An 18-Year-Old Man with Idiopathic Non-cirrhotic Portal Hypertension

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ABSTRAK

Hipertensi portal non-sirosis merupakan salah satu penyebab dari hematemesis dan melena yang cukup jarang terdiagnosa. Seperti pada pasien sirosis hepatis, pada pasien dengan hipertensi portal non-sirosis, hematemesis dan melena terjadi karena ruptur varises esofagus. Namun berbeda dengan sirosis hepatis, pada pasien hipertensi portal non-sirosis tidak ada tanda-tanda gagal hati, yang merupakan ciri khas dari sirosis hepatis. Telah dilaporkan pasien pria dengan hipertensi portal non-sirosis yang mengalami hematemesis akibat ruptur varises esofagus

Kata kunci: *hipertensi portal non-sirosis, hematemesis, ruptur varises esofagus.*

ABSTRACT

Non-Cirrhotic Portal Hypertension (NCPH) is a rare cause of hematemesis and melena. Like in cirrhotic patient, hematemesis in NCPH patient was caused by rupture of esophageal varices. But unlike in cirrhotic patient, in NCPH there are no sign of liver failure, because liver physiology is still normal. We reported case of male patient with NCPH that had hematemesis because of rupture of esophageal varices.

Keywords: *non-cirrhotic portal hypertension, hematemesis, rupture variceal esophagus.*

INTRODUCTION

Non Cirrhotic Portal Hypertension is a disease characterized by periportal fibrosis and involvement of small and medium branches of the portal vein, resulting to intrahepatic portal hypertension in the absence of cirrhosis or other cause of liver disease.[1]

NCPH is more prevalent in Asia than in western country. NCPH related with hygiene and socioeconomy status, worsen status increase prevalence of NCPH. In Asia, male and younger patient were more frequent diagnose had NCPH than in western country.[1-6]

Symptoms of NCPH is caused by increase of portal vein pressure. Splenomegaly, thrombocytopenia, variceal esophagus are some sign of portal hypertension.

Treatment of NCPH is non specific, usually to prevent further complication. The most prevalent complication of NCPH is rupture of esophageal varices.

CASE ILLUSTRATION

A 18-year-young male patient came to Saiful Anwar Hospital with bloody vomiting since 6 days ago reffered from previous small hospital

and got treatment from it but no improvement. He complained bloody vomiting 3-4 times a day with each vomit approximately 250 cc and blacktarry stool approximately 100 cc a day. He felt weak and epigastric pain before vomit and complained his stomach become bigger than before. Last year, he also hospitalized with same complaint and already consumed 10 mg propranolol twice a day, 100 mg spironolactone once daily and 30 cc lactulose syrup three times a day if difficult to passing stool. His grandfather already passed away with liver disease but the family could not mention the name of the disease. History of consumed traditional potion, pain killer drugs, and alcohol were denied. Patient had just graduated from senior high school 1 year ago and because his disease he can not do heavy work.

Physical examination revealed blood pressure 120/60 mmHg, pulse rate 100x/minute, respiratory rate 18x/minute, axillary temperature 36.7°C, BMI 21.4 kg/m², in head and neck examination we found anemic conjunctiva, and there was no alopecia, in chest examination we did not find gynecomastia, spider naevus, heart and lung were normal. Liver span was 10 cm with flat surface, sharp edge, dense, and no pain in pressure, shifting dullness was positive, and traube space dullness, spleen Schuffner I. In his extremities we did not find edema and there was no palmar erythema.

