

## Left-Sided Portal Hypertension: A Case Series

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### ABSTRACT

*Left-sided portal hypertension is rarely found, but this condition may cause gastrointestinal tract bleeding and can be life-threatening. The exact incidence of left-sided portal hypertension is unknown as it is rarely found, approximately 1-5%, and most cases were misdiagnosed. We reported 3 cases of left-sided portal hypertension in male patient aged 34 years old, female patient aged 29 years old, and female patient aged 35 years old. Most diagnosis was made based on the clinical findings by excluding the diagnosis of cirrhotic portal hypertension. Splenic vein angiography remains the gold standard in diagnosing left sided portal hypertension. Left-sided portal hypertension is difficult to differentiate from cirrhotic portal hypertension because in these both abnormalities, varices can be present. Left-sided portal hypertension can be considered as a diagnosis in patient with upper gastrointestinal tract bleeding due to oesophageal varices, gastric varices, or portal hypertension gastropathy, accompanied with hypersplenism without the presence of hepatic abnormality or cirrhosis.*

**Keywords:** *left-sided portal hypertension, upper gastrointestinal tract bleeding, hypersplenism*

### ABSTRAK

*Left sided portal hypertension merupakan kasus yang jarang ditemukan tetapi merupakan penyebab perdarahan saluran cerna yang dapat mengancam jiwa. Angka kejadian left sided portal hypertension tidak diketahui secara pasti karena kejadiannya yang sangat jarang, yaitu sekitar 1-5%, dan sebagian besar kasus salah diagnosis. Kami melaporkan 3 kasus left sided portal hypertension pada pasien laki-laki 34 tahun, pasien perempuan 29 tahun, dan 35 tahun. Sebagian besar diagnosis ditegakkan berdasarkan kondisi klinis dengan mengeksklusi diagnosis hipertensi portal sirotik. Angiografi dari vena splenikum tetap menjadi standar baku dalam mendiagnosis left sided portal hypertension. Left sided portal hypertension sulit dibedakan dari hipertensi portal sirotik karena kedua kelainan tersebut dapat ditemukan varises. Left sided portal hypertension dapat dipertimbangkan sebagai diagnosis pada pasien dengan perdarahan saluran cerna bagian atas yang disebabkan karena varises esofagus, varises gaster, atau gastropati hipertensi portal, disertai hipersplenisme tanpa ditemukannya kelainan hepar atau sirosis.*

**Kata kunci:** *left sided portal hypertension, perdarahan saluran cerna bagian atas, hipersplenisme*

## INTRODUCTION

Left-sided portal hypertension or also known as sinistral hypertension is a rare case but may cause life-threatening gastrointestinal tract bleeding. This is usually found as a result of splenic vein obstruction.<sup>1-4</sup>

The incidence of left-sided portal hypertension keeps increasing because clinicians start to notice this term and also due to the advancement of diagnostic technology. Most patients are asymptomatic and did not suffer from complication; thus, the precise incidence is unknown. Left-sided portal hypertension represents 5% from cirrhotic portal hypertension cases. Initially, left-sided portal hypertension case is frequently diagnosed as cirrhotic portal hypertension due to the low incidence rate.<sup>5-10</sup>

In this case series, we discussed 3 interesting cases of patients with left-sided portal hypertension.

## CASE ILLUSTRATION

### Case 1

A 34-year-old male came to the Emergency Department (ED) of Dr. Hasan Sadikin Hospital with the chief complaint of coffee ground vomit with sticky tarry stool since 1 day before hospital admission. Patient had previously been diagnosed to suffer from paroxysmal nocturnal haemoglobinuria (PNH), and received azathioprine, folic acid, and propranolol. Physical examination showed anaemic conjunctiva, dull Traube's space, with no gynecomastia, spider nevi, or palmar erythema. Blood examination revealed Hb 7.5 g/dL, leucocyte 6630/mm<sup>3</sup>, thrombocyte 166000/mm<sup>3</sup>. Serology for hepatitis was negative. Gastroscopy examination showed large varices, starting from the mid to the distal, 3 columns, curved in the oesophagus and large fundal varices in the gaster. Abdominal ultrasound showed the presence of large vein and lineal vein dilatation. Patient was diagnosed with left-sided portal hypertension e.c paroxysmal nocturnal haemoglobinuria with complication of oesophageal and fundal varices rupture. During hospitalisation, patient received pharmacologic therapy of octreotide and packed red cell (PRC) transfusion; bleeding stopped on day-2 hospitalisation.

### Case 2

A 29-year-old female came to the Emergency Department (ED) Dr. Hasan Sadikin Hospital with the chief complaint of coffee ground vomit and black

stool since 1 day before hospital admission. Patient felt that her abdomen was enlarged. Previous history of jaundice was not known. These symptoms had been experienced for 3 times by the patient and she had undergone abdominal ultrasound and endoscopy in 2015. From physical examination revealed anaemic conjunctiva, subicteric sclera, dull Traube's space.

During the previous hospitalisation, abdominal ultrasound was performed, showing hepatosplenomegaly with ascites, multiple cholelithiasis, no dilatation in lineal vein, no enlargement of paraaortic/parailiac lymphadenopathy, no abnormalities in pancreas, bilateral kidney, or urinary bladder.

