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Hematemesis as Initial Presentation of Non-Cirrhoticportal Hypertension

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Abstract: ***Idiopathic non-cirrhotic portal hypertension (INCPH) is a rare disease characterized by intrahepatic portal hypertension in the absence of cirrhosis, other causes of liver disease and splanchnic venous thrombosis. We report a case of a18-year-old female who presented to us with a history of pain abdomen from 7 days, fever and generalized weakness from 4-5 days, black coloured stools from 2 days and one-episode of hematemesis on the day of admission. All known causes of hypersplenism were ruled out and she was diagnosed to have idiopathic massive splenomegaly with portal hypertension and hypersplenism.***

Keywords: Hematemesis, Non-cirrhotic Portal hypertension, Hypersplenism, Ascitis, Oesophageal varices, Pancytopenia

1. Introduction

The term idiopathic noncirrhotic portal hypertension (INCPH) has been recently proposed to replace terms, such as hepatoportal sclerosis, idiopathic portal hypertension, incomplete septal cirrhosis, and nodular regenerative hyperplasia, used to describe patients with a hepatic presinusoidal cause of portal hypertension of unknown etiology, characterized by features of portal hypertension (esophageal varices, nonmalignant ascites, porto-venous collaterals), splenomegaly, patent portal, and hepatic veins and no clinical and histological signs of cirrhosis.3 Idiopathic non-cirrhotic portal hypertension (INCPH) is a rare disease characterized of intrahepatic portal hypertension in the absence of cirrhosis or other causes of liver disease and splanchnic venous thrombosis. The etiology of INCPH can be classified in five categories: 1) immunological disorders (i. e. association with common variable immunodeficiency syndrome, connective tissue diseases, Crohn's disease, etc.), 2) chronic infections, 3) exposure to medications or toxins (e. g. azathioprine, 6-thioguanine, arsenic), 4) genetic predisposition (i. e. familial aggregation and association with Adams-Oliver syndrome and Turner disease) and 5) prothrombotic conditions (e. g. inherited thrombophilias, myeloproliferative neoplasm, antiphospholipid syndrome). Roughly, INCPH diagnosis is based on clinical criteria and the formal exclusion of any other causes of portal hypertension. A formal diagnosis is based on the following criteria: 1) presence of unequivocal signs of portal hypertension, 2) absence of cirrhosis, advanced fibrosis or other causes of chronic liver diseases, and 3) absence of thrombosis of the hepatic veins or of the portal vein at imaging. Patients with INCPH usually present with signs or symptoms of portal hypertension such as gastro-esophageal varices, variceal bleeding or splenomegaly.4

2. Case Report

A 18 year old girl, non-alcoholic, student by occupation came to our hospital with chief complaint of hematemesis about 120 ml 6-7 hours back. She was having complaint of Pain Abdomen from 2 weeks, Fever-from 4-5 days, Generalised weakness from 4-5 days and black coloured stools from 2 days. Patient was asymptomatic 2 week back then she started having pain in left flank region which was not associated with food intake or change in position. Pain was low in intensity, non-progressive, non-radiating. Pain was associated with fever and generalized weakness from 4-5 days. Fever was low grade. Patient was having one episode of fever per day. Fever subsided with antipyretics medications. Patient was having black coloured stools from 2 days but no active lower GI bleeding was present. Patient was not having any history of any long-term illness. Patient was not having any history of long-term drug intake. Patient was not having any history of any allergy. Patient was vegetarian by diet, non-alcoholic, non-smoker and was having normal bowel and bladder habits.

On general examination patient was well built and was conscious, oriented to time place and person. Patient BP was 116/76 mmhg, pulse rate-86 b/min which was good in volume and regular. Patient was febrile on touch. Patient temperature was 99.8 degree Fahrenheit. Patient respiratory Rate was 18 per minute. Patient was not having any pallor,

icterus, clubbing, cyanosis, koilonychia and lymphadenopathy. Patient was not having any pedal edema.

On systemic examination patient was not having any abnormality in CVS, CNS and respiratory system. On GIT examination patient was having gross splenomegaly crossing midline which was non tender on palpation. Liver was not palpable. Abdomen was normal in shape. Skin over abdomen was normal. No scars, no dilated veins and no visible pulsations were seen over abdomen. All signs of chronic liver disease were absent. On percussion dullness was present in bilateral flanks. On auscultation bowel sounds were normal.

