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Surgical approach towards management of Banti's syndrome: A rare disorder

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Abstract
Banti's Syndrome is characterized by splenomegaly, anemia in absence of any hematological disorder and idiopathic portal hypertension. It is a rare disease in the West, although its incidence in countries like Japan, India and Pakistan is relatively higher.
A 45-year-old male presented with pain in abdomen, generalized weakness, massive splenomegaly, mild hepatomegaly and pancytopenia. Splenectomy was performed and post-operative recovery of the patient was excellent with subsidization of abdominal pain and increase in platelet count, hemoglobin and total leucocytic counts. Histopathology report revealed congestive splenomegaly confirming our diagnosis.
This condition is more commonly seen in socio-economically disadvantaged people. The management of this disease is primarily based on treatment of gastro-intestinal bleeding due to portal hypertension and treatment of hypersplenism, ranging from variceal ligation and endoscopic sclerotherapy, emergency stunts, beta blockers to splenectomy
Banti's syndrome is a rare disorder causing splenomegaly, idiopathic portal hypertension and anemia. Surgical management by performing splenectomy can help improve the general condition of the patient as well as avoid the complications of portal hypertension and splenomegaly.

Keywords: Banti's syndrome, splenomegaly, portal hypertension, pancytopenia, surgical management

Introduction
Banti's Syndrome was described by Guido Banti (Professor of pathological anatomy, Florence, Italy) in 1894 as congestive splenomegaly in absence of any intrahepatic or extrahepatic obstruction [1]. In Guido Banti's words, it is a disease which progresses slowly, but it inexorably drags the patient to death. It is characterized by splenomegaly, anemia in absence of any hematological disorder and idiopathic portal hypertension. Banti's syndrome is also known as non-cirrhotic portal hypertension (NCPH) in India and Idiopathic Portal Hypertension (IPH) in Japan [2]. We report a case of Banti"s Syndrome in a 45 year old male presenting with splenomegaly, pain in abdomen and anemia.

Case report
45-year-old male presented to our OPD for evaluation of long standing dull-aching pain in left side of abdomen. The patient also complained of black tarry stool, easy fatigability, generalized weakness and loss of appetite. The patient did not give any history of fever, urinary complaints or yellowish discoloration of eyes and skin. Family history was not significant and the patient did not give any drug history, or history of alcohol consumption.
On general examination, the patient was afebrile, hemodynamically stable, with a BMI of 17.4. Pallor was present and there was no evidence of lymphadenopathy or icterus. Cardio-respiratory evaluation revealed a systolic flow murmur in the mitral area. Abdomen was distended with massive splenomegaly with splenic edge reaching just below the umbilicus. Liver was palpable 6 cm below the right sub-coastal margin. Laboratory investigation revealed pancytopenia (Hemoglobin 8.30 gm/dl, WBC 2100/cumm with 37% PMN and 58% lymphocytes, Platelet 30,000/cumm). Peripheral blood smear showed aniso-poikilocytosis, few elliptocytes, occasional tear drop cells and hypochromic microcytic anemia. Six serial malaria parasite tests were negative. Liver and kidney function tests were within normal limit and patient had normal coagulation profile. Bone marrow aspiration report revealed few micro-normoblasts with maturation arrest of myeloid and erythroid series cells. Clinical diagnosis of massive splenomegaly with mild hepatomegaly was confirmed on contrast enhanced CT Scan of abdomen showing splenic enlargement measuring 27.6 X 10.2 cm. Splenic vein appeared dilated and tortuous measuring 1.5 cm in diameter. Liver was 17.2 cm in size with normal texture and there was no evidence of liver cirrhosis.

USG Doppler of abdomen revealed multiple collaterals with a hepato-portal flow of 23cm/second. Upper GI endoscosy and colonoscopy revealed normal study.

