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RESEARCH ARTICLE

BANTI'S SYNDROME: AN UNUSUAL CAUSE OF HYPERSPLENISM WITH PANCYTOPENIA: A RARE CASE REPORT

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ABSTRACT

Non cirrhotic portal fibrosis (NCPF) also known as idiopathic portal hypertension (IPH) well also known as Banti`s syndrome is characterized by perivenular fibrosis of the small along with the medium branches of the portal vein ultimately leading to portal hypertension with no change in the structure or function of the liver. As of now the etiology still remains a mystery but postulated to be chronic infections, thrombophilia, immunological disorders, exposure to medications or toxins or genetic abnormalities.The majority of the patients presents with well tolerated episodes of upper gastrointestinal bleed, a long-standing mass or heaviness in the left upper quadrant suggestive of splenomegaly, along with other features of hypersplenism like pancytopenia. Herein we delineate a case of non-cirrhotic portal fibrosis causing pancytopenia in a young male.

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INTRODUCTION

Non cirrhotic portal fibrosis is a disorder of unknown etiology, characterized by fibrosis of small and medium sized branches of the portal vein resulting in portal hypertension. It belongs to the group of vascular conditions of liver wherein the pressure gradient between the portal vein and inferior vena cava is greater than 5mmhg in the absence of cirrhosis[1]. The condition was initially described in 1889 by an Italian pathologist named Banti after which NCPF also came to known as Banti`s syndrome. The increased portal pressure and resistance to splanchnic flow eventually opens the portosystemic collateral circulation and causes congestive splenomegaly. Clinical manifestations of NCPH include variceal bleeding which is typically well tolerated due to preserved liver function. Late stage NCPH may occasionally lead to ascites[2]. Laboratory tests reveal preserved liver function but anemia, leukopenia, and thrombocytopenia i.e. pancytopenia is common due to hypersplenism. Histological findings are nonspecific and comprise a wide range of features from minor changes to obliterative portal-venopathy, incomplete septal fibrosis/cirrhosis and nodular regenerative hyperplasia attributed due to elevated portal venous pressure[3].

Diagnosis requires the exclusion of other causes of liver diseases and portal hypertension. The mandatory diagnostic tests employed are medical history of the patient, blood tests, abdominal imaging, and liver biopsy. The optional diagnostic tests include HVPG, transient elastography (fibroScan) and ARFI[4]. NCPH holds no specific treatment and is based on managing its complications. TIPSS is an effective alternative in patients who fail to respond to medical and endoscopic therapy. Herein we present a case of a 30-year-old male having hematemesis caused by esophageal varices and hypersplenism caused by massive splenomegaly.

CASE REPORT

Male, 30 years old was admitted to our hospital on 21st of May 2024 with chief complaint of recurrent episodes of hematemesis from 15 days. Patient also had fever, bloating, abdominal distension and general weakness. On examination patient was conscious and oriented, vitals were stable maintaining saturation on room air. Pallor was present but no icterus, clubbing or edema was present. Abdominal examination shows abdominal distension with splenomegaly.

Patient had history of similar complaints in the past. Around 7 years back (2017) he suffered recurrent episodes of hematemesis for which he got hospitalized and initially diagnosed with typhoid and malaria. He was treated accordingly and patient was symptomatically better. Ultrasound whole abdomen shows coarse and inhomogeneous echotexture of liver with moderate splenomegaly with dilated portal and splenic vein. UGIE done, in view of persistent episodes of hematemesis, andhe underwent two esophageal bands insertion for variceal bleed. The patient was prescribed medications and was asked for one year follow up. Final diagnosis made was Child-A CLD, with NCPF with splenomegaly with hypersplenism.
However due to covid pandemic he missed follow-up.
Again after 6 years the patient experienced the similar complaints and got admitted in the intensive care unit (ICU) at NIIMS. On presentation CBC reports shows pancytopenia with severe anemia.