All routine investigations were done. Patient Hb was 4.5 gm%, TLC were 4000/mm^{3}and Platelets were 50, 000. Patient LFT, RFT, serum ferritin, ESR, LDH, serum copper, serum ceruloplasmin, serum vitamin B12 level, serum folic acid level were within normal range. HIV, hepatitis B and C were non-reactive. Widal test, MP by card test and dengu serology were negative.

On USG whole abdomen liver was Normal in size and echo, pancreas was normal, gall bladder was normal, spleen was22.3 cm in size (enlarged). Splenic infarct was present. Kidneys were normal in size and echo. Mildascites was present.

On CECT Abdomen-Spleen was grossly enlarged in size with prominent spleno-portal axis and shows multiple peri-gastric, peri-pancreatic, peri-portal, peri-esophageal and splenic hilar collaterals were seen-suggestive of Portal Hypertension. Multiple tiny calcified granulomas were seen in splenic parenchyma. Minimal inter-bowel free fluid was seen. Fatty infiltration of liver was seen. Wedge shaped hypodense areas were seen in inferior pole of splenic parenchyma-likely Splenic Infarct.

On USG colour doppler portal vein diameter was 19mm. Splenic artery show normal flow. Splenic Vein show normal flow, no thrombus was seen. Portal vein show normal flow, no thrombus was seen.

On Ascitic Fluid Analysis-Protein were 2.8g/dl, Sugar was 136 mg/dl. Total cells were 270 cells (70%-Mononuclear cells and 30%polymorphs). SAAG was Transudative, ADA was 13 (<25-Normal).

On upper GI endoscopy patient was having large varices (Grade 3) with mild portal gastropathy.

All the causes of portal hypertension were ruled out. Final Diagnosis of Non cirrhotic Portal Hypertension was made.

Patient was started on antibiotics, diuretics, Beta blockers. Whole blood was transfused to improve all cell lines. Endoscopic band ligation was done for esophageal varices. Patient improved well and was discharged on beta blockers and diuretics and splenectomy was advised.

3. Discussion

Massive splenomegaly presenting with hypersplenism, pancytopenia and portal hypertension, without any underlying known cause is known as Banti's syndromeor non-cirrhotic portal hypertension. There are various causes of splenomegaly. When all the known causes of portal hypertension are ruled out, it is termed as idiopathic non cirrhotic portal hypertension.1Idiopathic portal hypertension (IPH) is a disorder that is also known as non-cirrhotic portal fibrosis and hepatoportal sclerosis.2Portal hypertension is characterized by an increase in portal pressure (> 10 mmHg) and could be a result of cirrhosis of the liver or of noncirrhotic diseases. When portal hypertension occurs in the absence of liver cirrhosis, noncirrhotic portal hypertension (NCPH) must be considered. The prognosis of this disease is much better than that of cirrhosis. Noncirrhotic diseases are the common cause of portal hypertension in developing countries, especially in Asia. NCPH is a heterogeneous group of diseases that is due to intrahepatic or extrahepatic etiologies. In general, the lesions in NCPH are vascular in nature and can be classified based on the site of resistance to blood flow. In most cases, these disorders can be explained by endothelial cell lesions, intimal thickening, thrombotic obliterations, or scarring of the intrahepatic portal or hepatic venous circulation. Many different conditions can determine NCPH through the association of these various lesions in various degrees. Many clinical manifestations of NCPH result from the secondary effects of portal hypertension. Patients with NCPH present with upper gastrointestinal bleeding, splenomegaly, ascites after gastrointestinal bleeding, features of hypersplenism, growth retardation, and jaundice due to portal hypertensive biliopathy. Other sequelae include hyperdynamic circulation, pulmonary complications, and other effects of portosystemic collateral circulation like portosystemic encephalopathy. The management of these patients include managing hypersplenism and variceal bleeding. In about 95 % of patient's variceal ligation is enough to cease bleeding. Others need shunt surgery. Variceal recurrence has been seen in 20 % of patients, while recurrent bleeding is seen in 3 % of patients. Surgery is indicated in patients with recurrent variceal bleeding or severe anemia requiring repeated blood transfusions or recurrent splenic infarcts.1

4. Conclusion

Non cirrhotic portal hypertension is a diagnosis of exclusion, after all the other causes of portal hypertension and splenomegaly have been ruled out. It has good outcomes after prompt and appropriate treatment. So, Non-cirrhotic portal hypertension should be kept in mind in case of hypersplenism, especially in young males and should be treated appropriately. Splenomegaly may be the main symptom of IPH without liver cirrhosis. Histopathological assessment of liver biopsy can exclude liver cirrhosis, which has major consequences for treatment. In such patients, splenectomy is usually sufficient, without the need for liver transplantation.

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