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CASE REPORT

Portal Hypertension

by

INDA D. ARIF, MARDJANIS SAID and E.M. HALIMUN

(Departments of Child Health and Surgery, Medical School University of Indonesia, Jakarta)

Abstract

A case of portal hypertension in a 9-year-old Indonesian female child is presented. The diagnosis was made by demonstrating oesophageal varices radiologically and surgically, and by measuring the portal venous pressure. Portocaval side to side shunt was done resulting in diminishing of the oesophageal varices, disappearance of hypersplenism, and a normal portal venous pressure. Continuous observation is recommended despite the relative good prognosis.

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Introduction

Obstruction of the flow of blood at any site from the splanchnic venous system along the course of the portal blood flow through the liver to the heart produces portal hypertension (Garabedian and Whitcomb, 1971). The obstruction may occur in the portal vein itself or in the liver. Cirrhosis of the liver is the most common etiology in the adult, while thrombosis of the portal vein is a more frequent cause of portal hypertension in the child (Foster et al., 1963).

Portal hypertension may be divided into (Whipple, 1945):

1. Extrahepatic portal hypertension
   — pre-hepatic block
   — post-hepatic block

   The prehepatic block is often caused by thrombosis of the portal vein as a result of omphalitis in the neonatal period (Ehrlich et al., 1974), peritonitis in the neonatal period (Trusler et al., 1962), or thrombosis of the umbilical vein due to catheterization in the neonatal period (Trusler et al., 1962; Shaldon and Sherlock, 1962). The physiological obliteration of the umbilical vein and ductus venosus rarely causes a prehepatic block. A block in the hepatic vein is an example of posthepatic block. Myers and Robinson (1973), and Patton (1963) reported that cavernomatous transformation is a frequent cause of extrahepatic portal hypertension.
2. Intrahepatic portal hypertension :
   The intrahepatic portal hypertension may be due to liver cirrhosis, malignant hepatoma, and intrahepatic venous thrombosis secondary to hamartoma.

This paper is a report of a case of portal hypertension in a 9-year-old female child, on whom the authors had the opportunity to diagnose and treat.

Case report

S., a 9-year-old Indonesian female child, was admitted for the first time to the Department of Child Health, Dr. Cipto Mangunkusumo Hospital Jakarta, on April 17, 1975, with a diagnosis of portal hypertension. The history revealed that 4 days prior to admission she suffered from hematemesis followed by melaena; since then she looked very pale and became weak. Six days before, she had cough and a running nose. There was neither fever nor history of epistaxis, gum bleeding, nor subcutaneous bleeding. At the age of two she developed jaundice. The neonatal history was uneventful. Physical examination on admission revealed a 9-year-old Indonesian female child with a body weight of 23 kg. She was alert, in a sufficient nutritional state, looked rather pale, and weak. Cyanosis, jaundice, and dyspnoea were absent. There were no enlargements of the lymphnodes. The body temperature was 37.4° C. The heart and lungs were normal. The abdomen was supple with slight ascites but venectasy was absent; the liver was just palpable, the spleen was enlarged (S III). Slight edema was present at both legs, and there were ecchymoses, petechiae, and hematomas.

Laboratory findings

Hb.: 4.8 gm.%; RBC: 1.16 mill./mm^3; reticulocytes: 49‰; WBC: 4,600/mm^3; platelets: 154,000/m^3; differential count: 0/-/2/63/31/4; the blood smear revealed poikilocytosis, hypochromia, and the presence of target cells.

Bleeding time: 3.30 min.; clotting time: 4 min.; recalcification time: 117 sec; prothrombin time : 26 sec.; PTT : 42 sec.; TGT: normal; clot retraction: 60%.

Blood analysis

Albumin: 3.41 gm.%; globulin: 2.63 gm.%; ureum: 24 mg.%; creatinine: 1.18 mg.%; cholesterol: 114 mg.%; direct bilirubin: (—); indirect bilirubin: 0.8 mg.%; alkaline phosphatase: 10.8 K.A.U; TTT: 2.7 Kingburg U.; Kunkel: 10.9 U.; SGOT: 14,544 mU/ml.; SGPT: 9,999 mU/ml.; BSP retention test: 3% after 45 minutes.

No abnormalities were detected in the urine and stool. Bone marrow smear revealed a hyperactive erythropoietic system. Histologic examination (P.A. No. 753489) of the liver biopsy specimens revealed a non-specific hepatic reaction and hydropic degeneration. Post-necrotic scarring was also observed. The possibility of post-necrotic cirrhosis could not be excluded, if these changes are found throughout the whole liver.

