obstruction is relatively rare and the causes are myeloprolifrative disor-ders, deficiencies of protein C and S and anti thrombin III leading to hypercoagulable state and portal vein thrombosis. Other rare causes in older age groups are tumours in liver, bile ducts, and pancreas.[5]

Patients with extra haptic portal venous obstruction present mainly with recurrent episodes of variceal bleeding. The remainder present with asymptomatic spleno megaly or features of hypersplenism. Ascites is a rare finding in young children. The diagnosis of extra hepatic portal venous obstruction (EHPVO) is easily made by characteristic clinical features, normal liver function tests and ultra sonography or spleno porto venography. Definitive diagnosis of portal vein thrombosis is made by selective angiography of the celiac axis, superior mesenteric artery and splenic vein.

Endoscopy will define the severity and bleeding risk of varices. Hematologic assessment should focus on hyper coagulable states particularly in patients with no other evident risk factors. Management of extra hepatic portal venous obstruction (EHPVO) is two fold. One is management of bleeding varices by either vasopressin (Pitressin) or somatostatin, sengstaken esophageal compression, endoscopic sclerotherapy or porto systemic shunts. Surgery can also be utilized in emergency state, subsequent management includes planned surgical procedures. The second choice is the management of splenomegaly and hypersplenism by either distal spleno renal shunt (DSRS) or splenectomy. Extra hepatic portal venous obstruction (EHPVO) is the major cause of portal hypertension and bleeding varices second to cirrhosis of liver (secondary to viral hepatitis) in Pakistan. The natural history and prognosis of the disease is variable. Rupture of esophageal varices is the most serious complication of portal hypertension and is associated with significant mortality.

Patients with EHPVO presents mainly with recurrent episodes of massive haematemesis, on examination they have large spleens but the liver is of normal size and consistency, ascites is a rare finding. The diagnosis of EHPVO is easily made by characteristic clinical features, normal liver function tests, histology and by duplex ultrasound of portal venous system. High incidence of EHPVO, with all the complications of the disease and its affects on patients and their families, led us to study the cases of EHPVO in our indigenous population and to analyze their causes. The study also intended to clinically differentiate EHPVO from cirrhosis of liver and to see the effect of EHPVO on liver function tests.

METHODOLOGY

This was an observational study conducted in the department of gastroenterology, Pakistan Institute of Medical Sciences, Islamabad from January 1995 to September 1996.

Patients of all ages with the features of portal hypertension were included in this study. After careful history and physical examination patients were subjected for laboratory investigations including liver function test, renal function test, blood CP, PT, APTT, HbsAg and anti HCV,[6] other specialized procedures including endoscopy, liver biopsy and ultra sound was also done in all patients while proctoscopy was carried out in patients with fresh rectal bleeding.

RESULTS

Twenty five patients were enrolled with age range between 12 to 55 years, mean age was 20 years, majority of the patients were less then 20 years of age, males were predominant 15 as compared to 10 females.

Main clinical presentation in all patients were upper GI bleeding as hematemesis and melaena as well, splenomegaly was observed in 88% of patients, whereas hypersplenism

was observed in 28% patients accompanied with pancytopenia. Moderate ascites was observed in two patients secondary to advanced metastatic renal cell carcinoma and hypoalbuminemia, 76% patients had hypochromic microcytic anemia with mean hemoglobin 6gram/dl. Liver size was normal in all but two patients, secondary to metastatic renal cell carcinoma and accessory right lobe, whereas liver function tests were normal in 92% patients. PT, APTT and renal function tests were also normal in all patients

Endoscopy revealed that 90% patients had Grade III varices while proctoscopy revealed 24% patients had rectal varices. Duplex ultrasound showed that almost all patients had dilatation of major portal channels, portal vein measuring greater than 1.5cm in diameter, 88% patients showed the evidence of portal vein thrombosis, out of these 50% patients showed cavernous transformation of portal vein with hepato-petal portal flow.

Evidence of cirrhosis was not found in any patient with EHPVO, 80% patients had normal liver histology while 16% showed evidence of portal fibrosis, one patient had infiltration of metastatic renal cell carcinoma. Two patients were positive for HBsAg and one for Anti HCV.

