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Extra Hepatic Portal Hypertension with Clubbing, Cyanosis, Chest Deformity- A Case Report

Md. Benzamin[1*], Md Wahiduzzaman Mazumder[2], Fahmida Begum[3], Mukesh Khadga[1], Zannatul Ferdous Sonia[1], Md. Rukunuzzaman[2], A.S.M Bazlul Karim[4]

[1]Resident (phase-B), Department of Pediatric Gastroenterology and Nutrition, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka, Bangladesh

[2]Associate Professor, Department of Pediatric Gastroenterology and Nutrition, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka, Bangladesh

[3]Assistant Professor Department of Pediatric Gastroenterology and Nutrition, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka, Bangladesh

[4]Chairman, Department of Pediatric Gastroenterology and Nutrition, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka, Bangladesh

***Corresponding author**
Md. Benzamin

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Abstract: Portal hypertension is not uncommon in children. Extra hepatic portal venous obstruction (EHPVO) constitutes about 75% of portal hypertension. Several risk factors predispose to development of EHPVO such as neonatal sepsis, umbilical catheterization, severe dehydration, abdominal trauma or surgery etc. But risk factors are usually detected in less than half of patients. Patient commonly presents with variceal bleeding with splenomegaly. Here we are presenting a case with some atypical features.

Keywords: Portal hypertension, Children.

INTRODUCTION

Portal hypertension is a condition that occurs due to the formation of portal–systemic collaterals which shunt a portion of the portal blood flow to the systemic circulation, bypassing the liver. It can arise from disorders with blood flow at any level within the portal system [1]. Extra hepatic portal venous obstruction (EHPVO) is an important cause of portal hypertension which constitutes 68-76% of portal hypertension in children from developing countries [2]. The relation of haematemesis, splenomegaly and portal hypertension was first recognized by Banti almost a century ago. Majority of cases are due to primary thrombosis of the portal vein [2].

CASE REPORT

Zahid, 10 year old boy, 1st issue of non-consanguineous parents presented with history of haematemesis and melena for 10 days and bluish discoloration of lip since 4 years of his age. His informant mother stated that Zahid developed bluish discoloration of lip and finger tips initially during physical activity but later on it also appeared during rest condition and persisting till now. Zahid had haematemesis 2 times and melena 3 times in last 10 days.

On query, mother gave history of low grade evening rise of temperature for last 1 year associated with dry non-productive cough. He was not growing like other peers. He had no history of jaundice, contact with TB patient, neonatal sepsis, umbilical catheterization, abdominal trauma, severe dehydration or any surgery.

On examination, Zahid was dyspneic, cyanosed (figure1), anicteric, conjunctiva congested, no lymphadenopathy, pigeon chest deformity (figure 2), BCG mark present, clubbing present (in all 4 limbs,stage 4 ,figure 1), SPO_2 at room air: 70% and with 4ltr/min O_2: 80%. Anthropometrically, Zahid was severely wasted, moderately stunted. On systemic examination, vesicular breath sound with few crepitations present in both lung fields, no murmur was found. Hepatomegaly(3cm) and Splenomegaly (7cm) present, no ascites seen. On locomotor system examination, gibbus present at T6-T7 level.

Investigations: revealed Hb%: Increased (17.1gm/dl), high ESR (60 mm in 1st hr), liver function test: Normal, esophagogastrodudenoscopy showed Grade IV esophageal varices, MT: Negative, chest Xray: Miliary mottling (figure 3), sputum for AFB and Gene Xpert: Negative, lung perfusion test: Normal, echocardiography: Normal, transesophageal

echocardiography: Mild Atrial Septal Defect(ASD), contrast enhanced Echocardiography: Pulmonary arterio-venous shunt.(table 1).

