Case Report

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Case Report on Extrahepatic Portal Venous Obstruction

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Abstract

Extra Hepatic Portal Venous Obstruction (EHPVO) is an major cause of Portal Hypertension. Pediatric EHPVO is idiopathic in most of the cases. A child who had normal growth and development, having complaints similar to upper GI bleeding underwent video Oesophago Gastro Duodenoscopy (VOGD Scopy) and was found having sclerosed Oesophageal varices in the form of snake like pattern over the entire mucosa. Child was treated with Sclerotherapy.

Keywords: EHPVO; Portal hypertension; Esophageal varices; Sclerotherapy.

Clinical Report of Patient: An 8 year old child admitted to hospital with complaints of hematemesis, sudden in onset, consistent blood clots, in large bouts. Child had melena episode, low grade fever, and per abdomen no organomegaly found. Child is diagnosed as a case of Grade II Esophageal varices, Extrahepatic portal venous obstruction with Portal Hypertension. On Physical examination had a soft non tender abdomen with significant splenomegaly. Child was treated with Inj. Octreotide infusion 1mcg/kg bolus, then IV infusion 1mcg/kg/hour for 24 hours according to body weight, Tab. Proponolol 40 mg ¼ Bd, Tab. Rantac ½ Bd, and child got sclerosed of esophageal varices for the entire mucosa. After sclerotic therapy child had no evidence of bleeding. But child was advised to continue sclerotherapy once in every 3 months till gets relieved from variceal recurrences.

INTRODUCTION

Extrahepatic portal vein obstruction (EHPVO) is commonly due to portal vein thrombosis, resulting in portal hypertension, and is a major cause of upper GI bleeding in children.

Definition

EHPVO is characterized by cavernomatous transformation of Extrahepatic portal vein as which results in portal hypertension and its consequences. The common site where it occurs is portal vein and at the junction of splenoportal axis.[1]

Incidence

According to the WHO, EHPVO is considered as a rare disease with 0.05% prevalence. But in few developing countries, amounts to 30–55% of variceal bleed and 70% of pediatric patients

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with portal hypertension. Most the data in India shows that EHPVO is the cause for 54% of portal hypertension and 85-92% of bleeding of upper gastrointestinal tract in children.[1]

Etiology

The etiology is heterogeneous with respect to age and geographical locations:[1]

1. Anomalies of Portal vein such as agenesis, atresia or stenosis.
2. Venous anomalies and anomalies of vitelline vein.
3. Gene mutation, activated protein C resistance and myeloproliferative disorders.
4. Umbilical Sepsis and Umbilical vein catheterization.
5. Other causes include omphalitis, neonatal sepsis and portal vein.
6. Other causes include any prothrombic state that can induce thrombus formation likepancreatitis, omphalitis, neonatal sepsis and direct injury to portal vein.

Pathology

Portal cavernoma is the hallmark of EHPVO. This thrombus formation leads to absence of bile flow in the portal vein and results information of collateral vessels in common bile duct with transportation of bile to liver. Cavernoma formations can occur in any of anatomical structures present around the liver. The formed collateral vessels are inadequate to decompress the portal pressure which produce porto-systemic shunt and results in esophageal varices.

EHPVO Classification

Site	Type 1:	Only trunk involved
	Type 2:	Only branch: 2a–One; 2b-both branches
	Type 3:	Involvestype 1 and type 2
Presentation	R:	Recent
	Ch:	Chronic (with portal cavernoma and PHT)
Type of underlying liver disease	C:	Cirrhotic
	N:	Noncirrhotic diseases of liver
	H:	Hepatocellular carcinoma and other near by malignancies
	L:	Post liver transplantation
	A:	Absence of underlying liver disease
Degree of portal venous system occlusion	I:	Incomplete flow to the portal vein as seen radiographically
	T:	Total no flow in portal vein lumen on imaging
Extent of PV system occlusion	S:	Splenic vein
	M:	Mesenteric vein or both

Clinical Features[1]

Clinically, it is manifested as esophageal and/or gastric varices, foregut bleeding, splenomegaly, hypersplenism, growth retardation and/or neurocognitive impairment:

1. ***Variceal Bleeding:*** It can manifest as a sudden episode or secondary to a precipitating event. More often than not, EHPVO patients tolerate bleeding episodes better.
2. ***Ascites:*** Ascites is usually transudative and transient, seen during episodes of hemorrhage or immediately following surgery.
3. ***Growth Retardation:*** Growth retardation is because of reduced hepatic portal flow which deprives the body of hepatotropic hormones.
4. ***Splenomegaly and Hypersplenism Splenomegaly:*** It is the second common form that is present with abdominal mass and pain. Mild enlargement of spleen is seen in 40 – 80% of these patients.
5. ***Portal Biliopathy/Portal Cavernoma Cholangiopathy (PCC):*** PCC means any abnormalities of intra or extrahepatic bile ducts and gallbladder wall which are noticed on ERCP and ERCP changesin portal biliopathy are classified as;

Type I: Involvement of only extrahepatic bile duct.