DISCUSSION

Portal hypertension caused by extra hepatic obstruc-tion occurs when the site of block in the portal vein before the blood reaches the liver. In western countries such patients comprise 5-10 % of all cases of portal hypertension but in developing countries this percentage is much higher.[7] EHPVO is the commonest cause of major upper Gastro intestinal bleeding in childhood.[8,9]

Patient with EHPVO are usually young mostly between 10-20 years of age and in good general health. The disease generally does not lead to severe malnutrition and there are no chronic liver disease stigmata therefore oedema, ascites, gynecomastia, spiders naevi or porto systemic encephalopathy (PSE) are very rarely observed. The most important clinical feature is splenomegaly, the spleen is enlarged and painless, splenomegaly was observed in 88% of our patients, the reason for this high percentage is due to late presentation (after 2-3 years) of patients to hospital after first bleeding and secondly malaria is endemic in Pakistan.

Hypersplenism is one of the most common finding in patients with EHPVO, patients with 50,000/- platelet are frequently observed. Platelet function such as adherence and aggregation is however normal, this is the reason that symptomatic hypersplenism in the form of gingival bleeding, nose bleeding, ecchymoses and hematoma are rarely observed. However leucopenia was common finding in our patients.

Anaemia was also observed in majority of our patients with haemoglobin ranged between 5-9gm/dl.[6] Such a high percentage of anaemia was possiblly due to frequent variceal bleeds before presentation to a hospital and associated worm infestation which is common in our rural areas. A large proportion of patients presents with episodes of upper GI bleeding in the form of haematemesis and melaena.[10] Due to good liver functions in EHPVO, patients withstand the hemorrhage well and do not have mental confusion or other clinical sings of porto systemic encephalopathy. When the bleeding is more severe mild ascites may be observed for a transient period. Contrary to patients with cirrhosis there is no hepato renal syndrome. The liver is normal in size and consistency, stigmatas of liver disease such as jaundice or vascular spiders are absent.

From laboratory stand point the disease is characterized by normal liver function tests with no laboratory evidence of hepato cellular damage. However portal block of long duration may result in liver atrophy with deranged liver function test.[11] In addition to normal liver function tests confirmation of normal liver is best obtained by liver biopsy or indirectly by measurement of Wedge hepatic vein pressure (normal 3-12mm Hg). The findings of an elevated splenic pulp pressure (normal 8-12mm Hg) and demonstra-tion of block by simultaneous splenic portography confirm the diagnosis of extra hepatic portal hypertension.

Sonography identified the cause of portal hypertension either intra hepatic (cirrhosis) or extra hepatic. In cirrhosis the liver may be enlarged, normal in size or shrunken and lobulated when end stage disease is present. Coarse parenchymal echoes are identified in 65% while remaining 35% have a normal echo pattern.[12] In the presence of portal hypertension, if the liver appears sonographically normal, one should look for extra hepatic obstruc-tion of the main portal or splenic vein. Using duplex doppler ultrasound the rate and direction of portal blood flow can be measured.

Portal vein thrombosis is known to be a silent cause of portal hypertension in infants whilst often not presenting clinically until young adulthood, it has recently been recognized to be associat-ed with other conditions, these includes chronic pancreatitis with inflammatory involvement of splenic vein or malignant infiltration secondary to carcinoma of the pancreas or hepatoma. These patients had umbilical sepsis or septicaemia in their neonatal life or childhood. So the incident of portal hypertension can be decreased by educating the peoples about prevention of neonatal and childhood sepsis by improving sanitation.

In a young child with splenomegaly and upper GI bleeding and normal liver function the diagnosis of EHPVO is nearly certain. It could be confirmed with the help of ultrasonography, doppler studies and upper GI endoscopy, rarely other investigations like spleno porto venography, superior mesenteric angiography (venous phase), CT abdomen or liver biopsy are needed to confirm the diagnosis. In adults a search for the primary cause should be under taken. The prognosis of EHPVO depends upon the site of block, immediate resuscitation after bleeding, the effectiveness of sclerotherapy and the availability of suitable vessels for shunt surgery.

CONCLUSION

Portal vein thrombosis (PVT) is the major causes of EHPVO. The diagnosis of EHPVO is almost certain in young patient with splenomegaly and upper GI bleeding in the absence of stigmatas of chronic liver disease. Distinction from cirrhosis of liver can be easily made by the absence of peripheral stigmatas (spiders, gynecomastia, levconyc-hia palmar erythema) of chronic liver disease, normal liver function tests and histology.

ACKNOWLEDGEMENT

We are indebted to Clinision for their assistance in drafting the final manuscript. We are also grateful to Dr. Rizwan Qazi and Dr. Qurat-ul-Ain Haider for their valuable comments on the earlier draft of the manuscript.

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