Esophageal Varices Bleeding in Portal Hypertension due to Portal Vein and Splenic Vein Obstruction

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ABSTRACT

Based on its relation to the liver sinusoid, increased pressure of portal vein can occur at three levels: presinusoid, sinusoid ,and postsinusoid. Obstruction of the presinusoid veins can be caused by extra-hepatic condition such as venous thrombosis.

We reported a case of portal hypertension with esophageal varices bleeding was a result of obstruction due to thrombosis of the splenic vein and portal vein under hypercoagulant conditions due to thrombocytosis. The management of esophageal varices was sclerotherapy while for overcome the thrombosis the patient was given hydroxy urea.

***Key Words**: Esophageal varices bleeding, portal hypertension, vein obstruction*

INTRODUCTION

Portal hypertension is defined as increased portal vein pressure of over 30 cmH_20, often caused by increased pressure on the portal blood flow. Based on its relation to the liver sinusoid, increased pressure can occur at three levels, presinusoid, sinusoid, and postsinusoid. Obstruction of the presinusoid veins can anatomically be caused by extra-hepatic conditions (such as venous thrombosis and increased splenic blood flow), or at the proximal of the liver sinusoid, preventing the liver parenchyme from exposure towards increased venous pressure (such as schistosomiasis). Postsinusoidal obstruction may be caused by the Budd-Chiari syndrome and veno-occlusive diseases. Cirrhosis causes portal hypertension at the sinusoid level.[1,2,3]

Liver cirrhosis is the most common disease that causes portal hypertension. Portal vein obstruction is the second most common cause, which can be idiopathic or may be related to liver cirrhosis, infection, abdominal trauma, congenital malformations of the portal vein, post-surgical conditions, autoimmune diseases, changes in the walls of the portal vein due to inflammation, malignancy, pancreatic disorders, etc.[1,3,4,5,6] Pancreatic disorders may take the form of pancreatitis, carcinoma, and cysts.[3] Pancreatic cysts may take the form of pseudocysts, serous cystadenoma, mucinous cyst neoplasm, and mucinous cystadenocarcinoma.[7] Malignant disorders may cause portal vein thrombosis through direct invasion of the portal vein, external compression of the portal vein, or periportal fibrosis due to surgery or radiotherapy.

Chronic pancreatitis is often accompanied by splenic vein thrombosis.[4]

Venous obstruction may also be caused by thrombosis under hypercoagulant conditions, such as venous policytemia, thrombocytosis, or deficiency of protein C, protein S, or antithrombin III. Thrombocytosis can be primary (in myeloproliferative conditions) or secondary (such as in iron deficiency anemia, chronic inflammatory diseases, post-surgical conditions, malignancy, and so forth). [1,3,6,8]

Clinical manifestations of portal hypertension include abdominal discomfort, gastric and esophageal varices bleeding, splenomegaly, hypersplenism, ascites, and acute and chronic encephalopathy if severe. Under non-cirrhotic conditions, patients with splenomegaly with variceal bleeding should be suspected of splenic vein or portal vein thrombosis. [1,3,8,9] Normal liver biopsy may demonstrate mild changes in portal hypertension due to extra-hepatic portal vein obstruction. [5, 10, 11] Radiologic imaging such as ultra-sonography, Computerized Tomography Scan (CT-scan), Magnetic Resonance Imaging (MRI), angiography, and splenoportography may assist the investigation of the cause of portal vein obstruction. [3, 4, 8]

Treatment is usually aimed at alleviation of the complications of portal hypertension. Bleeding must be controlled. Avoid administration of aspirin. Somatostatin and sclerotherapy may be used.[3,12] If bleeding has been managed, investigate the cause of portal hypertension. If esophageal varices bleeding is caused by portal vein thrombosis due to thrombocytosis, reduction of platelet count by means of cytotoxic drugs is more advisable than surgery.[4] Beta-adrenergic blockers such as propanolol or nadolol, in addition to being capable of reducing portal pressure through a vasodilatation effect on the splancnic arterial flow and the portal vein system, in addition to by reducing cardiac output. Splenectomy is indicated in excessively large spleens, hypersplenism, and obstruction of the splenic vein. [1, 2, 3]