Type II: Involvement of only intrahepatic bile duct.

Type IIIa: Involvement of extrahepatic and unilateral intrahepatic bile duct.

Type IIIb: Involvement of extrahepatic and intrahepatic ducts bilaterally.

DIAGNOSIS

A clinical diagnosis can be made with a detailed history and physical examination demonstrating splenomegaly.

Thrombosed portal vein, cavernoma formation is evidenced on USG.

Other Noninvasive imaging studies like CT angiography and MR angiography are an alternative for invasive splenoportography.[3]

MANAGEMENT

It includes the treatment of variceal bleeding, growth retardation, portal biliopathy, hypersplenism, and massive splenomegaly. Therapeutic options range from conservative medical therapy, endotherapy and endoscopic variceal ligation to surgical intervention.[3]

Treatment of Variceal Bleed Medical Management Acute variceal hemorrhage requires emergency treatment. In common the treatment options available are – Medical management, endoscopic variceal obliteration and surgical options.

In an acute variceal bleeding, somatostatin/ octreotide/terlipressin can decrease the variceal pressure and help in stabilizing the patient before an endoscopic therapeutic intervention.

Endoscopic sclerotherapy (EST) and endoscopic variceal ligation (EVL) are successful means to achieve hemostasis with success upto 96%.

Endoscopic Sclerotherapy (EST) is well established treatment for bleeding GI varices, accomplishes vascular obliteration by injection of a sclerosing agent. When Sclerotic agents are injected into the site or adjacent to blood vessels it causesvascular thrombosis and endothelial damage leading to endofibrosis and vascular obliteration.

The most commonly used sclerosants are the synthetic chemicals and fatty acid derivaties like sodium tetradecyl sulfate, polidocanol, sodium morrhuate and ethanolamine oleate.[5]

Pediatric dose is not standardly prescribed for the sclerosants. Many pediatric GI specialist use 1/4th of adult dose for less than 12 years of age, depending upon the size.

Technique for Sclerotherapy: To manage bleeding, a sclerosant is sent into the cavity with a sclerotherapy needle through an endoscope. In children usually 25 gauge needleis used. Sclerosants may be injected into variceal locations. The sclerosants induces the thrombosis and occlution of the lumen of the varix. Paravariceal injection, occludes the varix by tamponade and induces fibrosis of tissue around the varix. It can sometimes cause temporary bleeding during the procedure. There is no evidence that one technique is better than the other.[5]

Endoscopic variceal ligation (EVL) is the procedure of choice in primary prophylaxis. EST is effective in eradicating esophageal varices in 88–100% cases. Initially endoscopy is done to detect the severity and site of varix. 5-10 bands are placed from gastroesophageal junction and progressing upward to 5-8 cm distance. 2–4 sessions are required to treat recurrence of variceal bleeding with an interval of 2–4 weeks.

SURGERY

When the bleeding cannot be controlled, emergency surgery such as shunt procedures or devascularization is required. Surgery is done when there is failure of endoscopic management or bleeding is not controlled to endoscopic treatment.

The preferred surgical interventions are portosystemic shunts (PSS) and ablative procedures.

Porto-Systemic Shunts (PSS) PSS diverts blood flow from the high pressure portal circulation to low pressure systemic circulation by anastomosis between a tributary of the portal vein (splenic, superior mesenteric, and left gastric, left gastroepiploic) and a systemic vein (renal, inferior vena cava, and adrenal). The shunts may be either selective or non selective.[4]

Variceal Ablative Procedures These procedures include splenectomy alone or esophageal devascularization. These procedures are indicated when no suitable vein for shunt is available or as salvage therapy for failed endoscopic variceal bleeding control.[4]

SUMMARY

The prognosis is good in children with EHPVO without cirrhosis. During acute bleeding, pediatric nurses are responsible for maintain of airway with intubation and carefully monitor the child who is on octreotide administration. Children should follow up for every 3–6 months. Nurses responsibility is to educate the parents on further sclerotherapy and make adequate visits to the hospital for the same.

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