

## A CASE REPORT OF ESOPHAGEAL VARICES, PANCYTOPENIA, SPLENOMEGALY DUE TO EXTRA HEPATIC PORTAL VEIN OBSTRUCTION

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**ABSTRACT**

**Introduction:** Extrahepatic portal vein obstruction (EHPVO) with Esophageal varices, pancytopenia and splenomegaly is rare in India with low prevalence, Esophageal varices can lead to recurrent gastrointestinal bleeding due to portal hypertension, pancytopenia due to hypersplenism and chronic blood loss. **Case presentation:** we report a case of 14 year old girl from north India with recurrent episode of gastrointestinal bleeding from Esophageal varices, with low haemoglobin, leukopenia and thrombocytopenia. In CECT and USG Doppler suggestive of extra hepatic portal vein obstruction. **Discussion:** Noncirrhotic portal hypertension is common in Extrahepatic Portal Vein obstruction. Its occurrence in the paediatric population is very rare. Portal hypertension can cause variceal bleeding and splenomegaly. Pancytopenia due to hypersplenism. The diagnosis of the EHPVO by imaging investigations like Doppler ultrasound, computed tomography, or magnetic resonance imaging required. A proper team channel including gastroenterologists, interventional radiologists, surgeons, and intensivists is crucial role in the management of varices. **Conclusion:** Extrahepatic Portal Vein obstruction in children is rare condition and difficult to diagnose and management.

**KEYWORDS :****INTRODUCTION**

Extrahepatic portal vein obstruction (EHPVO) is a disease of hepatic vein and is defined as extrahepatic portal vein obstruction either involvement of the intrahepatic portal vein, splenic vein, or superior mesenteric vein. In children and adolescent EHPVO is rare, with a prevalence of <5 cases per 10,000<sup>1</sup>. Portal vein thrombosis (PVT) is one of the most common causes of EHPVO in adults; however, half of EHPVO cases in children remain idiopathic<sup>2</sup>. Unexplained intestinal bleeding and splenomegaly in children should raise the physician's suspicion of EHPVO<sup>3</sup>. The non-invasive Diagnostic imaging of choice for EHPVO is abdominal Doppler ultrasonography, which visualizes the portal vein blood flow velocity with a sensitivity of 91 % and specificity of 100 %. Esophagogastroduodenoscopy is necessary to diagnose gastroesophageal varices. We report a case of portal hypertension with esophageal varices and pancytopenia in northern India.

**Case Report**

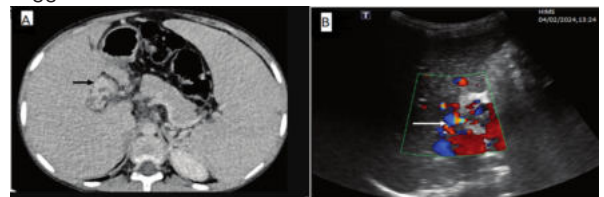
14 years old female from north India region presented to the emergency department with complaints of Blood in vomiting since 5 days which was sudden in onset, red in color, 2-3 episodes per day, around 50-60ml in each episode, non-projectile, not containing food particle. Patient also complaints of black color stool since 3 days, 4-5 episodes per day which was black tarry in color, foul smelling, semi-solid and sticky in consistency, associated with pain abdomen. The patient had a similar episode of complaints two year back for which she did not seek medical treatment and the episode was relieved on its own without any further similar episodes in between. There is no history of bleeding manifestation in the patient or among the family members.

On examination, patient conscious, cooperative and oriented

to time, place and person. BMI – 12.2 kg/m<sup>2</sup>, Pallor present, afebrile, tachycardia present, blood pressure of 100/60 mmHg, respiratory rate-18/min with a SpO<sub>2</sub> of 99% in room air. There was no jaundice or lymph node enlargement, clubbing, cyanosis, systemic examination revealed tenderness with massive splenomegaly. The other systemic examination was unremarkable.

On blood investigations, haemoglobin was 4.9g/dL, RBC-2.62millions/mm<sup>3</sup>, TLC 2200/mm<sup>3</sup>, PCV-17.8, Platelets-67000/mm<sup>3</sup>, total bilirubin 1.83 g/L, direct bilirubin 0.90 g/L, ALP 281 with other blood parameters within normal range. In view of low haemoglobin, a blood transfusion was done. For the evaluation of suspected lower gastrointestinal bleed, an upper endoscopy was performed which shows grade IV esophageal varices, isolated gastric varices. (Fig. 2 A, C)

Ultrasonography her abdominal ultrasound with Doppler studies and Contrast-Enhanced Computed Tomography (CECT) (Fig.1) abdomen revealed liver normal sized, enlarged spleen and numerous collateral vessels in region of porta hepatis consistent with cavernous malformation of portal vein with dilated splenic vein, all these findings were suggestive of EHPVO



**Fig. 1.** Contrast enhanced computed tomography (CECT) axial section (A) shows cavernoma of portal (black arrow) splenomegaly and relatively smooth liver parenchyma. Color

Doppler image (B) in a patient with extrahepatic portal vein obstruction (EHPVO) shows multiple tortuous collaterals at the porta instead of by main portal vein.

