Banti's Syndrome Presenting as Hematemesis - A Case Report

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INTRODUCTION

Massive splenomegaly presenting with hypersplenism, pancytopenia and portal hypertension, without any underlying known cause is known as Banti's syndrome. There are various causes of splenomegaly. When all the known causes of portal hypertension are ruled out, it is termed as Banti's syndrome. This syndrome was discovered by Guido Banti in 1882 and is named after him. Banti's syndrome is also known as idiopathic portal hypertension or non-cirrhotic portal fibrosis.[1] Banti's syndrome is commonly found in India and Japan than in the West. [2]

There is absence of any haematologic cause, primary hepatic cause or any tumour or mass lesion involving the spleen. Banti had stated that the primary organ involved was spleen and not the liver leading to secondary splenomegaly. Other features include normal liver function tests, varices seen in endoscopy, cytopenia of one or more cell lines, absence of cirrhosis, patent hepatic veins and elevated portal pressure with multiple collaterals. The complications include rupture of varices and massive bleeding.[3] We report a case of a 20-year-old male who presented to us with a history of fever for 7 days and one-episode of hematemesis on the day of admission. All known causes of hypersplenism were ruled out and he was diagnosed to have idiopathic massive splenomegaly with portal hypertension and hypersplenism.

PRESENTATION OF CASE

A 20-years-old male, non-alcoholic came with complaint of one episode of hematemesis about 150 ml 1 hour back. There was no history of jaundice, diarrhoea, easy bruising, melaena, distension of abdomen, swelling over feet, cough, altered sensorium, chest pain, palpitations, breathlessness, orthopnea or paroxysmal nocturnal dyspnoea (PND). Patient was not a known case of any chronic illness. He did not have similar complaints in the past.

On general examination patient was well built and well oriented. He was afebrile. His pulse was 92 / min, regular, normal volume and character. Blood pressure was 110 / 70 mmHg. There was no sign of pallor, icterus, clubbing, cyanosis, lymphadenopathy and oedema. Jugular venous pressure (JVP) was normal. On abdominal examination massive splenomegaly was present 13 cms below left costal margin, palpable edge beyond umbilicus; (grade 4) (Figure 1). It was non tender. Liver was not palpable. Abdomen was of normal shape and skin over the abdomen was normal with no dilated veins, scars and no visible pulsations. All signs of liver cell failure were absent.

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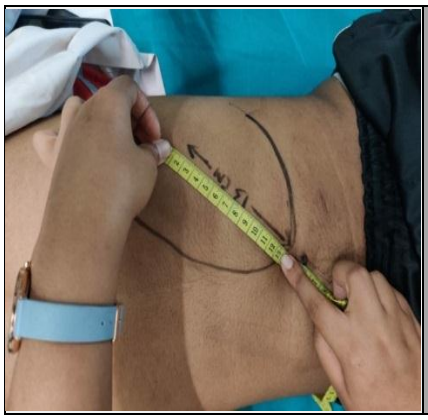

Figure 1. Massive Splenomegaly Extending Beyond the Umbilicus

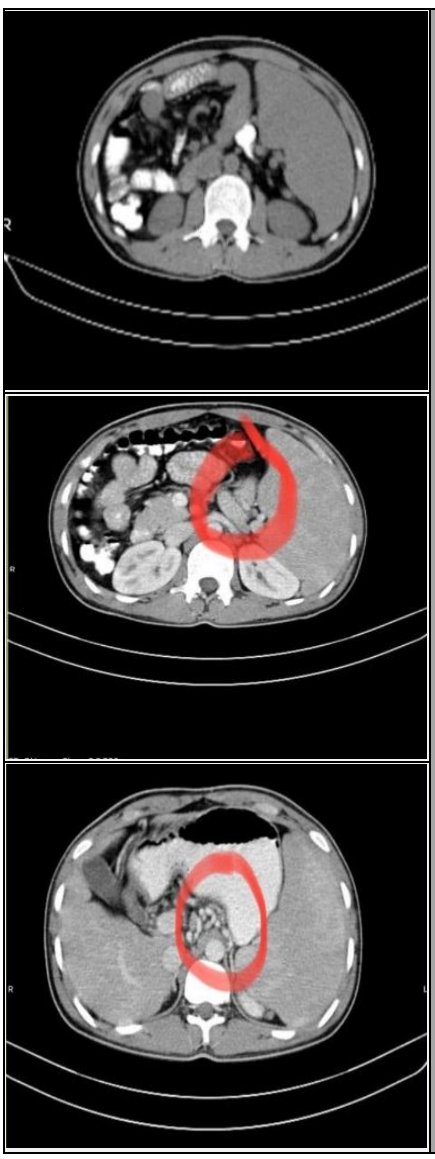

Figure 2. (i) CT Abdomen Showing Massive Splenomegaly (ii) CT Abdomen Showing Dilated Splenic Vein (iii) CT Abdomen Showing Formation of Multiple Collaterals

