

## Case Report

# Esophagoduodenal Varices in Non-cirrhotic Portal Hypertension with Myelodysplastic Syndrome: A Case Report

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**Abstract:**

**Background:** Esophagogastroduodenal varices are dilated submucosal of distal esophageal, gastric, and duodenal veins connecting the portal and systemic circulation. This case report aims to describe a unique case of a child with esophagoduodenal varices due to myelodysplastic syndrome.

**Case:** We reported a case of 3-year-old girl who came to Hasan Sadikin General Hospital on April 3 2022, complaining of black stools 1 time per day for two days before admission. She had previously been diagnosed with esophagogastroduodenal varices since 2019. On initial examination, the patient was fully conscious and appeared pale. The patient's clinical condition improved after adequate treatment of blood transfusion, octreotide, omeprazole and propranolol. However, patient later developed pancytopenia and underwent bone marrow puncture examination which revealed a myelodysplastic syndrome.

**Discussion:** Myelodysplastic syndrome is a condition where ineffective hematopoiesis occurs and can lead to blood malignancy, especially acute myeloblastic leukemia. In this patient, she presented with unequivocal hypertensive gastroesophageal varices, splenomegaly, absence of fibrosis and thrombocytosis supporting subsequent diagnosis of idiopathic non cirrhosis portal hypertension. On the other hand, non-cirrhotic portal hypertension can also be caused by myelodysplastic syndrome as described in this case report.

**Conclusion:** Myeloproliferative malignancies can be a cause of idiopathic non cirrhosis portal hypertension. Pancytopenia often occurs in patients with portal hypertension due to splenomegaly or myelodysplastic syndrome, which can lead to acute myeloblastic leukemia, an example of a myeloproliferative malignancy.

**Keywords:** esophagoduodenal varices, extrahepatic portal vein obstruction, myelodysplastic syndrome

## Introduction

Esophagogastroduodenal varices are dilated submucosal of distal esophageal, gastric, and duodenal veins connecting the portal and systemic circulation.<sup>1</sup> Every minute, 1500 cc of blood circulates through the portal vein throughout the body. This large flow causes an increase in portal vein pressure. Several etiologies cause increased pressure in the portal vein, mostly due to complications from hepatic cirrhosis. Other etiologies unrelated to hepatic cirrhosis are extrahepatic portal vein obstruction (EHPVO), congestive heart failure (CHF), nodular regenerative hyperplasia (NRH), nonalcoholic fatty liver disease (NAFLD), hepatic sinusoidal obstruction syndrome (SOS), metabolic diseases (Gaucher's and Zellweger Syndrome), schistosomiasis, and hepatportal sclerosis.<sup>2,3</sup> Due to this increase in portal vein pressure, the body will respond by forming collateral flows and dilating blood vessels. When the flow is higher, the varicose veins widen and rupture, leading to gastrointestinal bleeding.<sup>1</sup>

The clinical manifestations of non-cirrhotic portal hypertension are similar to those of patients with cirrhosis, namely the presence of collateral circulation, ascites, and splenomegaly in the absence of cirrhotic stigmata.<sup>4</sup> The main complication of this situation is bleeding from ruptured varices, this can be prevented by administering non-selective beta-blockers and varicose ligation per endoscopy.<sup>5</sup> The morbidity and mortality of non-cirrhotic portal hypertension are better than that of patients with liver cirrhosis.<sup>6</sup>

Pancytopenia may occur in patients with portal hypertension due to splenomegaly. Other causes of pancytopenia may result from myelodysplastic syndromes. This situation occurs due to ineffective hematopoiesis and can develop into acute lymphoblastic leukemia.<sup>8</sup>

In this case report, a 3-year-old girl with esophagogastroduodenal varices was reported without signs of hepatic cirrhosis. The patient underwent further examination, namely bone marrow puncture and the results were obtained. On further examination, it was found that the child also had myelodysplasia syndrome. Esophagogastroduodenal varices caused by myelodysplasia are rare and will be discussed in this case.

## Case

A 3-year-old girl came to Hasan Sadikin General Hospital on April 3 2022, complaining of black stools 1 time per day for two days before admission. Complaints accompanied by vomiting of blood 2-3 times since the day before entering the hospital with a volume of half a glass. The patient looked paler and weaker. Patient experience purpura without any complaints of bleeding including nosebleed, bleeding of gums or bruising. There are no complaints of fever. The patient was previously taken to the

local hospital and then referred to our hospital for further examination and management.

Patients have experienced complaints of black stools and vomiting of blood since two years ago. Currently, the patient takes propranolol 5 mg every 24 hours orally. The patient had previously been diagnosed with esophagogastroduodenal varices since 2019 from an endoscopy examination on September 17 2019, with results of grade III-IV esophageal varices, fundal varices, gastroduodenitis, minimal grade II duodenal varices, gastric antral hypoperistalsis, duodenal a/r nodularity. The patient also had a CT angiography examination on October 15 2019, with the suspected results of duodenal varices starting from the superior mesenteric vein to the hepatic portal vein, and there may still be abnormalities in vein formation. Since then, the patient has been receiving propranolol 10 mg every 8 hours orally. The last time the patient had hematemesis and melena was in March 2020.