Table-1: Investigations

CBC : Hb - 17.1 gm/dl, HCT: 55.7% ESR- 60 mm in 1st hr TC of WBC: 4,480/cmm DC: N-50%, L-46% Platelet: 77,000/cmm
Liver Function Test(LFT): S. ALT- 14 U/L PT: 14.2 sec S. Albumin: 3.6 gm/dl
Endoscopy of Upper GIT: Grade IV oesophageal varices seen.
USG of Whole Abdomen: Coarse hepatic parenchyma with splenomegaly. X-ray of thoraco-lumbar spine: Dorsal kyphotic curvature exaggerated. Destruction with vertebral body height reduction and end plate irregularity noted at D6,D7 vertebra with reduction of intervening intervertebral disc causing focal kyphosis.(figure 4)
Colour Doppler USG of HBS: Coarse hepatic parenchyma, portal vein shows hepato-petal flow, mean velocity 10.22 cm/s, no thrombus or cavernous transformation seen which is consistent with portal hypertension.
MT: Negative Chest X-ray: Miliary mottling present. Sputum for AFB: Negative. Sputum for Gene Xpert: Negative.
Echocardiography: Normal
Pulmonary Perfusion Scintigraphy: Normal perfusion(bilateral)
Contrast Echocardiography: Pulmonary Arteriovenous Shunt. Transesophageal Echocardiography: Mild ASD.
Bone Marrow Study: Erythroid hyperplasia with increase number of plasma cells. Other marrow elements maturing.

We finally diagnosed this case as Congenital Cyanotic Heart Disease (Pulmonary arteriovenous malformations) with Extra-hepatic portal hypertension with disseminated Tuberculosis.

DISCUSSION

Portal hypertension develops from obstruction of portal circulation, either within or without the liver. Extrahepatic portal venous obstruction (EHPVO) is the commonest cause of portal hypertension and variceal bleeding in children in developing countries [2]. As per the Asia Pacific association for study of Liver (APASL) consensus 2006, EHPVO is defined as ‘‘a vascular disorder of liver, characterized by obstruction of the extrahepatic PV with or without involvement of intrahepatic PV radicles or splenic or superior mesenteric veins” [3]. Variceal bleed (49–85%) and splenomegaly (63–88%) with no stigmata of chronic liver disease are the commonest presentations in children. Mean ages of first bleeding episode are around 3.8–5.2 years [4, 5].

Our patient presented with variceal bleeding in the form of haematemesis and melena, splenomegaly and no stigmata of chronic liver disease (CLD) except clubbing. The risk factors are usually detected in less than half of patients. Common local causes are:

Omphalitis, portal vein phlebitis, umbilical vein catheterization (UVC), pancreatitis, liver abscess, surgery around portal vein (splenectomy, cholecystectomy, Billroth-II procedure) and malignancies (pancreatic, hepatic or duodenal) [6]. Others: diarrheal illnesses, abdominal sepsis, and nephrotic syndrome [7]. In our patient, no risk factors were found.

For diagnosis of extrahepatic portal hypertension, Liver function test (LFT): Usually normal. Elevations of alkaline phosphatase and gamma-glutamyl transpeptidase are seen with development of portal biliopathy, while hypoalbuminemia may be seen during bleed episodes. Doppler to see PVT(portal vein thrombus) and portal vein cavernoma (sensitivity and specificity of >95%) [8]. In our patient, LFT was normal, esophagogastrodudenoscopy showed Grade IV esophageal varices, Doppler ultrasonography of hepatobiliary system showed hepato-petal flow, mean velocity 10.22 cm/s, no thrombus or cavernous transformation seen which is consistent with portal hypertension. As thrombus can be formed at any site of

portal tributaries, our patient might have thrombus in intrahepatic portal tributaries.