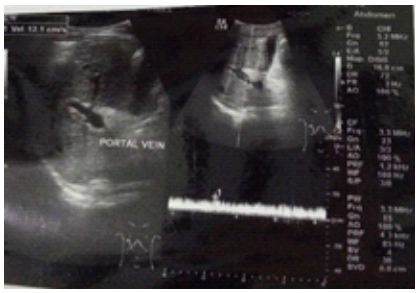

Figure 1. Doppler USG

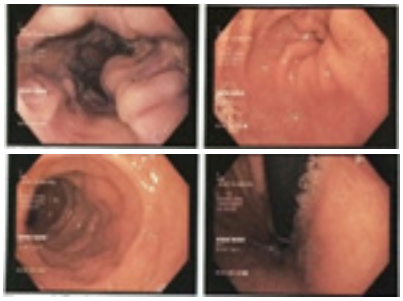

Figure 2. Endoscopy show esophageal varices grade III-IV

Laboratory tests showed hemoglobin levels of 5.6 g/dl, leucocyte count 8.300/µL, thrombocyte count 71,000/µL. His serum albumin levels was 2.26 g/dL with globulin levels of 1.51 g/dL, his liver function, renal function, serum electrolyte, bilirubin, hepatitis B and C serology marker, ANA test, and hemostatic function were within normal range. Chest X-ray within normal range, Abdominal USG revealed splenomegaly and ascites, Doppler USG revealed PSV 12.1 cm/s, portal hypertension but no thrombus in portal vein

Endoscopy showed esophageal varices grade II-III and liver biopsy showed metavir F1 (portal fibrosis without septa) and the ascites fluid analysis reveals transudate with Rivalta test negative.

Patient was diagnosed with Idiopathic Non-Cirrhotic Portal Hypertension. He was done gastric lavage with normal saline every 8 hours until the bleeding stop, 20 drops/minute of IVFD of normal saline (0.9% Nacl solution), PRC transfussion 2 packs/day until Hb> 8g/dL, 50 µg octreotide intravenous continued with 250 µg/hour until 72 hours bleeding stop, 1 gram daily ceftriaxone, 60 mg lansoprazole continued with 6 mg/hour, 10 mg metoclopramide three times daily, and 30 cc lactulose syrup three times daily via nasogatric tube. He suffered from recurrent bleeding until day 10 and finally we decided to do emergency endoscopy and band ligation of esophageal varices and the bleeding was stop, 3 days after bleeding stop and the vital sign were stable, he was go home and prescribed with 10 mg propranolol three times daily, 100 mg

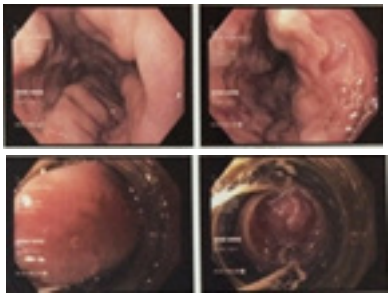

Figure 3. Endoscopy post band ligation

spironolactone once daily, and 30 cc lactulose syrup three times a day. We educate the patient about the compliance to prevent the bleeding occurred.

DISCUSSION

The Asian Pacific Association for the Study of the Liver (APASL) defines INCPH as a disease of uncertain etiology characterized by a periportal fibrosis and involvement of small and medium branches of the portal vein, resulting in the development of portal hypertension.[1]

Diagnosis of INCPH is rendered in patients that have portal hypertension, after excluding portal vein thrombosis, Budd-Chiari syndrome, exposure of toxin and medication, cirrhosis hepatis, chronic viral hepatitis, and other liver disease.

INCPH is commonly reported in developing countries and in lower socioeconomic groups. INCPH is a common cause of portal vein hypertension in Japan and India, about 30-40%, but only 3-4% porta hypertension were caused by INCPH in western hemisphere. However the true prevalence of INCPH might be higher, since patient misdiagnosed as cirrhosis patient. In India and western m INCPH is more frequent in male, while in Japan, female is more frequent. In India age of onset INCPH is younger (25-25 year old) compare in Japan (43-56 year old) and western (median 40 years old).[1,3-5]

No definitive etiology of INCPH. Roughly, the potential mechanisms involved in INCPH pathogenesis can be classified in five main categories: immunological disorders, chronic infections, exposure to medications or toxins, genetic disorders and prothrombotic conditions.[5]

Clinical sign of INCPH is related to hypertension porta without sign of liver failure. Variceal bleeding is the most common sign, but unlike in cirrhotic patient, prognose of variceal bleeding in INCPH is usally good. Ascites is reported in up to 50% of cases, and it usually develops in the context of precipitating factors such as variceal bleeding or infections. Generally, it is easily controlled with low dose of diuretics and resolution of the trigger. Hepatic encephalopathy is a rare complication and it is also related to precipitating factor. Splenomegaly also not uncommon in INCPH.[3]

In our patient, he had hematemesis that related to variceal bleeding (diagnosed with endoscopy) and also splenomegaly and ascites. He also did not show any sign of liver failure, eg; gynaecomastia, palmar erythema and also no edema in his lower extremity.