Lab	Reports
Hb:	5.1g/dL
HCT:	18.9%
MCV:	75.9fL
MCH:	20.50pg
MCHC:	27g/dL
RDW:	19.8%
Platelet count:	20,000 cells/µL
TLC:	1270 cells/µL

Remarks were mild anisocytosis and microcytic hypochromic anemia with pancytopenia. USG whole abdomen shows coarse liver echotexture with mild surface irregularity with prominent main portal vein with gross splenomegaly ad moderate ascites. Repeat CBC done and he underwent blood transfusion to maintain the blood corpuscles. Meanwhile patient's blood parameters did not improve much and the pancytopenia continued except an improvement in haemoglobin (7.1 gm%) following blood transfusions. With all these findings a diagnosis of Banti's syndrome also known as Non-Cirrhotic Portal fibrosis (NCPF) was made. Liver biopsy not done due to persistent thrombocytopenia. Patient was discharged on propranolol 40 mg to reduce portal pressures with other supportive medications and was kept on regular follow-up. Patient's blood counts remained on lower side; however, there was no episode of bleeding and hence splenectomy was not considered.

The CBC data of pre transfusion and post transfusion is given as follows.

Pre transfusion lab reports	Post transfusion lab reports
Hb: 3.4g/dL HCT: 11.9% MCV: 77.9fL MCH: 22.2pg MCHC: 28.5g/dL RDW: 25.2% Platelet count: 50,000 cells/µL TLC: 920 cells/µL KFT = S. uric acid: 10mg/dL LFT = S. bilirubin direct: 0.84mg/dL Total protein: 5.06g/dL Albumin: 2.66g/dL	Hb: 7.1g/dL HCT: 23.3% MCV: 82fL MCH: 24.9pg MCHC: 30.3g/dL RDW: 20.3% Platelet count: 1,90,000 cells/µL TLC: 1580 cells/µL

DISCUSSION

Banti syndrome is a rare condition also known as noncirrhotic portal fibrosis (NCPF) or idiopathic portal hypertension (IPH)[5]. Banti's syndrome is a rare disorder characterised by chronic congestive enlargement of spleen leading to hypersplenism resulting in blood cell destruction causing pancytopenia due to excessive destruction of blood cells. Common symptoms of Banti's syndrome include weakness, fatigue, anaemia, and splenomegaly. Patient gradually presents with hematemesis and melena, causing anaemia which is microcytic and hypochromic[6]. Complications of portal hypertension dominate the clinical features present in patients with NCPF, the most common being variceal bleeding[7]. Unlike cirrhotic patients, prognosis of variceal bleeding in NCPF is usually good due to the preserved liver function. Banti syndrome can be associated with cirrhosis of the liver which can cause ascites. The major symptoms in patients with Banti's syndrome are GI haemorrhage and splenomegaly, which were found in our patient[8]. Endoscopic sclerotherapy and endoscopic variceal band ligation were performed. Beta-blockers for primary prophylaxis should be given for portal hypertension even in the absence of cirrhosis. Splenectomy may be indicated for recurrent bleeding and severe anaemia requiring multiple blood transfusions[9]. In our case, patient presented with dull aching upper abdominal pain and abdominal distension. On examination, there was severe pallor with hepatosplenomegaly. On routine tests, there was pancytopenia with normal liver function. Bone marrow examination shows hypercellular marrow, negative for immune histochemical staining for lymphoma. Surgery is also indicated for patients with symptomatic hypersplenism, spontaneous bleeding episodes or severe anaemia requiring repeated transfusions or repeated splenic infarcts[10]. Our patient was discharged on beta-blockers to reduce portal pressures and is under regular follow-up. Patient has had no episode of bleeding till now, and thus splenectomy is not being considered.

CONCLUSION

Banti's syndrome is a rare disorder so it is diagnosis of exclusion, after all the other causes of portal hypertension, splenomegaly and anaemia have been ruled out. Patients improve significantly after early and appropriate treatment.

So, Banti's syndrome should be kept in mind in case of hypersplenism, especially in young males and should be treated promptly.

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