Pulmonary arteriovenous malformations (PAVM) are rare pulmonary vascular anomalies. The incidence of PAVM is 2–3 per 100 000 population [9]. About 13% to 55% of patients are asymptomatic [10]. The common presenting symptom is dyspnoea on exertion, which is seen in 31% to 67% of patients. Others include epistaxis, haemoptysis, palpitation, chest pain, cough, bruit/thrill, clubbing, cyanosis, telangiectasia, polycythaemia [10, 11]. In one study, the classic triad of dyspnoea, cyanosis, and clubbing was found in 10% of patients with a PAVM [12]. Chest radiography, Contrast echocardiography, Contrast enhanced computed tomography, pulmonary angiography (gold standard) are diagnostic tools [11, 13]. In our patient exertional dyspnoea, cyanosis, clubbing, conjunctival congestion were present, SPO_2: 70% in room air and these features were seen from early childhood. Polycythemia was evidenced by high hemoglobin and bone marrow study. His chest x-ray, echocardiogrphy did not give any clue but contrast echocardiography demonstrated a delay of 1 sec before the bubbles were visualised in the left atrium which showed pulmonary arterio-venous shunt (figure 4). Due to this cyanotic congenital heart disease, patient developed polycythemia which is a risk factor for developing thrombus resulting in Extra-hepatic portal hypertension.

Polycythemia related to hypertension and vascular abnormalities are caused by increased red cell mass. An episode of venous or arterial thrombosis like mesenteric, hepato-portal or splenic vein thrombosis should always raise suspicion of polycythemia vera as a possible cause [14].

Tuberculosis is an important global health problem and Bangladesh is one of the high tuberculosis burden countries in the world. It includes pulmonary and extrapulmonary form. Studies from Bangladesh showed that extrapulmonary form is more common in children [15, 16]. Disseminated tuberculosis is defined as tuberculous infection involving the blood stream, bone marrow, liver or involvement of two or more non-contiguous sites or miliary tuberculosis [17]. It is common among the infants and children [18]. Our patient had low grade fever for 1 year with cough, anorexia, failure to thrive. On examination he was severely wasted, moderately stunted, vesicular breath sound with few crepitations present in both lung fields, hepatomegaly (3cm), splenomegaly (7cm), gibbus present at T6-T7 level. Investigations showed raised ESR, Miliary mottling on Chest Xray, destruction of ribs (T6-T7) with kyphosis. Disseminated TB can also result in Extra-hepatic portal hypertension as it causes phlebitis which facilitates thrombus formation [19-21].

The patient was managed with Endoscopic variceal band ligation, anti-TB regimen according to National Guideline along with nutritional support. He was given brace for kyphosis (figure 6). For pulmonary arterio-venous shunt, patient was referred to Paediatric Cardiology Department for Cardiac Catheterization and shunt closure.

