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Banti's Syndrome: Case Report and Review of Literature

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Introduction

In 1898 Banti described a disorder characterized by splenomegaly and hypersplenism, resulting in portal hypertension and anemia in the absence of hematological disease. 1 Banti's syndrome is also 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. 2,3 Banti's syndrome is a disorder of unknown etiology, clinically characterized by portal hypertension (varices and portosystemic collateral vessels), splenomegaly, and anemia (hypersplenism). 3 It has been reported from Indian subcontinent. 4-6 In a Pakistani case series of portal hypertension, 18 out of 37 patients were found to have IPH as the etiology. 6 We report a case of Banti's syndrome in an 20-year old girl presenting to us with anemia and splenomegaly.

Case Report

A 20 year old female was referred to us for the evaluation of symptomatic anemia. She also complained of longstanding dull, dragging left sided abdominal pain. Review of systems revealed menorrhagia. On examination she was a lean, pale-looking girl. There was no lymphadenopathy or icterus. She was afebrile, her vitals were stable and cardiovascular examination revealed a systolic flow murmur in the mitral area. Her liver was not palpable, however, she had massive splenomegaly with the splenic edge reaching just below the umbilicus. Laboratory data revealed pancytopenia (Hb=8.4 gm/dl, WBC=2200/mm³ with 67% PMN and 22% lymphocytes, Platelet = 35,000/mm³) and peripheral blood film showed anisocytosis and hypochromic microcytic anemia. Her coagulation profile was normal, as were her liver function tests. Anemia workup revealed iron deficiency and five serial malarial parasite tests were negative. Coomb's test and autoimmune profile were negative and hypercoagulable states were ruled out. Her hepatitis serology was negative and she had normal [(0)] ceruloplasmin and alpha-1 antitrypsin levels. Her bone marrow revealed a cellular specimen showing all hematopoietic cell lines and massive splenomegaly was confirmed on CT Abdomen, which demonstrated a splenic size of 21.7 cm in cranio-caudal extent. Moreover, it showed large varices in the splenic hilum, gastro-splenic and gastro-pancreatic ligaments and cavernous transformation of the portal vein, indicative of portal hypertension (Figure 1). Her liver size and texture was normal on CT scan and there was no evidence of cirrhosis (Figure 2). Ultrasound Color Doppler demonstrated numerous patent collaterals resulting from this cavernous transformation, with a hepatoportal flow velocity of 24 cm/sec. Esophagogastroduodenoscopy and sigmoidoscopy were normal. A presumptive diagnosis of Banti's syndrome was made on the basis of pancytopenia, splenomegaly and non cirrhotic portal hypertension. She was put on iron and folate supplements. In addition, low dose propranolol was also started, to reduce portal

pressures. Splenectomy was not considered as she has no evidence of active bleeding. Her blood counts improved on supportive medical management.

Discussion

Patients with Banti's syndrome may present with a long-standing mass in the left upper quadrant due to splenomegaly and consequences of hypersplenism, as did our patient, or they may present with one or more well-tolerated episodes of gastrointestinal variceal bleeds resulting from portal hypertension. ⁷ Development of jaundice, hepatic encephalopathy and ascites is uncommon and the liver function tests are preserved in majority of patients. ^{7,8} Anemia in these patients may be microcytic, hypochromic due to gastrointestinal blood loss or normocytic, normochromic due to hypersplenism. ⁷ Our patient had microcytic hypochromic anemia despite normal endoscopy. Thalassemia was ruled out by serum protein electrophoresis. Anemia in this patient could be multifactorial, with menorrhagic blood loss and nutritional deficiency compounding hypersplenism. Leukopenia ($<4000/\text{mm}^3$) and thrombocytopenia ($<50,000/\text{mm}^3$) may occur in these patients on account of increase in plasma volume and splanchnic pooling of the blood while the bone marrow is hypercellular. Symptomatic hypersplenism is rather rare and necessitates intervention. ⁷ Banti's syndrome is a rare disease in the West, although its incidence in other countries, such as Japan and India is relatively higher. ⁹ The condition has been commonly seen in people who are socioeconomically disadvantaged, both in India and Iran. ^{7,10} 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. ¹¹ The 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. ⁷ The etiology of Banti's syndrome is not well-understood. 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. ^{7,12} Immunologic and immunogenetic hypotheses have also been proposed, supported by reduction in suppressor/cytotoxic T lymphocytes and predominant Th1 cells seen in this disorder. ^{7,13} The HLA-DR expression in portal microvessels may be an initiating factor leading to immunologic assault on portal microvessels. ¹⁴ 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. ⁸ The intrasplenic and portal vein pressures are markedly elevated in patients with Banti's syndrome, as are splenic and portal vein blood flow, suggestive of hyperdynamic circulatory state. 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. 6,7 CT portography and CT hepatic angiography have also been used to demonstrate the aberrant vessels in these patients. The key management issues in patients with Banti's are gastrointestinal hemorrhage 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. Variceal recurrence has been reported in approximately 20% of patients, though only 3% of these bleed recurrently. Beta-blockers are efficacious in primary prophylaxis even in non-cirrhotic patients with portal hypertension. 7 Surgery is also indicated for patients with symptomatic hypersplenism -spontaneous bleeding episodes or severe anemia requiring transfusion or repeated splenic infarcts. Prognosis for patients with Banti's disease is excellent. Even in patients with acute variceal bleeds, the mortality is significantly lower than seen in cirrhotic patients. After successful eradication of such gastroesophageal varices, the 5-year survival is reported to be as high as 100%. 7

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