On the 10th day of hospitalization she developed hematemesis and melaena. Oesophagogram showed prominent oesophageal varices at a distal portion of the oesophagus. The gastric mucus layer was normal. The I.V.P. was normal. Splenoportogram revealed a complete filling of the contrast in the portal vein up to the hilus of the liver. There was neither contrast seen in the liver structure nor cavernomatous transformation. The portal venous pressure was 280 mm. H_2O Three days after complete examinations she underwent portocaval (side to side) operation. On exploration no thrombosis in the portal vein as well as in the splenic vein was found; large varices were seen around the upper part of the duodenum. Preoperatively ampicillin and blood transfusions were administered. Fultrexin was given orally and also added in the laxative agent. Ampicillin was continued for 10 days postoperatively.

Postoperative observation was uneventful as seen from the following results:

	Pre-operative	Post-operative
Hb. (gm.%)	3.5 — 6.4	10 — 10.5
R.B.C. (mill./mm^3)	1.6 — 2.2	3.5 — 4.5
W.B.C. (per mm^3)	3,500 — 5,000	5,800 — 9,800
Platelets (per mm^3)	75,000 — 154,000	240,000 — 270.000
Spleen	S III	S I
Ascites	+	—
Edema	+	—
Portal venous pressure (mm H_2O)	280	165
Liver function tests	normal	normal

She was discharged 12 days postoperatively in a good condition. At the last visit 60 days thereafter she did not look pale, the heart and lungs were normal; ascites and edema were not found, neither were subcutaneous bleeding. The liver and spleen were not palpable. Hb.: 10 gm.%; WBC: 9,800/mm^3; platelets: 270,000/mm^3. Portal venous pressure : 165 mm H_2O. The oesophagogram showed diminishing varices.

Discussion

The diagnosis of portal hypertension in this case was based upon the following findings: enlarged spleen with consequently symptoms of hypersplenism, oesophageal varices (radiologically and surgically), splenoportogram, and the increase of the portal venous pressure. Kaye (1969) and Foster et al. (1963) found that liver function tests were still normal in cases of extrahepatic portal hypertension. The first symptoms in the first years of life were ascites and hepatomegaly (Ehrlich et al., 1974). At the older age the main symptom is bleeding from the gastrointestinal tract as a result of damage of the oesophageal varices, whereas in extrahepatic portal hypertension this occurs in the first years of life (Trusler et al., 1962). Besides hemorrhoid and caput medusae on the abdominal wall, pancytopenia and hypersplenism are the most frequent findings.

In our case the first symptoms appeared at the age of nine. Ehrlich et al. (1974) reported that gastrointestinal bleeding was not prominent until the age of six. The presence of oesophageal varices was demonstrated in the oesophagogram and proved during the operation.

Though there were recurrent bleedings, hypersplenism, ascites, and edema, the liver function tests were still normal. Trusler et al. (1962) and Foster et al. (1963) found that the liver function tests were normal in cases of liver cirrhosis in children, whereas in adults these were always impaired. Furthermore, the diagnosis of portal hypertension can be established by the demonstration of oesophageal varices by means of oesophagography or oesophagoscopy and must be confirmed by measuring the portal venous pressure percutaneously or surgically. In our case the diagnosis was intrahepatic portal hypertension based upon the findings as mentioned above.

Portal hypertension due to liver cirrhosis has usually a bad prognosis, particularly in adults (Trusler et al., 1962; Foster et al., 1963). The treatment of portal hypertension consists of conservative and surgical management. The conservative treatment includes bedrest, sedation, blood transfusions, and pitressin injections. If these are not successful, surgical intervention has to be considered. The definitive correction is to diminish the portal venous pressure by shunting the portal to the systemic circulation. This can be done if the diameter of the vein (portal, mesenterial, or splenic) is more than 1 cm. which can be demonstrated by splenoportography. If the condition is poor and the diameter of the veins is not large enough for shunting, bleeding can be stopped by e.g. ligation of the vein, implantation of Boerema button, or gastic transection. In our case recurrent bleedings can be overcome by conservative treatment. Shunting was made electively as suggested by other authors, since there were recurrent bleedings, hypersplenism, and normal liver function, while the diameter of the veins is large enough (Trusler et al., 1962; Shaldon and Sherlock, 1962; Ehrlich et al., 1974).

Portocaval shunt side to side was done since this procedure was the ideal one for our case and the possibility of rebleeding was low (Patton, 1963), although we were still aware of the possibility of encephalopathy postoperatively. Splenectomy was not necessary since by decreasing the pressure of the splenic vein hypersplenism can be abolished; and if the shunt is blocked later on, splenorenal shunt can still be performed in the forthcoming years. Within a 2-month period postoperatively, hypersplenism disappeared and rebleeding did not occur. The portal venous pressure dropped to normal. Since the liver function tests are still normal, we may expect that the prognosis will be relatively good. Anyhow, a close lifelong observation is recommended.

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