Examination of all other systems was unremarkable. Complete blood count revealed; haemoglobin (Hb) - 11 gm %, mean corpuscular volume (MCV) was 82, platelet count was 20,000, white blood cell count (WBC) count was 2300. In differential leukocyte count (DLC) lymphocytes were 40 %, neutrophils were 50 %, eosinophils were 5 %, monocytes were 5 % and basophils were 0 %. Peripheral smear showed normocytic normochromic red blood cells (RBCs), platelets reduced on smear, antigen presenting cell (APC) - 80,000 cells / cumm, no hemoparasites seen. IgM for malaria was negative. Kidney function tests and liver function tests were within normal limits. Prothrombin time activated partial thromboplastin time and international normalised ratio (INR) was normal. Urine showed no (RBCs). Lactate dehydrogenase (LDH), erythrocyte sedimentation rate (ESR) and serum ferritin all were within normal limits. Markers for human immunodeficiency virus (HIV), hepatitis B and hepatitis C were negative. Serum copper and serum ceruloplasmin was within normal range.

Ultrasonography of abdomen and pelvis was suggestive of massive splenomegaly measuring 22.8 cm and an isolated dilated splenic vein measuring 9.8 mm. Computed tomography (CT) scan of the abdomen was suggestive of gross splenomegaly measuring 22 cm, and portal hypertension with multiple varices in the left gastric, lower oesophageal and splenic hilar region. (Figure 2. i, ii, iii.)

Upper gastroendoscopy was suggestive of large oesophageal varices, mild portal gastropathy (PGP) and mild gastric antral vascular ectasia (GAVE). Endoscopic band ligation was done for the varices. (Figure 3)

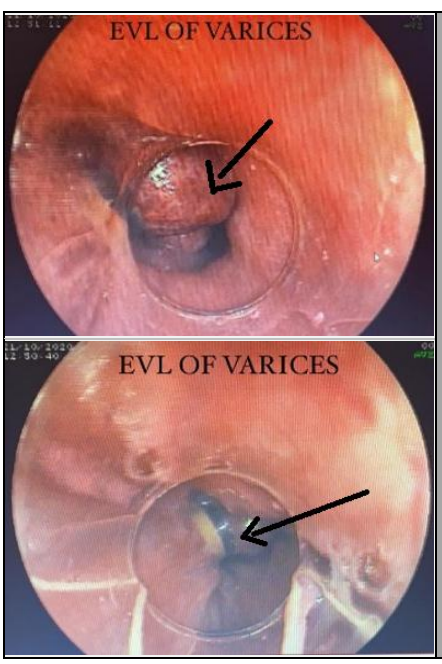

Figure 3. (i) Endoscopic Image Showing a Dilated Oesophageal Vein (ii) Endoscopic Image Showing Ligation of the Oesophageal Varix

DISCUSSION OF MANAGEMENT

Patient was started on beta blocker prophylaxis, tablet propranolol 40 mg OD, for portal hypertension. Meanwhile he was also given supportive treatment and plenty of hydration for his dengue infection. Patient improved symptomatically and was discharged.

DISCUSSION

Banti's syndrome patients present with massive splenomegaly with or without varices. Jaundice rarely develops in these patients. Pancytopenia is due to hypersplenism.[4] Leukopenia and thrombocytopenia is due to peripheral pooling of blood. Bone marrow is hypercellular.[4,5] Complications like hepatic encephalopathy and ascites rarely develop in these patients.

Symptomatic hypersplenism is rare in such patients.[4] Several cases in India, male predominance was found, while in the West and Japan female preponderance is found. Also it affects a more younger group of patients ranging from 25 to 35 years.[4] Various hypothesis are stated such as hepatitis B infections, various intra-abdominal and systemic infections, abnormalities of clotting of blood and also chronic exposure to substances like arsenic causing fibrosis of small portal veins.[6,7] Several genetic and immune mediated mechanisms are also implicated related to rise in T helper1 cells and reduction in cytotoxic / suppressor lymphocytes. Association of HLA-DR has been associated with the immunologic process.[8] Sarin et al. proposed that in genetically predisposed individuals, thrombosis of small and medium branches of the portal vein leads to splenomegaly. The portal and splenic veins are seen as dilated on ultrasonography, and similar finding was seen in our patient.[7] 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.[7] Surgery is indicated in patients with recurrent variceal bleeding or severe anaemia requiring repeated blood transfusions or recurrent splenic infarcts. Prognosis of Banti's syndrome is very good and after successful ligation of varices the 5-year survival is said to be 100 %.[7]

CONCLUSIONS

Banti's syndrome 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, 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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