On initial examination, the patient was fully conscious and appeared pale. The patient weighs 15 kg and is 101 cm tall. The patient's nutritional status was within normal limits. On examination, blood pressure was 90/50 mmHg, respiratory rate 20 times per minute, heart rate 100 times per minute, 97% oxygen saturation in room air, and a temperature of 36.8 Celsius. The conjunctiva looked anemic, the sclera was not icteric. There is no enlargement of the lymph nodes. The liver was palpable 4 cm below the costal arch, and the spleen was palpable as far as Schuffner II. The acral feels warm, looks pale, and the capillary refill time is below 3 seconds. Other examinations are within normal limits. Laboratory examination showed a hemoglobin level of 5.8 mg/dl, hematocrit 21.3%, leukocytes 13,030/mm<sup>3</sup>, and platelets 233,000/mm<sup>3</sup>. **Table 1** showed the laboratory test results from admission to day-9 of admission. **Figure 1** showed endoscopic view of the patient and **Figure 2** showed CT angiography of the patient.

**Table 1.** Laboratory test results

	Admission	Day-2	Day-3	Day-4	Day-6	Day-9
Hemoglobin (mg/dl)	5.8	6.1	8.2	10.5	11.4	9.8
Hematocrit (%)	21.3	22.5	28.3	34.4	36.5	31.3
Leukocytes (mm <sup>3</sup> )	13030	9640	3770	2910	5750	3490
Thrombocyte (mm <sup>3</sup> )	233000	182000	93000	54000	113000	64000
MCV (fl)	57.7	60.2	66.4	69.8	66.7	67.7
MCH (pg)	60.2	16.3	19.2	21.3	20.8	21.2
MCHC (%)	27.2	27.1	29.0	30.5	31.2	31.3

The patient was initially diagnosed with upper gastrointestinal bleeding et causa esophageal rupture with anemia gravis et causa underlying disease. The patient was placed on a nasogastric decompression tube, given a blood transfusion for 250 ml (15 ml/kg body weight), gastric lavage, vasoactive octreotide maintenance therapy of 15 mcg/hour and a proton pump inhibitor. Once the patient could be given oral medication, we discontinued the vasoactive therapy and started beta blockers. The patient received propranolol 10 mg every 8 hours orally, omeprazole 20 mg every 12 hours, and sucralfate 10 cc every 6 hours. The patient's clinical condition improved, marked by the stopped bleeding.

On the second day of treatment, the patient found complaints of paleness without being accompanied by black stools and no blood vomiting. The patient was re-examined after the transfusion, and laboratory results were obtained. Laboratory tests showed a hemoglobin level of 6.1 mg/dl, hematocrit 22.5%, leukocytes 9640/mm<sup>3</sup>, and platelets 182,000/mm<sup>3</sup>. On the third day of treatment, there was a picture of pancytopenia. Laboratory tests showed 8.2 mg/dl hemoglobin levels, hematocrit 28.3%, leukocytes 3770/mm<sup>3</sup>, and platelets 93,000/mm<sup>3</sup>.

Afterwards, the patient was consulted to the hemato-oncology division regarding suspicions of bleeding from other sources and underwent a bone marrow puncture in relation due to complaints of purpura and pancytopenia from laboratory examination on April 12, 2022, according to suspicions towards pancytopenia the bone marrow puncture was performed. The result of bone marrow puncture was myelodysplastic syndrome. The final diagnosis for the patient was esophagogastrroduodenal varices et causa myelodysplastic syndrome.



