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RESEARCH ARTICLE

A CASE OF BANTI'S SYNDROME IN A YOUNG MALE – AN ATYPICAL PRESENTATION

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ABSTRACT

In 1898 Banti described a disorder characterized by splenomegaly and hypersplenism, resulting in portal hypertension and anemia in the absence of hematological disease. It usually occurs around 3rd to 4th decade of life. It is known as non-cirrhotic portal hypertension (NCPH) in India and Idiopathic Portal Hypertension (IPH) in Japan. Hepatoportal sclerosis seems to be its counterpart in the United States. Banti's syndrome is a disorder of unknown etiology. It has been reported from Indian subcontinent. We report a case of atypical presentation Banti's syndrome in 34-year old man presenting to us with perianal and scrotal swelling. He was subsequently diagnosed to have Banti's syndrome.

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INTRODUCTION

Banti's syndrome is a rare and an eponymous disease first described by Guido Banti, an Italian physician in the year 1898. He described it as a disorder characterized by splenomegaly and hypersplenism, resulting in portal hypertension and anemia in the absence of haematological disease (Banti *et al.,* 1889). It is known by various names globally. It is called non cirrhotic portal fibrosis in India, idiopathic portal hypertension in Japan, hepato-portal sclerosis in USA and Banti's syndrome in Europe (Sarin *et al.,* 2007). Though it is a rare disease, some Indian studies consider one-fifth of the cases of portal hypertension to be non-cirrhotic portal fibrosis implying that many times it is under-diagnosed (Bhargava *et al.,* 1991). The disease occurs worldwide, though it is more common in developing countries. It has been reported from Indian subcontinent (Datta, 1976; Makharia *et al.,* 2001; Qureshi *et al.,* 1991). The age of onset is usually 3rd or 4th decade (Dhiman *et al.,* 2002). We report a case of Banti's syndrome with an unusual presentation.

CASE REPORT

A 34 year old male presented to the emergency department with complaints of Perianal and scrotal swelling since 3 days. This was associated with Fever with chills on and off, foul smell, severe pain and discomfort. He had a history of 1 episode of haematemesis 4 months back for which endoscopy was done and it showed grade 2 oesophageal varices. Patient was asymptomatic with respect to the same and did not follow up subsequently. He also had episodes of loose stools 1 week back, was diagnosed to have acute gastroenteritis and received treatment for the same. On examination, he was well built and had pallor. He was febrile (Temp-102°F), had tachycardia (Pulse rate – 122/min) and Hypotension (BP- 100/60mmHg). Local examination revealed scrotal swelling associated with necrotic skin changes, foul smelling discharge and tenderness. Boggy, tender swelling was present over the left perianal region with marked necrotic skin changes. Abdomen appeared to be distended. No scars, sinuses or dilated veins were seen. It was non- tender on palpation and there was massive splenomegaly with splenic edge reaching just below the umbilicus. His cardiovascular system, respiratory system and central nervous system examination were normal. Local Tissue Ultrasound revealed – diffuse scrotal wall oedema with left sided perianal abscess suggestive of infective etiology. All relevant blood investigations were sent and the patient was posted for emergency incision and drainage of perianal abscess with scrotal exploration.

Laboratory data revealed the following:

Hb: 9.2 gm/dL,
RBC: 3.2 x 10^6 / mm^3
WBC: 21600mm^3 (Neutrophils-83%, Eosinophils-1%, Lymphocytes-12% Monocytes-4%)
Platelet count: 30000/mm^3
Peripheral blood smear: RBCs - mild anisocytosis with normocytic normochromic anaemia. WBCs – Polymorphonuclear leucocytosis with toxic changes. Platelets-showed marked reduction with few giant platelets.