CASE REPORT

Mr. S, 52 years old, middle class, was hospitalized on the fourth floor of the B in-patient ward of Cipto Mangunkusumo Hospital from the 28th of May, 1999, due to complaints of pain on the upper left abdomen since 1 year prior to hospital admission. The pain was sudden in nature, recurrent, did not spread and was so severe that the patient was unable to walk. There was no nausea and vomiting, and there was no fever. The patient defecated as usual. The patient was hospitalized at the Red Cross Hospital, Bogor, for one week and received 3 kinds of medications. The patient was released when he was relieved of his symptoms.

Ten months following admission there was swelling of the upper left abdomen, sometimes accompanied by intense pain, which recurred and did not spread. The patient was readmitted to the Red Cross Hospital.

Three months following the second admission, the patient vomited and defecated black stool. The patient was admitted for the third time to the Red Cross Hospital. He received 10 packages of red transfusion and 4 packs of yellowish transfusion. The patient was advised to be treated at Cipto Mangunkusumo Hospital. The patient denied any history of alcoholism, herb drinking, and smoking. The patient often suffered from generalized weakness.

During physical examination, we found the patient to be moderately ill, fully conscious, with a blood pressure of 110/70 mmHg, a pulse rate of 90 times/minute, a temperature of 37°C, and a respiratory rate of 20 times/minute. His conjunctiva were not pale. His sclera showed no signs of jaundice. His jugular venous pressure was 5-2 cm H_2O. His lungs were sonor, vesicular, without and rales/crackles or wheezing. First and second heart sounds were pure, without any gallop or murmur. His abdomen was not distended. His liver was not palpable. His spleen was palpable at Schuffner III, pliant, smooth, with rounded edges and without tenderness. No mass was palpable. There was no lymph node enlargement. There was no palmar erythema and spider nevi.

Laboratory findings were as follows: Hemoglobin level 11,2 g/dL, Hematocryte 38 vol %, leukocyte count 27.800/mL, platelet count 1.499.000/mL, LED 10 mm/hour. Urine evaluation findings were as follows: specific gravity 1020, pH 6, leukocyte count 4-5/LPB. No erythrocyte was found in the urine. The patient's albumin level was 4,7 g/dL, his globulin level 3,3 g/dL, and total bilirubin 0,5 mg/dL.

Chest x-ray demonstrated a cardiothoracic ratio (CTR) of less than 50%. There was no infiltrates in the lungs.

During admission, the following problems were established: 1. Splenomegaly, 2. Anemia, 3. Thrombocytosis and leukocytosis.

During further evaluation, abdominal ultrasound demonstrated chronic hepatitis with multiple pancreatic cysts and splenomegaly. Abdominal CT-scan without contrast demonstrated mass on the pancreatic head and splenomegaly. Abdominal CT-scan using contrast demonstrated splenomegaly, chronic liver disease, and suspected tu-