Figure 1. Endoscopic View

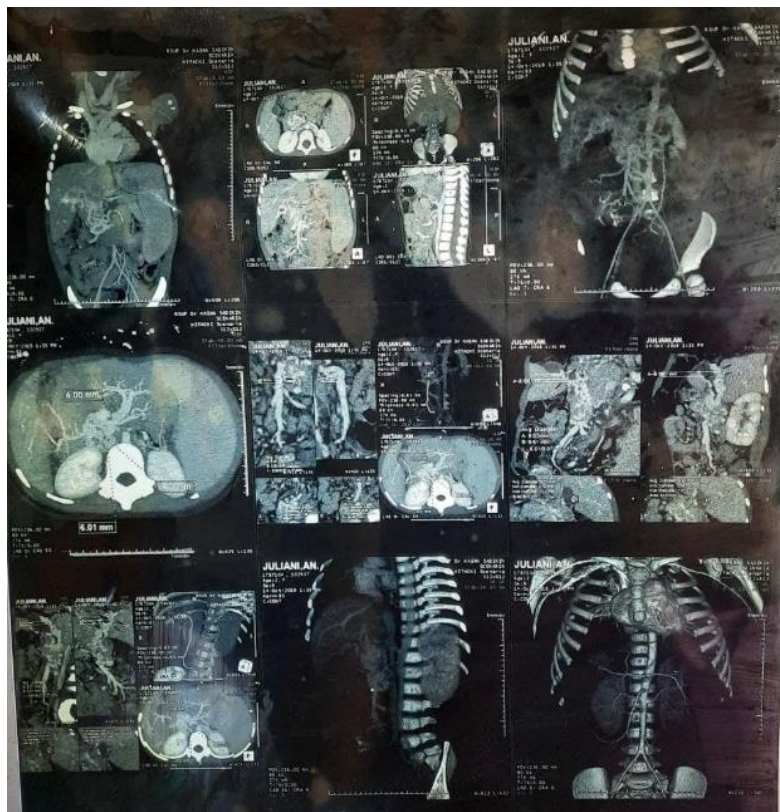


Figure 2. CT angiography of the patient



## Discussion

Non-cirrhotic portal hypertension (NCPH) is a condition of increased portal pressure without signs of cirrhosis. Diagnosis of NCPH is challenging because there is no gold standard for diagnosing this condition. NCPH can occur due to EHPVO (54%), others can occur due to CHF, NRH, NAFLD, SOS, metabolic diseases (Gaucher's and Zellweger Syndrome), schistosomiasis, and hepatoportal sclerosis, or occur idiopathically. Liver biopsy is the main examination for diagnosing patients with suspected NCPH to rule out hepatic fibrosis and cirrhosis.<sup>2,3,4</sup>

Clinical manifestations of NCPH include the presence of collateral circulation, ascites, and splenomegaly without any signs of cirrhotic stigmata. In western countries, the main manifestation of INCPH is splenomegaly and/or increased liver function, whereas in India, most present with gastrointestinal bleeding. The prognosis of patients with NCPH depends on the underlying disease. Complications from increased portal venous pressure must be managed, such as administering diuretics in patients with ascites, administering non-selective beta-blockers, and/or varicose veins ligation.<sup>4,13</sup>

Idiopathic non-cirrhotic portal hypertension (INCPH) is a diagnosis of exclusion if no signs of hepatic cirrhosis are found, and must meet the following 5 criteria, namely: (1) clinical manifestations (splenomegaly, esophageal varices, ascites, increased hepatic vein pressure gradient (PG), or the presence of collateral portal vein flow), (2) no signs of cirrhosis were found on liver biopsy, (3) the absence of chronic liver diseases such as hepatitis B or C, steatohepatitis, autoimmune hepatitis, hereditary hemochromatosis, Wilson's disease, or primary biliary cirrhosis, (4) non-cirrhotic diseases that cause increased portal pressure such as CHF, sarcoidosis, schistosomiasis were not found, (5) portal and hepatic vein patency from Doppler ultrasound or CT scan results.<sup>4</sup> INCPH is more common in Asia than in western countries. Several things can be associated with this event: immune disorders, infections, drugs and toxins (thiopurine derivatives, arsenic, vitamin A), genetic disorders, and prothrombotic disorders (thrombophilia, myeloproliferative malignancy, antiphospholipid syndrome). In this patient, since the bone marrow puncture revealed MDS, we diagnosed the patient as NCPH due to Myelosplastic Syndrome.<sup>7,13</sup>

In this case, apart from NCPH, the patient also experienced pancytopenia. Pancytopenia can be caused by many diseases including malignancy, Evans syndrome, malaria infection and chronic liver disease.<sup>12</sup> Portal hypertension can also cause pancytopenia due to splenomegaly, but in this case, the patient had BMP performed due to myelodysplastic syndrome. Patient was diagnosed with myelodysplastic syndrome after BMP was performed.<sup>8</sup>

Myelodysplastic syndrome (MDS) is a condition where ineffective hematopoiesis occurs and can lead to blood malignancy, especially acute myeloblastic leukemia. In this patient, presence of unequivocal of hypertension gastroesophageal varices, splenomegaly absence of fibrosis and thrombocytosis supporting subsequent diagnosis of INCPH but on the other hand, NCPH is probably caused by MDS.<sup>11</sup> MDS is more common in adults, and cases in children are around 1.8 – 4 cases per 1,000,000 population. MDS in children can occur due to the deletion of chromosome 5q and/or sideroblasts. Children with MDS can be managed by administering a hematopoietic stem cell transplant (HSCT).<sup>9</sup> About 20% of MDS can turn into acute myeloblastic leukemia.<sup>10</sup>

Our weakness in managing this patient is that we have not performed a liver biopsy to rule out liver fibrosis or cirrhosis. It is necessary to carry out periodic follow-ups of patients to prevent varicose rupture and evaluate the possibility of malignancy in patients.

## Conclusion

In conclusion, NCPH is a condition that is rarer than portal hypertension with cirrhosis. Treatment of NCPH is similar to therapy in portal hypertension patients with liver cirrhosis. A histopathological examination needs to be done to rule out liver fibrosis or cirrhosis. Myeloproliferative malignancies can be a cause of INCPH. Pancytopenia often occurs in patients with portal hypertension due to splenomegaly or MDS, which can lead to acute myeloblastic leukemia, an example of a myeloproliferative malignancy.

## Conflict of Interest

None declared

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