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Case Report

Rupture of esophageal varices due to portal hypertension

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Variceal bleeding is the most common cause of serious upper gastrointestinal (UGI) bleeding in children. Most variceal bleeding is esophageal.[1] Hemorrhages from esophageal varices due to portal hypertension are a major cause of morbidity and mortality. There is a 30% mortality rate following an initial episode of variceal hematemesis. Mortality increases to 70% with recurrent variceal hemorrhage. Moreover, the one year survival rate after variceal hemorrhage is often poor (32 to 80%).[2-4]

We report a case of esophageal varices rupture caused by portal hypertension, an emergent case in the Pediatric Gastrohepatology division.

Case Report

An 8 ½ year-old girl, was hospitalized in the pediatric ward at M. Djamil Hospital, Padang for 30 days (August 21st – October 19th 2007). She was referred from Achmad Mochtar Hospital, Bukittinggi, complaining of hematemesis lasting 7 days. Hematemesis occurred 4 to 7 times per day, each incident yielding emesis volume of ½ to 1 glassful. The emesis contained fresh blood mixed with food. The last hematemesis episode occurred 4 days prior to hospitalization. She had a tar-like stool 2 days prior to admission. She had a high, continuous fever without shivering beginning 1 day prior to admission. There was no history of cough, breathlessness, or spontaneous bleeding of the skin, nose, gum or other sites. She had no history of abdominal pain and urination was normal. She had been hospitalized in Achmad Mochtar Hospital for 7 days, and had received a blood transfusion prior to transfer to M. Djamil Hospital.

The patient was never jaundiced and was taking no long-term medications or herbs. She had experienced recurrent bloody vomiting 4 years and 2 years previously, and had been hospitalized in Achmad Mochtar Hospital for 9 days and 11 days, respectively. There was no family history of jaundice or liver disease. She was vigorous at birth and had a normal growth and development pattern.

Physical examination revealed a conscious, moderately ill child. She had normal vital signs including blood pressure 100/70 mmHg, pulse 126 times/minute, respiration 28 times/minute, and temperature 38.6 ^{O}C. Her body weight was 19 kg and height was 122 cm. She was undernourished (weight for age 67.85%, height for age 93.12%, weight for height 86.36%). Her skin was warm and showed no petechiae, purpura, bruising, cyanosis, icterus or palmar erythema.

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There was no lymph node enlargement. The patient's head, pupils, ears and nose were normal. Conjunctiva was anemic and sclera was not icteric. No anomalies were found during neck and chest examination. The abdominal wall was tender without distension, caput medusae, or venectasia. Shifting dullness was negative. The liver was not palpable, but the spleen was slightly enlarged (S1). Genitalia were normal at pubertal stage A1 M2 P1. Extremities were normal and showed no pretibial edema.

Laboratory results revealed hemoglobin was 10.5 g/dl, hematocrit 34%, platelet count 170,000/μl, erythrocyte 3.2 million/μl, reticulocyte 7%, MCH 30.88 pg, MCV 97 fl, and MCHC 32.81%. Leukocyte count was 20,200/μl, with 2% bands, 89% segments, 7% lymphocytes, and 2% monocytes. A peripheral blood smear showed polychromatic erythrocyte and anisocytosis. Urine and stool examinations were normal. Total bilirubin was 1.12 g/dl, indirect bilirubin 0.74 mg/dl, and direct bilirubin 0.38 mg/dl. Serum aspartate aminotransferase was 64 U/L, alanine aminotransferase 28 U/L, alkaline phosphatase 69 U/L, and γ-glutamil transferase 6 U/L. Total protein was 4.5 g/dl, with albumin 2.7 g/dl, and globulin 1.8 g /dl. Prothrombin time (PT) was 13.1s and activated partial thromboplastin time (aPTT) was 38.3s. Liver function tests showed slightly abnormal results (increased levels of serum aspartate aminotransferase and hypoalbuminemia). Hepatitis markers (HBsAg, anti HBc, and anti HCV) were absent.