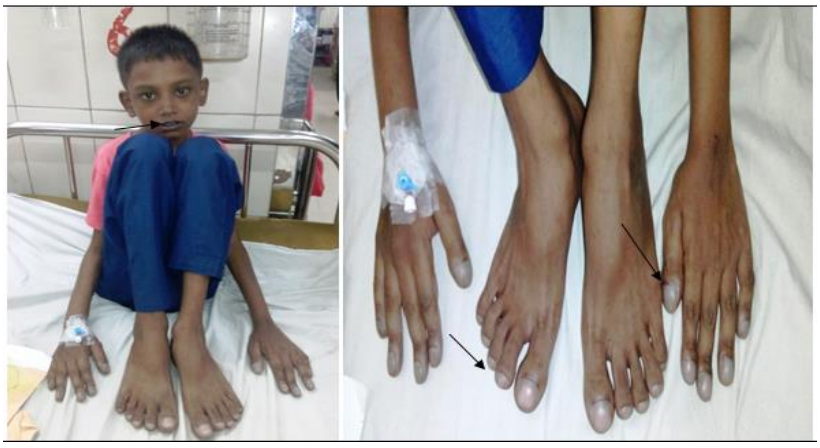

Fig-1: Cyanosis, clubbing

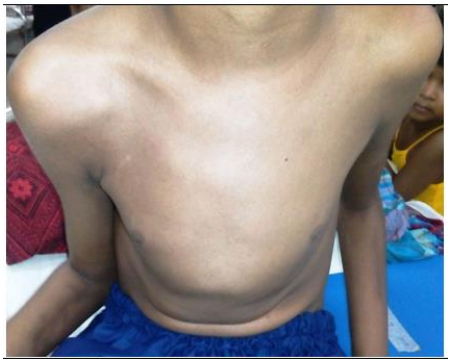

Fig-2: Pigeon chest deformity

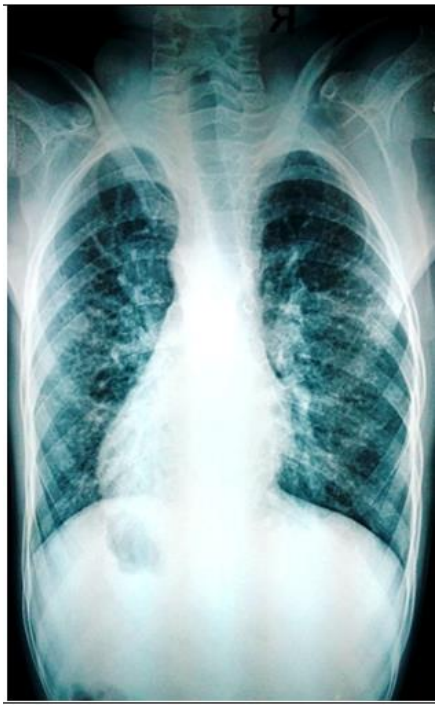

Fig-3: Miliary mottling

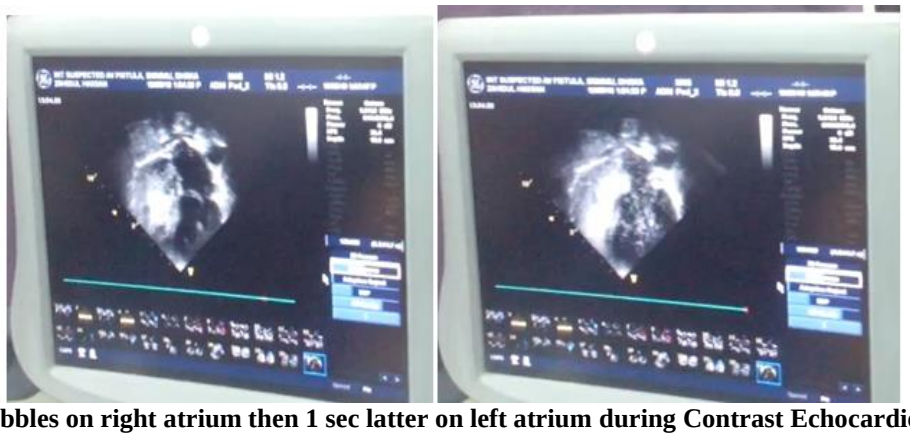

Fig-4: Bubbles on right atrium then 1 sec latter on left atrium during Contrast Echocardiography.

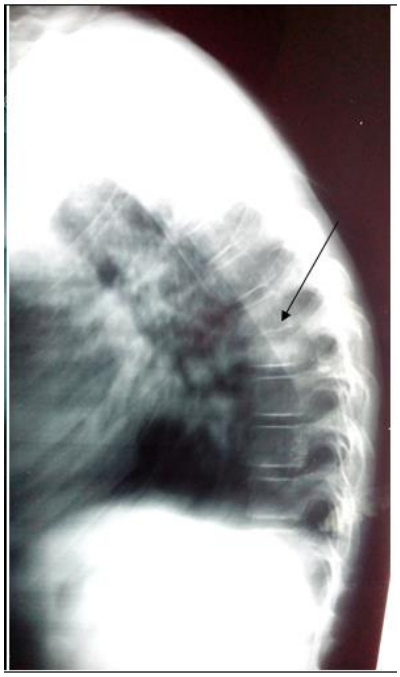

Fig-5: Destruction with body height reduction D6, D7 vertebra

Fig-6: After 4 weeks of treatment

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