During hospitalisation, patient was planned for bone marrow aspiration examination, but she declined because was not ready for the procedure. Patient brought results of endoscopy examination performed in 2015 which showed grade III oesophageal varices, mild portal hypertension gastropathy, bile acid reflux. Ligation at 3 sites was performed to the patient. Patient was diagnosed with: (1) Left-sided portal hypertension with complication of hematemesis melena e.c oesophageal varices rupture; (2) Haemolytic anaemia e.c Paroxysmal Nocturnal Haemoglobinuria DD/ Autoimmune Haemolytic Anaemia.

Patient came with Hb of 3.3 g/dL, received 3 times of 2 units PRC transfusion during hospitalisation. During hospitalisation, patient received red blood cells transfusion and octreotide, bleeding stopped on the first day of hospitalisation. Patient received propranolol after discharged from the hospital.

### Case 3

A 35-year-old female came with the chief complaint of feeling weak since 1 month before hospital admission, which worsened since 1 week before hospital admission. Patient felt abdominal enlargement and looked pale. There was no previous history of hepatitis. From physical examination, it was found that patient had anaemic conjunctiva, icteric sclera, and spleen was palpable at schuffner V. There was no spider nevi, palmar erythema, or asterixis.

During hospitalisation, patient underwent endoscopy examination which revealed grade 3 oesophageal varices with moderate portal hypertension gastropathy. Abdominal ultrasound indicated multiple cholecystolithiasis without any signs of cholecystitis, splenomegaly with the possibility signs of portal hypertension, and chronic pancreatitis. Abdominal CT-scan showed the presence of hepatosplenomegaly with well-defined border in large hypodense lesion

in the anterior of the spleen, hepatolithiasis, multiple cholelithiasis with bile duct sludge, ascites. Patient was diagnosed with left-sided portal hypertension, Evans syndrome DD/paroxysmal nocturnal haemoglobinuria. Initial laboratory examination showed Hb 4.6 g/dL; during hospitalisation patient received washed red cell transfusion. Patient received propranolol 3 x 10 mg, oral methylprednisolone 1 x 40 mg, oral folic acid 1 x 5 mg, and oral calcium carbonate 3 x 500 mg after discharged from the hospital.

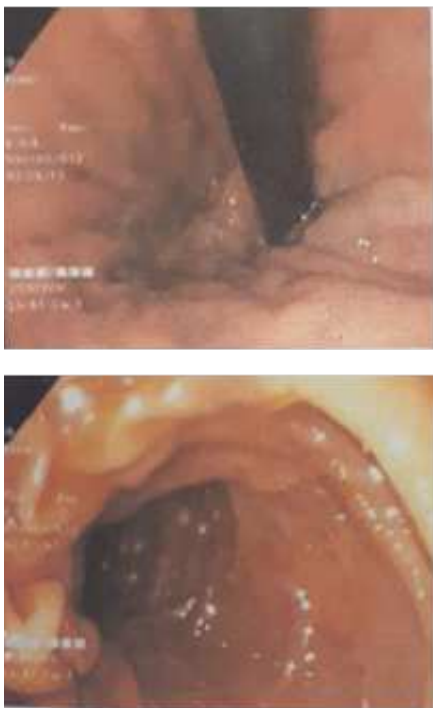


Figure 1. Upper gastroeintestinal endoscopy results

## DISCUSSION

These 3 cases of left-sided portal hypertension happened in male and female in the second and third decade of life. The exact incidence of left-sided portal hypertension is unknown because the case is rare, approximately 1-5%, and most cases were misdiagnosed as cirrhotic portal hypertension. From a study involving 13 patients with left-sided portal hypertension, it was found that the peak incidence was in the fourth decade of life with the age range of 31-68 years old, and more than 50% of patients were male. This is usually caused by thrombus occlusion in splenic vein which may result in lieno (spleno)-gastric varices formation to decompensate the increasing pressure in splenic vein.<sup>2,3,6</sup> Diagnosis of left-sided portal hypertension is usually considered with the presence of 3 conditions, which are upper gastrointestinal tract bleeding, splenomegaly, and normal liver function.<sup>3</sup>

Most diagnosis was made based on clinical findings by excluding the diagnosis of cirrhotic portal hypertension, supporting examinations play important role in confirming the diagnosis in most cases. Splenic vein angiography remains the gold standard in diagnosing left-sided portal hypertension.<sup>3</sup> The most common cause of left-sided portal hypertension include chronic pancreatitis, pseudocyst pancreas, and pancreatic neoplasm. From these 3 cases, 1 of them had chronic pancreatitis found in the abdominal ultrasound examination.<sup>4</sup>

Patient with left-sided portal hypertension frequently is asymptomatic. The most common clinical manifestations are upper or chronic upper gastrointestinal tract bleeding, abdominal pain, or chronic anaemia.<sup>6</sup> From these 3 cases, 2 of them came with the chief complaint of upper gastrointestinal tract bleeding, and 1 of them came with anaemia.

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