The patient underwent abdominal ultrasonography (USG) and CT-scan. The USG showed liver enlargement with a granular surface, rough heterogeneous parenchyma, and a blunt edge (**Figure 1**). Portal vein diameter was 11.6 mm, with normal biliary duct and enlarged spleen. The USG suggested hepatic cirrhosis, splenomegaly, and portal hypertension. The CT-scan showed a normal-sized but hyperdense liver, with widening of the vascular diameter (**Figure 2**). The bile duct and gallbladder were normal in size, and no gallstones were observed. However, the gall bladder wall was thickened. Pancreas and kidneys appeared normal. The CT-scan confirmed the hepatic cirrhosis and portal hypertension diagnoses. Confirmation of the diagnosis of hepatic cirrhosis by liver biopsy was not performed due to lack of parental consent.

On the second day of hospitalization, the patient had 500 ml of bloody emesis. She was somnolent, pale, and had labored breathing. Her blood pressure was 40/20 mmHg. Her heart rate was tachycardic at 150 times/minute, with respiratory rate of 32 times/minute, and body temperature of 36.2 ^{O}C. Her extremities were cold with poor perfusion. The patient was in hypovolemic shock caused by massive bleeding. Resuscitative measures were begun immediately by giving oxygen at 2 L/minute and a bolus of 20 ml/kg lactated Ringer's solution. Her state of hypovolemic shock was controlled within 30 minutes. She received a transfusion of 10 ml/kg whole blood because her hemoglobin had decreased to 4.8 g/dl. She received a maintenance solution (KaEN 1 B 65 ml/kg/day) while she temporarily fasted. She also received an octreotide drip (25 μg in 20 ml 5% dextrose), administered for 20 minutes, followed by 100 μg ocreotide in 100 ml 5% dextrose for the next 4 hours. Therapy also included intravenous injection of tranexamic acid (200 mg three times daily), ranitidine (20 mg twice daily), vitamin K (2 mg), cefotaxime (500 mg twice daily),

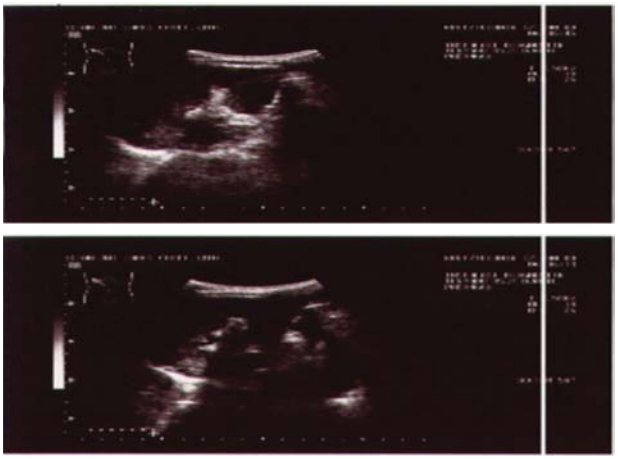

Figure 1. Liver ultrasonography (USG)

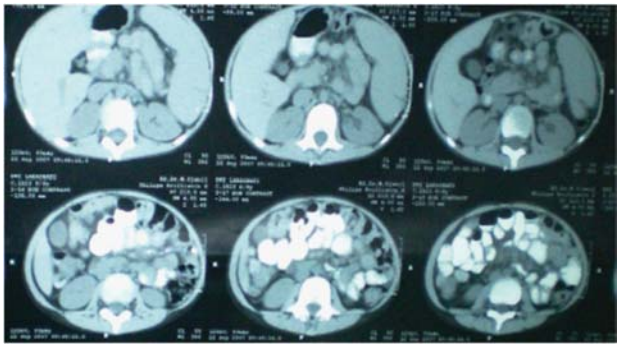

Figure 2. Abdominal CT-scan

and lactulose. Her vital signs, urination, and fluid balances were strictly monitored. A nasogastric tube was inserted to remove blood from the gastrointestinal tract. Blood gas analysis, electrolytes, random blood glucose examinations, and blood cultures were performed. Blood gas analysis was normal (pH 7.39, pCO_2 37 mmHg, pO_2 88 mmHg, HCO_3^- 20 mmol/L, BE -1.5 mmol/L, SO_2 97%). Random blood glucose (95 g/dl), sodium (Na 136 mg/dL), and potassium (K 3.6 mg/dL) were also normal.