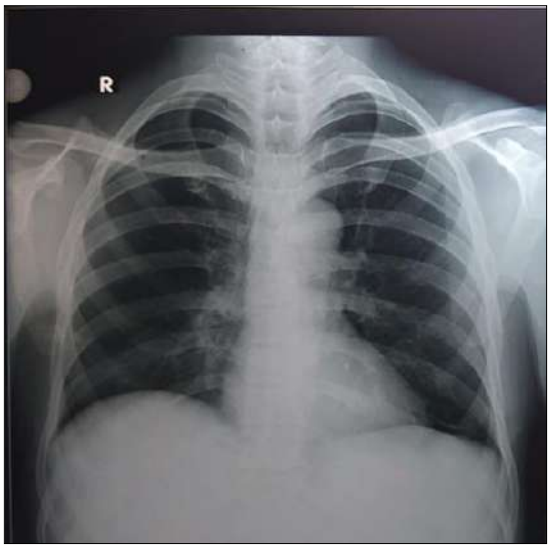

Fig 1: Chest X-ray PA view

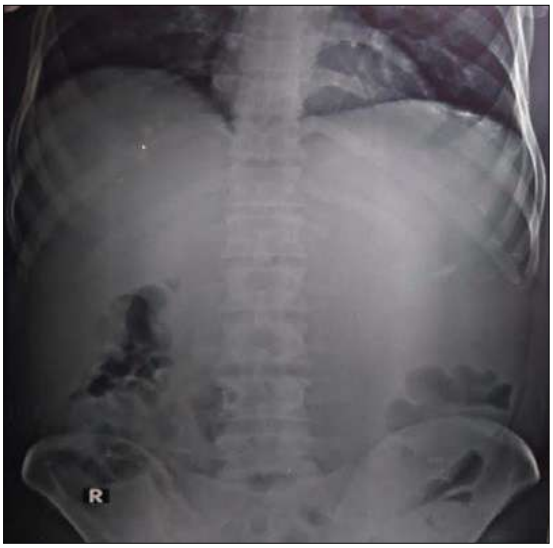

Fig 2: X-ray abdomen (Erect)

Considering the patient's cell lineage, and the risk of traumatic rupture of spleen, the decision of splenectomy was taken. Post-operative recovery of the patient was excellent and histopathology report (HP-13374-19) revealed congestive splenomegaly. There was a rapid increase in platelet level of the patient (pre-operative= 78,000/cumm, post-operative day (POD) 1 = 1,13,000/cumm, POD 3 = 1,79,000/cumm, POD 5 = 1,86,000/cumm, POD 7 = 2,10,000/cumm). The hemoglobin level increased from 8.3gm/dl pre-operatively to 9.7 gm/dl one month after surgery. The patient's general condition improved after surgery.

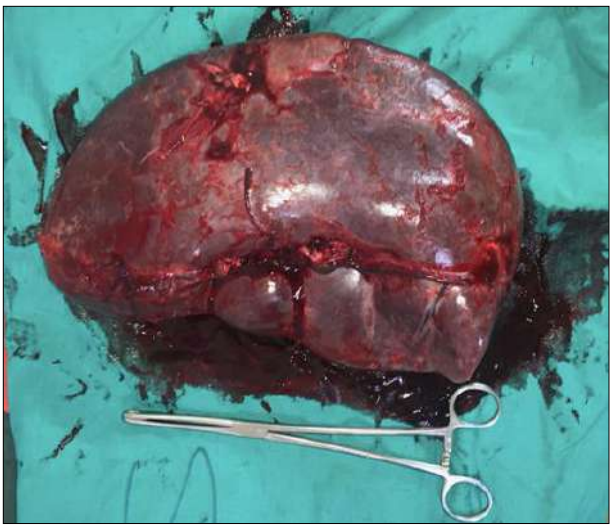

Fig 3: Total splenectomy done with spleen measuring 22 cm in cranio-caudal length and weighing 3.37 kg

Discussion

Banti's syndrome or chronic congestive splenomegaly is characterised by splenomegaly, pancytopenia and portal hypertension [1]. Five general mechanism behind development of splenomegaly [3] are: (i) reactive proliferation of lymphoid cells (ii) infiltration by lipid-laden macrophages or neoplastic cells (iii) extra-medullary haematopoiesis (iv) proliferation of phagocytic cells and (v) vascular congestion. Splenomegaly [4] is caused by one or combination of two or more of the above mechanisms and the various causes of splenomegaly are: infections (malaria, kala-azar, tuberculosis, viral hepatitis, infectious mononucleosis, CMV infection, sub-acute bacterial endocarditis etc.), inflammatory disorders (Felty's syndrome, SLE, serum sickness, rheumatic fever etc.), haematological disorders (thalassemia, sickle cell anaemia, haemolytic anemias etc.), neoplasia (myeloproliferative disorder, leukemia, lymphomas), storage disorders (gaucher's disease, niemann-pick disease), congestive splenomegaly (portal vein obstruction, splenic vein obstruction, Budd-Chiari syndrome, heart failure) or other miscellaneous cause like amyloidosis. Portal hypertension [5] is a term used to describe elevated pressures in the portal venous system, i.e. more than 5mm Hg greater than the inferior vena cava pressure. The cause of portal hypertension [5] could be supra-hepatic (heart failure, Budd-Chiari syndrome, inferior vena cava thrombosis), hepatic (viral hepatitis, liver cirrhosis, alcohol-induced liver disease, cholestatic liver disease), or intra-hepatic (arterio-venous malformation, portal vein thrombosis).