mor of the pancreatic head with a differential diagnosis of pancreatitis and carcinoma of the pancreatic head. Blood amylase level was 241 S. Somogryi (Normal: 90-150) and his blood lipase level was 250 U/I (Normal: <190). His CA was 19-9: 31 U/mL (Normal: <35). Doppler ultrasound demonstrated increased portal vein pressure, with a suspicion towards portal vein thrombosis. Liver biopsy demonstrated an image that was in line with chronic hepatitis, with a differential diagnosis of liver without significant disorder. The cause of portal hypertension seem to be infra-hepatic. HBsAg and anti-HCV IgM were negative. His SGOT was 38 U/I, SGPT 23 U/I, cholin-esterase 3,4 kU/I, ureum 40 mg/dL, and creatinine 0,44 mg/dL. Direct splenography demonstrated enlargement of the hillus, and enlargement and winding of the splenic vein1/3 proximal of the spleen,as well as enlargement of the left gastric vein (coronary vein). There was no enlargement of the portal vein. The findings indicated varices of the coronary vein, the 1/3 proximal splenic vein, with obstruction of the portal vein. The patient was then consulted with the Portal Team and was scheduled for splenectomy. On the 13 day of treatment, the patient vomited and defecated blood, suspected to be due to rupture of esophageal varices. Hemostasis evaluation demonstrated compensated DIC, with a Protombin Time of 14,6 (control: 11-14), aPTT of 32,5 (control: 30-40) and positive D-dimer. Endoscopy demonstrated grade II-III esophageal varices of the 1/3 middle to 1/3 distal esophagus, cardiac varices, portal hypertension gastropathy, and mild anthral gastritis. The patient then underwent endoscopic sclerotherapy and underwent seven sessions of sclerotherapy.

Further blood laboratory evaluation demonstrated 13% reticulocyte, with microcytic hypochrom morphology. Iron evaluation was 20, UIBC: 345. Benzidine test turned out negative. Bone marrow hemosiderin was reduced. The patient was suspected for iron deficiency anemia. During bleeding, the patient's hemoglobin level was 8,4. The patient received 250 cc of packed red cell (PRC) transfusion followed by 2x1 tablet of sulfasferosus. The following month, there was an increase of MCV to 70 m and MCH of 23 pg.

Bone marrow puncture demonstrated a hypocellular condition. Bone marrow biopsy demonstrated a hypercellular bone marrow tissue. Platelet physiology demonstrated platelet hyperaggregation. The patient was treated with 2x1 tablet of hydoxyurea, 1 x 300 mg of Allopurinol, and 3 x 500 mg of Bicarbonas natricus. On the eleventh day of treatment, the patient's platelet and leukocyte count dropped, and the dosage of hydroxyurea was adjusted.

DISCUSSION

Absence of cirrhosis during liver biopsy demonstrated that in this case, portal hypertension is a result of obstruction of the splenic vein and the portal vein, due to thrombosis of the splenic vein and portal vein under hypercoagulant conditions due to thrombocytosis. There is also the possibility of obstruction due to fibrosis of the pancreas, infiltrating the walls of the splenic vein, causing thrombus or cystic tumor of the pancreatic head, obstructing the portal vein and the splenic vein. [1,3,4] In this case, further exploration of the cause of obstruction during surgery is required.

In this patient, the problem of anemia was established based on history of generalized weakness, as well as bloody vomiting and defecation. Physical examination demonstrated a pale conjunctiva and a hemoglobin level of 11,2 g/dL. Anemia was though to be due to bleeding and iron deficiency. Iron deficiency anemia may also be the cause of secondary thrombocytosis. However, when treatment with hydroxyurea was terminated, there was an increase in hematocryte with normocytic normochrome erythrocyte morphology and a maintained increase in platelet count. Thus, primary thrombocytosis still cannot be crossed out.[13, 14, 15, 16]

The patient underwent sclerotherapy for esophageal varices, and no recurrent bleeding was found following sclerotherapy. This is in line with literature that stated that a drop in incidence and risk of recurrent bleeding is found in patients with extra-hepatic portal vein obstruction (with maintained liver function) undergoing sclerotherapy.[17, 18, 19]

Shunt construction, possibly without splenectomy, is the definite ideal treatment in patients with extra-hepatic portal hypertension with esophageal varices to avoid unwanted effects such as thrombocytosis, abdominal venous infarction, sensitivity towards infection, and the possibility of more significant bleeding due to loss of collaterals. Nevertheless, emergency conditions or other conditions with large varices, high risk, and difficulty in treatment with sclerosis alone, or absence of a suitable blood vessel for shunting, as in this case, splenectomy is a choice and is not always followed by an adversary outcome. [3, 20]

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