Endoscopic examination was done after the patient was in a stable condition (**Figure 3**). The esophagus appeared hyperemic and eroded, with bleeding and stigmas present. Endoscopy showed grade III esophageal varices. Sclerotherapy endoscopy (STE) was performed to prevent subsequent rupture of the varices (**Figure 4**).

Based on the data above, it was concluded that the patient had ruptured esophageal varices due to portal hypertension related to liver cirrhosis. Differential diagnoses included portal vein thrombosis and idiopathic portal hypertension. The patient was

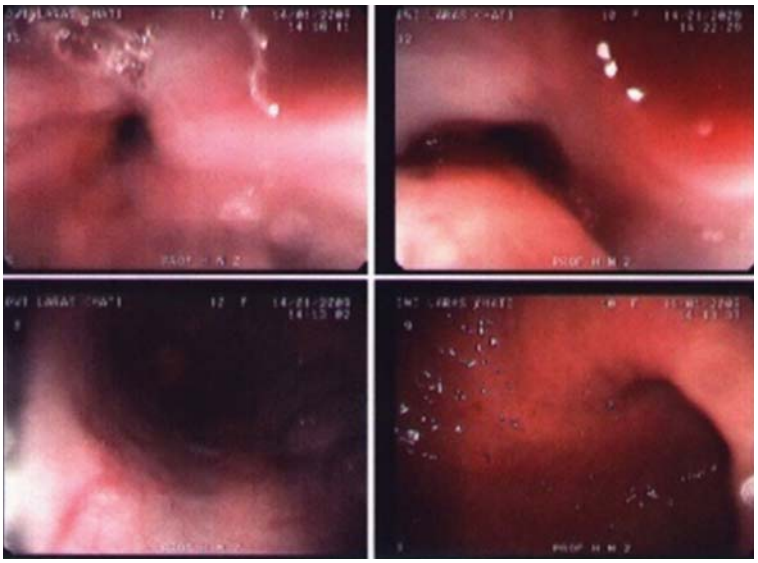

Figure 3. Endoscopy before STE

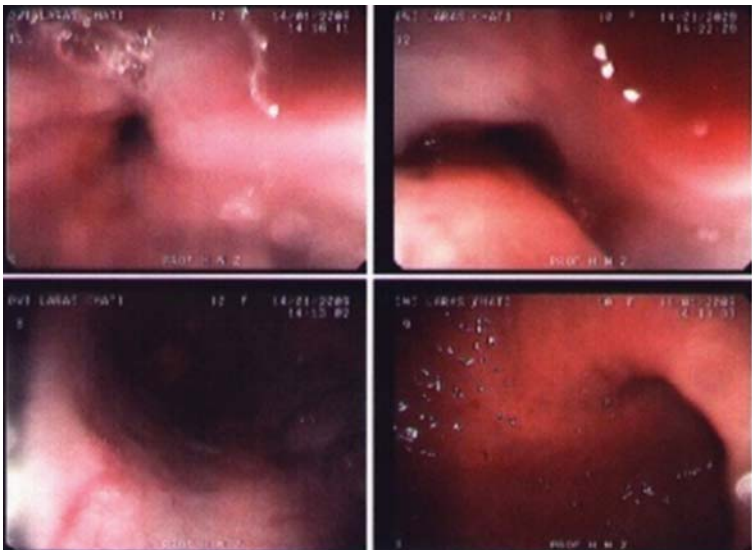

Figure 4. Endoscopy after STE

Table 4. The correlation of clinical variables with PCT and bacteremia

Clinical variables	PCT		P	OR (CI 95%)	Blood culture		P	OR (CI 95%)
	Pos	Neg			Pos	Neg		
Age*#)								
<1 years	14	11	0.378	2.29	11	14	0.003	23.00
>1 years	5	9		(0.49 to 11.04)	0	14		(2.82 to 187.5)
General appearance*								
Severe illness	13	4	0.006	8.67	7	10	0.158	3.15
Mild/moderate	6	16		(1.66 to 50.56)	4	18		(0.61to 17.38)
Blood culture								
Positive	9	2	0.025	8.10				
Sterile	10	18		(1.22 to 68.02)				