Patient could not be taken up for emergency surgery due to his low platelet count and was admitted to the ICU. The patient received i.v. antibiotics and blood transfusion (Random Donor Platelets, Fresh Frozen Plasma and Packed Cell Volume) and meanwhile other investigations were sent to look into the cause of severe thrombocytopenia. His coagulation profile was normal, as were his liver and renal function tests. Routine work up for fever was done and was negative for Malaria, Dengue, Enteric fever. Coomb's test and Autoimmune profile were negative. His hepatitis serology was negative. The patient was taken up for surgery the next day. Platelet counts of the patient continued to remain in the same range and he continued to receive platelet transfusions post surgery. He was advised bone marrow studies but he did not consent for the same. Blood culture reports showed no growth. Pus culture reports showed Aerobic growth and *E.coli* was isolated. Antibiotics were started in accordance with the culture sensitivity report. Patient developed abdominal distension, dyspnoea, hypotension on the 5th post-operative day. Emergency ultrasound showed Subacute intestinal obstruction with gross splenomegaly and peritoneal collection with multiple moving echoes. Patient was taken up for a laparotomy. Intra-operatively we found extensive adhesions, haemoperitoneum due to spontaneous splenic rupture. He underwent splenectomy with adhesiolysis. In due course, the patient gradually improved symptomatically. His vitals stabilised, counts became near normal. The patient was discharged after prophylactic pneumococcal and meningococcal vaccination and was advised to follow up for regular dressings for the scrotal and perianal wounds.

DISCUSSION

Banti's syndrome is a rare disease in the West, although its incidence in other countries, such as Japan and India is relatively higher (Oikawa *et al.,* 1998). The condition has been commonly seen in people who are socioeconomically disadvantaged, both in India and Iran (Sarin and Kapoor, 2002; Vakili *et al.,* 1992). Improved hygiene and standards of living could explain the relative rarity of the disease in the West and its declining incidence in Japan, indirectly suggesting a role of infection (Banti, 1889). Few studies done in the Indian subcontinent indicate a male predominance in contrast to the West and Japan, where the disease is more common in females. Furthermore it affects a much younger age group of patients, varying from 25 to 35 years (Sarin *et al.,* 2002). Our patient was a 34 year male and belonged to a weaker socioeconomic section. Banti's syndrome or Non cirrhotic portal fibrosis remains a disease of unknown etiology and a number of hypotheses have been proposed implicating the role of systemic or intra-abdominal infections, relationship with Hepatitis B virus, clotting abnormalities and chronic exposure to toxic substances such as arsenic leading to phlebosclerosis of small portal vessels (Sarin *et al.,* 2002; Orozco *et al.,* 1991). Immunologic and immunogenetic hypotheses have also been proposed, supported by reduction in suppressor/cytotoxic T lymphocytes and predominant Th1 cells seen in this disorder (Tokushige *et al.,* 2000). The HLA-DR expression in portal microvessels may be an initiating factor leading to immunologic assault on portal microvessels (Terada *et al.,* 1991). One of the main theories proposed is infection of the gut. Recurrent infection of the gastrointestinal tract by bacteria can lead to portal vein thrombosis due to septic embolization. This mural thrombus of portal vein leads to stellate cell activation. As stellate cells are known to cause fibrosis, they may lead to perisinusoidal fibrosis (Marra *et al.,* 1995). Our patient had an episode of acute gastroenteritis and it may have been a possible etiological factor. The other theory put forward especially in India is chronic arsenic poisoning due to contaminated drinking water (Datta *et al.,* 1979). In the study done by Datta et al, the liver biopsy of people with chronic arsenic poisoning showed periportal fibrosis and portal collaterals. This possibility is rare in our case as chronic arsenic poisoning occurs more commonly in farmers who are exposed to pesticides (Guha Mazumder, 2008). Another theory being proposed is that this may be an immunologically mediated disease as IgG, IgM, IgE and IgA are significantly elevated in this condition (Guha Mazumder, 1986). Some studies find cytotoxic lymphocytes to be significantly decreased in Banti's syndrome (Nayyar *et al.,* 1990). It is also found that tumour necrosis factor is increased in Banti's syndrome. Tumour necrosis factor is one of the factors implicated in liver damage. As in non cirrhotic portal hypertension there can be periportal fibrosis, tumour necrosis factor is implicated in it. Vascular cell adhesion molecule-1 (VCAM-1) is activated by tumour necrosis factor. The elevated levels of VCAM-1 suggest a possible immune aetiology (Yamaguchi *et al.,* 1999). Based on the above theories, Sarin *et al.* concluded that the disorder could develop in a genetically predisposed individual when prothrombotic events precipitated repeated microthrombotic insults in the small and medium branches of the portal vein. The main histopathologic findings are periportal fibrosis, intimal thickening of intrahepatic portal venous channels, obliteration of small portal venules and emergence of new aberrant portal venous channels. Although these abnormalities could be secondary to portal hypertension, it has been proposed that the vascular changes are the primary event that leads to portal hypertension (Dhiman *et al.,* 2002).