Patients with Banti's syndrome mainly present with long standing mass in left upper quadrant of the abdomen and vague gastro-intestinal complaints [1]. They may also present with complaints pertaining to hypersplenism, or one or more episodes of gastro-oesophageal bleeding resulting from portal hypertension. In vast majority of the patients the liver functions are persevered and hence development of jaundice or hepatic encephalopathy is uncommon. Ascitis is an uncommon finding in patients of Banti's syndrome. Anaemia, leukopenia and thrombocytopenia is a common finding is patients of Banti's syndrome despite normal bone marrow picture [7]. The cause of anaemia could be gastro-intestinal blood loss due portal hypertension or rapid sequestration of RBCs due to hypersplenism. All patients should be thoroughly evaluated for the cause of splenomegaly by various blood investigations, bone marrow evaluation, pathological and radiological investigations [7].

Banti's syndrome is a rare disease in the West, although its incidence in countries like Japan, India and Pakistan is

relatively higher [6]. This condition is more commonly seen in socio-economically disadvantaged people. Few studies done in Indian sub-continent is indicative of a male preponderance, whereas in western countries and Japan, the male: female ratio is equal. Furthermore, it is commonly seen in young adults in the age group of 25-35 years.

The etiology of Banti's syndrome is not well understood and hence characterised to be idiopathic in nature [9]. A number of hypotheses implicate the role of systemic or intra-abdominal infections, chronic exposure to toxic substance like arsenic causing phlebo-sclerosis of small portal vessels, clotting abnormalities and long term intake of immuno-suppressive medications like azathioprine [9]. Numerous hypotheses proposed in favour of immunological cause of Banti's syndrome are supported by reduction in suppressor/cytotoxic T-lymphocytes and predominance of Th1 cells in this disorder. The HLA-DR expression in portal micro-vessels may be an initiating factor leading to immunological assault on portal micro-vessels leading to congestive splenomegaly and portal hypertension [9]. Based on the above theories, Sarin *et al*. concluded that this disorder could develop in genetically predisposed individuals when pro-thrombotic events precipitate repeated micro-thrombotic insults in small and medium sized braches of portal vein [10].

The management of this disease is primarily based on treatment of gastro-intestinal bleeding due to portal hypertension and treatment of hypersplenism [3]. In patients with acute gastro-intestinal bleeding, variceal ligation and endoscopic sclerotherapy have proved to be equally efficacious in 95% of the patients with a recurrence rate of around 20% and 3% recurrent bleeding. Some patients may also require emergency shunt surgeries. Beta-blockers are used as prophylactic drug in patients with portal hypertension. Another drug, ethamolin, is an orphan drug approved by the US Food and Drug Administration in 1988 for treatment of oesophageal varices [11]. Surgical management is indicated in patients with symptomatic hypersplenism, severe anaemia requiring repeated blood transfusion or repeated episodes of splenic infarction. Splenectomy could be done by both open and laparoscopic method [12]. In open method, either a left sub-coastal or midline incision is taken or the spleen is mobilised to the midline by division of lateral and superior pole attachments. The spleno-colic and spleno-renal ligaments are divided at the lower pole. The short gastric vessels are ligated and splenic vessels are isolated. Finally, splenic hilum is held between three clamps and divided. After splenectomy the specimen should be sent for histopathological examination. The histopathological findings are concurrent with congestive splenomegaly [1]. Peri-portal fibrosis is seen with intimal thickening of intra-hepatic portal vein channels. There is obliteration of small portal venules and emergence of new portal veins. During surgery care should be taken to avoid injury to the tail of pancreas and accessory spleen should be searched for. The common complications of splenectomy are bleeding, left lower lobe atelectasis, sub-phrenic abscess formation, thrombosis of the splenic vein and injury to the tail of pancreas, post-splenectomy sepsis, overwhelming post-splenectomy infections, thrombocytosis and splenosis [12]. Vaccination against hemophilus influenza type B, influenza, pneumococcal, meningococcal, tetanus/pertussis/diphtheria is recommended atleast 14 days before elective surgery and 14 days after surgery if not taken pre-operatively.

Post-operative recovery and prognosis is excellent. Our patient underwent splenectomy due to severe anaemia requiring repeated transfusion and massive splenomegaly with risk of traumatic rupture. The post-operative recovery was excellent and there was a rapid increase in platelet level and steady rise in haemoglobin level in the follow up period.

Conclusion

Banti's syndrome is a rare disorder of the Eastern countries, affecting patients in the age group of 20-35 years mainly**.** The key to treatment of Banti's Syndrome is management of pancytopenia, hypersplenism and variceal bleeding. Beta blockers like propranolol is used for the treatment of portal hypertension and prevention of variceal bleeding. Variceal ligation, oesophageal banding and shunt surgeries are also performed for the treatment of gastro-oesophageal bleeding. Haematinic are given for treatment of anaemia. Surgery is considered in patients with features of hypersplenism and anaemia requiring multiple blood transfusions. Post-operative recovery in these patients in excellent.

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