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Case Report

A rare case of esophageal variceal bleeding from portal hypertension due to extrahepatic portal vein obstruction in a two-year-old

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ABSTRACT

Extrahepatic portal vein obstruction (EHPVO) is a type of liver vascular disease characterized by cavernomatous transformation and obstruction in the portal veins. Congenital defects, numerous transfusions, trauma, sepsis, dehydration, and hypercoagulable disorders are important risk factors. Since most patients have no symptoms, acute extrahepatic portal vein obstruction is frequently disregarded. Hematemesis and splenomegaly without hepatic decompensation are among the symptoms that might occur in subacute and chronic stages. Imaging studies aid in the diagnosis; Doppler imaging is added to ultrasonography to visualize portal vein blood flow. MRI and CT scans are used to visualize portal vein blockage. Prevention of acute bleeding is the cornerstone in the management. Studies have shown that transhepatic thrombolysis is the preferred choice to avoid systemic side effects. Treatment for extrahepatic portal venous thrombosis involves an intrahepatic portosystemic shunt, which is usually followed by conservative measures to stop variceal haemorrhage. When other forms of therapy are ineffective, liver transplantation is carried out. This case report describes a rare instance of EHPVO in a two-year-old boy who was hospitalized due to recurrent fever, increased belly circumference, and a persistent feeling of fullness in the abdomen. The patient had a surgically placed splenorenal shunt, which helped him respond well to treatment.

Keywords: Extra hepatic portal vein obstruction, Splenomegaly, Splenorenal shunt, Esophageal varices

INTRODUCTION

Extrahepatic portal vein obstruction (EHPVO) is a form of vascular disease in the liver that leads to structural changes in the portal veins, including cavernomatous transformation and obstruction. It may or may not involve the intrahepatic portal veins. The Asian population is prone to EHPVO. It is the primary cause of hematemesis in the pediatric population as a result of esophageal varices and a frequent cause of portal hypertension in children. It is not the same as occlusion of the portal vein brought on by neoplasms, chronic liver disease, or difficulties following surgery.[1] Congenital defects, dehydration, infection, trauma, hypercoagulable conditions, and repeated transfusions are among the risk factors. Splenomegaly and painless hematemesis without hepatic decompensation are the clinical presentation.[2] Liver function tests performed in a laboratory setting reveal a moderate increase of liver enzymes when cirrhosis or chronic liver disease are not present. Patients are frequently asymptomatic throughout the acute period, making it easy to miss EHPVO. Clinical signs and symptoms are usually the result of subacute and chronic stages of the illness. Imaging investigations are useful resources for making a diagnosis. Doppler imaging is added to ultrasonography as the initial imaging modality to see the portal vein's blood flow. The following measures to further visualize the blockage of the portal vein are MRI and CT studies. The only noteworthy histopathological result from the liver biopsy is apoptotic alterations in the non-perfused parts and enhanced mitotic activity in the remaining areas.[3]

CASE REPORT

A 2-year-old male infant who was admitted with an intermittent fever, an increase in belly circumference, and a feeling of fullness in the abdomen from birth. After episodes of high-volume hematemesis, the patient complains of being lethargic. No reports of reduced urine production. Additionally, he had fresh blood in his stools for the past two days and melena. The patient was brought in for additional care.

Development and growth are age-appropriate, and there has been a history of right herniotomy. His immunizations were current, and his birth history was unremarkable. He measured 98.6 F, had a heart rate of 118/min, a respiratory rate of 26/min, blood pressure of 106/60 mmHg, and a Spo2 of 96% in room air. In addition to being pale, the patient showed no symptoms of cyanosis, lymphadenopathy, icterus, clubbing, edema, or dehydration. Upon physical examination, the patient had substantial splenomegaly, a soft, non-tender abdomen, and was vitally stable. The systemic assessment continued in an unremarkable manner. The results of preliminary laboratory investigations revealed lowered levels of RBC, WBC, platelets and Haemoglobin. Serum total protein, serum albumin, urine routine/microscopy, renal function test, and serology panel were all within acceptable limits.

A 12.4 cm enlarged liver was seen on the whole abdomen CT scan; there were no intrahepatic biliary dilatations to be found. The size of the spleen was significantly increased (14.6 cm). Focused lesions were not observed. Hepatic veins and intrahepatic inferior vena cava were both normal; there was no intrahepatic biliary dilation. There are two hemangiomas visible: one with a maximum dimension of 1.6 cm along the lateral surface of the higher pole and one with a diameter of 1.2 cm along the medial surface of the lower pole. All of the CT findings point to splenomegaly, portal hypertension, and extrahelatic portal venous blockage.