Patients with Banti's syndrome may present with a long-standing mass in the left upper quandrant due to splenomegaly and consequences of hypersplenism or they may present with one or more well- tolerated episodes of gastrointestinal variceal bleeds resulting from portal hypertension (Sarin *et al.,* 2002). Development of jaundice, hepatic encephalopathy and ascites is uncommon and the liver function tests are preserved in majority of patients (Dhiman *et al.,* 2002; Sarin *et al.,* 2002). Ascites is only a rare finding in non cirrhotic portal fibrosis (Sarin *et al.,* 2007). Anemia in these patients may be microcytic, hypochromic due to gastrointestinal blood loss or normocytic, normochromic due to hypersplenism (Sarin *et al.,* 2002). Our patient had normocytic normochromic anemia and ascites. He also had a history of bleeding oesophageal varices in the past. Anemia in this patient could be thus be multifactorial, with blood loss and nutritional deficiency compounding hypersplenism. Thrombocytopenia (<50,000/mm3) may occur in these patients on account of increase in plasma volume and splanchnic pooling of the blood while the bone marrow is hypercellular. Hypersplenism usually causes leukopenia and thrombocytopenia (Dhiman *et al.,* 2002). Our patient on the contrary had leucocytosis which was secondary to septic foci in the form of perianal and scrotal abscesses. Severe thrombocytopenia leading to easy bruising, history of acute gastroenteritis and poor local hygiene together may be implicated in the formation of these abscesses. Patients with non cirrhotic portal hypertension have thrombosed portal vein occasionally accompanied with collateral circulation (Dooley *et al.,* 2011). On ultrasonography, the portosplenic axis is seen as dilated and patent, as was the case with our patient. Hepatic venography and radionuclide scintigraphy

have been used to distinguish between this entity and cirrhosis (Qureshi *et al.,* 1991; Sarin *et al.,* 2002). The key management issues in patients with Banti's are gastrointestinal haemorrhage and hypersplenism. For acutely bleeding patients, endoscopic sclerotherapy and variceal ligation are equally efficacious in 95% of patients and very few need to undergo emergency shunt surgery. Beta-blockers are efficacious in primary prophylaxis even in non-cirrhotic patients with portal hypertension. Surgery is indicated for patients with symptomatic hypersplenism i.e. spontaneous bleeding episodes or severe anemia requiring transfusion or repeated splenic infarcts. Splenic embolization may be attempted for hypersplenism. Prognosis for patients with Banti's syndrome is excellent. Even in patients with acute variceal bleeds, the mortality is significantly lower than seen in cirrhotic patients. After successful eradication of such gastro-oesophageal varices, the 5-year survival is reported to be as high as 100%. (Sarin *et al.,* 2002).

Conclusion

Our case highlights the fact that this condition is often under-diagnosed. It is imperative to diagnose the condition early and differentiate it from portal hypertension due to cirrhosis, as the management differs and once treated, the condition has excellent prognosis.

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