*) Absolute Fisher Test (performed on blood culture)

#) Formula Corrected OR (for blood culture test based on age)

discharged after 30 days of hospitalization. No bleeding was observed during a second STE. Upon discharge the patient was afebrile and able to eat soft foods. Her vital signs were normal. Her liver was not palpable and her spleen was S1. Hemoglobin was 14.4 g/dl, leukocytes 6100/μl, and platelets 98.000/μl. Propranolol 10 mg was given twice daily. A follow-up visit to the pediatric ambulatory department was suggested.

Fourteen months after discharge, the patient had recurrent hematemesis and was rehospitalized in M. Djamil Hospital, Padang for 28 days (December 20th 2008 – January 17th 2009). At that time hemoglobin was 7.0 g/dl. Liver function tests were within normal limits. PT and aPTT were also normal. She received whole blood transfusion, octreotide drip, propranolol (10 mg twice daily), ranitidine (20 mg three times daily), tranexamic acid (200 mg three times daily), and omeprazole (20 mg three times daily). Repeat abdominal USG revealed liver cirrhosis and splenomegaly. Endoscopy showed grade II and III esophageal varices. STE was repeated. The patient was subsequently discharged in stable condition.

Discussion

We have presented a case of an 8 ½ year-old girl with hematemesis caused by ruptured esophageal varices due to portal hypertension. The diagnosis was based on history taking, physical examination, laboratory and radiology findings. It was confirmed by endoscopy.

The patient had acute, massive, and recurrent hematemesis, not preceeded by abdominal pain. She had no history of taking erosive medications. Physical examination showed splenomegaly. There were no signs of bleeding on the skin, gums, or other sites, and there was no lymph node enlargement. Therefore, we excluded the diagnosis of upper gastrointestinal (UGI) bleeding caused by hemorrhagic disease. Review of literature suggests that any child with hematemesis and splenomegaly should be presumed to have esophageal variceal bleeding due to portal hypertension until proven otherwise.[1,2,4-6] Two-thirds of patients with portal hypertension hemorrhage before 5 years of age, and 85% do so by 10 years of age.[1]

The patient had slightly abnormal liver function and tested negatively for hepatitis markers. She had no family history of jaundice or liver disease. The abdominal USG and CT-scan revealed liver cirrhosis. Both modalities are considered to be highly sensitive in diagnosing liver cirrhosis. She appeared to have had compensated cirrhosis. Liver biopsy and angiography would have confirmed the diagnoses of liver cirrhosis, and portal vein thrombosis, respectively, but these tests could not be performed due to parents' refusal to consent.

Differential diagnoses of portal hypertension caused by portal vein thrombosis or idiopathic portal hypertension were excluded based on the presence of signs and symptoms of liver cirrhosis. Patients with idiopathic portal hypertension and portal vein thrombosis may be recognized during routine physical examinations if they have splenomegaly in the absence of liver abnormalities.[7]

The patient underwent STE to halt the hemorrhaging of the bleeding varices. She was also given octreotide to stop acute bleeding. The combination of sclerotherapy and octreotide is more effective than sclerotherapy alone.[8] Propranolol

was given to prevent recurrence of bleeding. A combination of STE and drug therapy reduces future variceal bleeding better than drug therapy alone. In patients with high risk esophageal variceal bleeding, endoscopic ligation of the varices is safer and more effective than propranolol for the prevention of future variceal bleeding.[9,10]

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