At the time of presentation conservative management was done, in view of low haemoglobin PRBC was transfused and given prophylactic antibiotics, patient admitted in medicine department, after upper GI Endoscopy Esophageal Variceal Ligation (EVL) done (Fig 2 B) and started non selective beta blocker.



**Fig. 2.** (A) In Upper GI endoscopy Esophageal varices shows, (B) Esophageal variceal ligation (EVL) done, (C) Gastric varices seen.

The patient was diagnosed with EHPVO with pancytopenia secondary to hypersplenism, and portal hypertension gastropathy. Patient managed symptomatically with pain relief and beta blocker. In anaemia 3 unit of packed red blood cells (PRBC) was transfused, with the haemoglobin rising to 8 g/dL. Esophageal Variceal ligation (EVL) was done. The patient was planned to be started on oral anti-coagulants for the EHPVO After resolution of thrombocytopenia and was discharged from the hospital in a hemodynamically stable condition.

## DISCUSSION

Extrahepatic portal vein obstruction which has also been called extrahepatic portal vein thrombosis and cavernous transformation of the portal vein.<sup>4-7</sup> Some patients in this category do have risk factors for thrombosis of the vessel such as neonatal umbilical vein catheterization, omphalitis, intraabdominal sepsis such as appendicitis or primary peritonitis, blunt abdominal trauma, neonatal dehydration and hypercoagulable states such as factor V Leiden deficiency or protein C and protein S deficiencies. The most common cause of extrahepatic portal hypertension is idiopathic. Variceal bleeding, anemia, and splenomegaly are the three most common features of EHPVO. At least one significant bleeding episode will occur in about 79 % of the affected children.<sup>8</sup> Variceal bleeding may occur as hematemesis or melena resulting from Esophageal and gastric varices, or it may occur with unexplained gastrointestinal bleeding, as in ectopic varices.<sup>9</sup> Moderate to massive splenomegaly is frequently present and may be accompanied by splenomegaly in 5% to 10% of patients<sup>10</sup>.

Our patient had hematemesis and melena and low haemoglobin due to chronic blood loss for the diagnosis of EHPVO, ABDOMINAL Ultrasonography can measure the echogenicity of the liver and spleen and Doppler USG shows portal vein obstruction and CTPV, followed by CT or MRI imaging. CT or MRI angiography may be used before surgical management. Esophagogastroduodenoscopy can detect gastroesophageal varices. And laboratory finding of pancytopenia due to hypersplenism (anemia, thrombocytopenia and leukopenia) support the diagnosis<sup>8,11</sup>.

In our case, there was splenomegaly measuring 21 \* 8 cm size in CECT of abdomen; splenic vein was dilated and tortuous with multiple collaterals along with the CTPV and dilated Porto systemic vascular channels. EHPVO management primarily focuses on containing the acute variceal bleed followed by secondary prevention. For patients with a recent EHPVO, Baveno V recommends anticoagulant therapy with LMWH followed by oral anticoagulants for at least 3 months. In case, the consensus fails to address the use of anticoagulants, and evidence in favour of TIPS and local

thrombolytic is also lacking. Endoscopic management is recommended for primary prevention of acute Variceal bleeding, and beta blockers are equally effective in secondary prevention<sup>12</sup>. In addition guidelines recommends left Mesenteric portal vein bypass (Rex Procedure) in children with chronic EHPVO although proximal splenorenal shunt has been shown an effective alternative in cases of difficult anatomical variation<sup>12, 13</sup>. With excision of the varices and timely performed shunt surgery, long-term survival are reaches 100% and 80% respectively<sup>5</sup>.

## CONCLUSION

EHPVO is a common cause of noncirrhotic portal hypertension in children and adolescents; however, it is less commonly diagnosed, and its treatment is challenging. Extrahepatic Stigmata of chronic liver disease are usually not present except for an enlarged spleen. The onset of bleeding typically occurs in a previously healthy patients. Prognosis is good. Frequency and intensity of the haemorrhagic manifestations decrease over the time, because of formation of collaterals. These patients may die from acute severe upper gastro intestinal bleeding. Although this condition is one of the cause of upper gastrointestinal bleeding in children and adolescent population but lacking of proper guidelines in the management of this condition. A careful diagnosis and multidisciplinary treatment are necessary to reduce the long-term morbidity and mortality associated with it.

## Ethical approval

Not required.

## Funding

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## Consent for publication

Written informed consent was obtained from the patients' father for publication of this case report and accompanying images. A copy of the written consent is

## Declaration of competing interest

None.

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