Gastroscopy was also done which showed 2 columns of large esophageal varices and 1 column of small varix in the esophagus and fundal varices in the stomach, owing to which the patient underwent endoscopic variceal band ligation. The child underwent spleenectomy+proximal spleenorenal shunt after all pre operative investigations. Intra operative and post operative period was uneventful. Postoperatively, the child was treated with iv antibiotics and analgesics. The patient was managed with 1 pint of fresh blood transfusion for reversing anemia. portosystemic shunt (spleno- renal) was done along with esophagogastric devascularization under general anesthesia. Multiple episodes of blood transfusion were also given to maintain a hemoglobin level at a acceptable level. On postoperative day 3 child underwent portosystemic shunt (spleeno-renal) Doppler, which showed a normal flow. A modified Makuuchi incision was created on the left side during the procedure, and the peritoneal cavity was opened layer by layer. The splenic vessels were severed and ligated. Spleno colic, dissected splenic hilum, and phrenico colic ligaments. Dissection of the splenic veins proximally. Dissected circumferentially and looped with a vascular sling, the left renal veins were discovered. Abdominal layers were closed with 2.0 vicryl and a 7.0 prolene proximal splenorenal shunt was performed. The child attained haemostasis and was hemodynamically stable.

He remained sedated and incubated. Dexmedetomidine, ketamine, and atracurium injections were used to maintain paralysis. Following surgery, the patient was given intravenous fluids, ranitidine, octreotide, intravenous antibiotics, and analgesics. Continuous monitoring was also carried out. Malena was observed in 5 instances, all of which were on the decline. The WBC count and CRP increased on the fifth postoperative day, and the condition improved in two days after clindamycin and meropenem injections were started.

Gradually, the patient's general condition improved, and the patient was discharged with Antibiotic prophylaxis and propranolol and planned for follow up after 2 weeks on an outpatient basis.

DISCUSSION

In paediatric patients, extrahepatic portal vein obstruction (EHPVO) is an uncommon cause of portal hypertension and frequently upper gastrointestinal haemorrhage, primarily from the gastric and esophageal varices.[4] Intestinal varices are uncommonly produced as new collaterals that accompany extrahepatic portal blockage and portal hypertension.[5]

Portal hypertension is mostly caused by extrahepatic portal venous obstruction (EHPVO), which is the second most prevalent cause worldwide. One of the main risk factors for EHPVO is portal venous thrombosis (PVT), a condition marked by partial or total blockage of blood flow in the portal vein due to a thrombus in the portal vein's main trunk, its intrahepatic branches, or the splenic or superior mesenteric veins.[6]

Portal hypertension is the most prevalent cause of upper gastrointestinal haemorrhages in children, accounting for up to 30% of cases that result in death. An EHPVO is typically the cause of paediatric portal hypertension. The etiology of portal venous obstruction and the variables that raise the risk of upper gastrointestinal bleeding in children with EHPVO are unknown. While drug usage or febrile conditions can cause some bleeding episodes, the majority occur spontaneously. With infrequent expansions into the superior mesenteric vein and the splenic vein, the portal vein is usually obstructed in EHPVO patients. Small amounts are only blocked at the hilum, where the portal vein terminates. An obstruction at the portal vein development is common, but it can occasionally affect the entire splenoportal axis.[7] The pathophysiology of EHPVO is typified by portal hypertension, which is caused by

prehepatic obstruction of portal blood flow. Furthermore, cavernous transformation of the portal vein (CTPV) is caused by the creation of hepatopeta collateral arteries around the destroyed portal vein.[8] EHPVO can arise from several variables such as congenital factors, omphalitis, prothrombotic conditions, dehydration, agenesis, and stenosis of the portal vein, as well as direct harm to the vein via catheterization of the umbilical vein.[9]

The three most typical presenting findings of EHPVO are splenomegaly, variceal hemorrhage, and anemic symptoms. Seventy-nine percent of the affected youngsters will experience at least one severe bleeding episode. In cases of esophagogastric varices, variceal bleeding may appear as hematemesis or melena; in cases of ectopic varices, it may subtly emerge as subtle gastrointestinal hemorrhage.[10]

In most cases, there is moderate to major splenomegaly, which in 5–10% of cases may be accompanied by hypersplenism. Short stature and slowed growth velocity may be caused by a decrease in hepatotropic growth factors. Doppler USG is the first-line radiological intervention that indicates portal vein obstruction and CTPV, followed by CT or MR imaging, for the diagnosis of EHPVO. Abdominal ultrasonography also enables for the measurement of liver and spleen echogenicity. Before shunt surgery, CT or MR angiography can be used to ensure adequate planning. Gastric varices can be identified by esophagogastroduodenoscopy. The diagnosis of hypersplenism is supported by the laboratory results of leucopenia, thrombocytopenia, and anemia.[10,11] The CT scan results in our case point to splenomegaly, portal hypertension, and extrahelatic portal venous blockage.

Controlling the acute variceal bleed and then moving on to secondary prophylaxis are the main goals of EHPVO care. The portosystemic shunt was created in our patient by utilizing proximal splenorenal shunt in conjunction with splenectomy. To lessen the esophagogastric varices, devascularization was additionally performed. Approximately 80% and 100%, respectively, of patients survive in the long run when the varices are properly removed and shunt surgery is done on time.

CONCLUSION

While EHPVO is a prevalent cause of noncirrhotic portal hypertension in children, it is difficult to treat and is not often identified. Although it is a frequent cause of upper gastrointestinal bleeding in children, there are still no appropriate guidelines for the management of this illness. A comprehensive approach to treatment and a precise diagnosis are essential to lowering the long-term morbidity and mortality associated with it.

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