Diagnosis of INCPH is diagnosis of exclusion, based of following criteria, 1) presence of unequivocal signs of portal hypertension (e.g., gastroesophageal varices, ascites, and/or splenomegaly); 2) absence of cirrhosis, advanced fibrosis or other causes of chronic liver diseases that can cause PH by appropriate serological, biochemical tests and liver biopsy and; 3) absence of thrombosis of the hepatic veins or of the portal vein at imaging studies performed at diagnosis.

Liver function test are normal, jaundice is rarely seen. Anemia, leukopenia and thrombocytopenia are common, because of hypersplenism.

In our patient, liver function test was normal, ratio albumin and globuline normal, there is no hyperbilirubrinemia and also normal coagulation test. He had anemia, maybe related to hematemesis but also because of hypersplenism. He also had thrombocytopenia because of hypersplenism. Marker for chronic viral hepatitis and also ANA test (to exclude autoimmune

Table 1. Diagnostic criteria of idiopathic non-cirrhotic portal hypertension[2]

Clinical sign of portal hypertension	- Splenomegaly - Esophageal varices - Ascites - Portovenous collateral
Exclusion of cirrhosis on,liver biopsy	Must had significant fibrosis
Exclusion of chronic liver disease causing cirrhosis/ Non cirrhotic portal hypertension	- Chronic viral hepatitis B and C - NASH(nonalcoholic Steatohepatitis) - Autoimmune hepatitis - Hereditary hemochromatosis - Wilson's disease - Primary biliary cirrhosis
Exclusion of condition causing non cirrhotic portal hypertension	- Congenital liver fibrosis - Sarcoidosis - Schistosomiasis
Patent portal and hepatic veins	

hepatitis) were normal. These test result showed that patient did not have any sign of liver failure.

Ultrasound is the first imaging modality study, in NCPH abdominal USG show normal liver, but sometime show chronic liver disease sign (nodularity), despite the lack of histological cirrhosis. Splenomegaly and echogenic thick walled portal vein. Contrast enhanced CT scan or MRI also can differentiated between cirrhosis and INCPH. Doppler USG also show increase in hepatic vein pressure gradien, normally 5 mmHg, but in NCPH hepatic vein pressure gradien more than 10 mmHG. From endoscopy we could find esophageal varices.

Our patient's imaging study show chronic liver disease with splenomegaly and ascites, from abdominal USG. It is still possible INCPH patients from USG had chronic liver disease and unrelated to histopathology result. CT scan angiography show hepar cyst segmen 8 and splenomegaly. His endoscopy show esophageal varices grade III-IV.

Liver biopsy is also an important diagnostic tool, to help differentiate between significant fibrosis (cirrhosis) and INCPH. In INCPH, we can find obliteration of small portal vein, and also minimal fibrosis. Our patient histopathology study show metavir F1 (that means fibrosis periportal without septa).

Management of INCPH is non specific, usually focus on management of its complication. The most common complication of INCPH is hematemesis cause by variceal bleeding. Principally management of hematemesis melena in INCPH and in cirrhotic hepatis is same. Vasoactive agents like octreotide or somatostatin used to stop bleeding, proton pump inhibitor (PPI) and ligation of variceal and beta blocker for prevention of rebleeding. In our cases, our patient bleeding could not control only by vasoactive agent (octreotide) and PPI (lanzoprazole), and finally in his 10th day treatment, we decided to do endoscopy emergency to stop the bleeding followed by ligation of variceal esophagus. After endoscopy, patient's bleeding is controlled, and after 13th day treatment he could continue his medication as outpatient clinic.

CONCLUSION

INCPH is not uncommon in Asia, sometimes we misdiagnosed it as cirrhotic patient, although initial treatment for complication is quite similar with cirrhosis patient, INCPH patient's had a better prognoses than in cirrhosis patient. Diagnosis of INCPH, is a diagnosis of exclusion,we must exclude other causes of hypertension portal before we diagnose INCPH. Laboratory finding, imaging study and also histopathology study help us to diagnose INCPH.

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