Case report

Banti's syndrome in a young male

Dr. Harish Rajaram[1], Dr Balasubramanian Rajaraman[2]

[1]Post graduate, Department of Internal Medicine, SMVMCH, Puducherry

[2] Associate Professor, Department of Internal Medicine, SMVMCH, Puducherry

Name of the Institute/college: Sri Manakula Vinayagar Medical College and Hospital

Corresponding author: Dr Harish Rajaram

Date of submission: 08 October 2014; Date of Publication: 10 December 2014

Abstract:

Banti's syndrome is a rare disease which is characterised by ascites, splenomegaly and portal hypertension without coexisting cirrhosis of liver. There is no geographical predilection for the disease. It is usually occurs around 3rd to 4th decade of life. Here we report a case of a 16 year old boy who presented with ascites, massive splenomegaly and pancytopenia. CECT abdomen revealed thrombosed portal radicals with normal liver. He was subsequently diagnosed to have Banti's syndrome. Through this case we deduce that Banti's syndrome must be considered in cases with portal hypertension though they may be of younger age group.

Key words: Banti's syndrome, portal vein thrombosis, non-cirrhotic portal fibrosis

Introduction:

Banti's syndrome is characterized by raised portal venous pressure due to either intrahepatic or prehepatic lesion which is accompanied by normal liver pathology. Here the characteristic picture is of portal hypertension in the absence of cirrhosis. It is known by various names globally. It is called non cirrhotic portal fibrosis in India, idiopathic portal hypertension in Japan, hepatoportal sclerosis in USA and Banti's syndrome in Europe.[1] It is an eponymous disease as it was first described by Guido Banti, an Italian physician in the year 1898. He described it as a disease with splenomegaly, accompanied by portal hypertension and anaemia with normal liver pathology. 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 underdiagnosed.[2]The age of onset is usually 3rd or 4th decade.[3] The disease occurs worldwide, though it is more common in developing countries. Its incidence is higher in people with lower socioeconomic status. Here we report a case of Banti's syndrome with unusually young age of onset.

Case details:

A 16 year old boy, who is a high school student, presented with fever, loose stools and vomiting for 3 days duration. He had continuous, high grade fever, which was not associated with chills and rigors. He had no history of cough and sputum. He also had loose stools for 3 days, with 4 episodes per day previously. It was semisolid in consistency. He also had vomiting for 3 days with one episode per day previously. It was non bilious and non projectile. Vomitus contained food particles.

He had past history of one episode of haematemesis 6 months back for which endoscopy done showed Grade III oesophageal varices. Endoscopic sclerotherapy was done for the same. Patient was asymptomatic and did not follow up subsequently.

On examination, he was febrile. His pulse rate was 104/mt. His blood pressure was 100/60 mmHg. He was pale.

On inspection his abdomen appeared distended and flanks appeared full. Umbilicus was slit shaped, appeared to be in midline. No Scars, sinuses or dilated veins were seen. On palpation there was no tenderness. Spleen was palpable. It was 15 cm below the left coastal margin just crossing the midline towards the left iliac fossa. On percussion of abdomen, shifting dullness was present. There was a dull note over Traube's space.

His cardiovascular system, respiratory system and central nervous system examination were normal.

Routine investigations were done. Complete haemogram showed pancytopenia. Peripheral smear confirmed the pancytopenia. The laboratory values are shown in Table 1.

Table 1 – Lab parameters of the patient

INVESTIGATION	REPORT
Red Blood Cell Count	3.3 x 10^6 mm^3
Hemoglobin	6.4 g/dl
Total Leukocyte Count	1100 cells / mm^3
Neutrophil	35
Lymphocyte	54
Monocyte	9
Eosinophil	2
Basophil	0
Platelet Count	51,000 / mm^3

Routine workup was done for fever. His reports came as negative for enteric, malaria and dengue fever. His blood culture was positive Staphylococcus aureus growth. His urine culture showed no growth of microorganisms. His liver function tests were normal. His renal function tests and electrolytes were within normal limits.

Thus he was admitted for fever with pancytopenia, with ascites and splenomegaly. Ascitic fluid analysis was done. Ascitic fluid sugar was within normal limits. SAAG ratio (Serum Ascites Albumin Gradient) was 2.7 indicating the ascites was due to portal hypertension. Ascitic fluid adenosine deaminase level was normal ruling out tuberculosis. No growth of microorganisms was seen in ascetic fluid culture.

Chest X ray showed blunting of right costophrenic angle indicating minimal right sided pleural effusion. There was no abnormality in ECG.

Abdominal ultrasound revealed normal liver, with splenomegaly and ascites. It also revealed mild luminal irregularities in portal vein. And right sided pleural effusion was present.

CECT of abdomen was done. It revealed that the portal radicals were thin and thrombosed and there was cavernous transformation surrounding it. Liver was normal. Hepatic veins were normal. Esophagus-stomach junction and lower oesophagus showed varices. Spleen was enlarged and spanned 20 cm. Splenic vein was dilated. Splenic vein was not continuous with SMV-MPV (superior mesenteric vein – main portal vein junction), but was draining through collaterals. Mesenteric and

www.ijbamr.com P ISSN: 2250-284X , E ISSN : 2250-2858

omental lymphadenopathy was present.(Fig 1 and 2). Subsequent tests were done to rule out hypercoagulable conditions. Therefore a diagnosis of Banti's syndrome was made.

He was started on amikacin based on blood culture and sensitivity report. He was also started on paracetamol, anti-emetics and anti-diarrhoeal agents. Platelet transfusion was done, when his platelet count fell below 30,000/mm^3 on day 4 of admission.

Tablet propronalol was started prophylactically for oesophageal varices. He gradually improved symptomatically and as his blood counts became near normal, he was referred for surgical management of varices and hypersplenism.

Figure - 1 CECT abdomen transverse section showing portal vein thrombosis (orange arrow) and mesenteric lympadenitis (blue arrow).

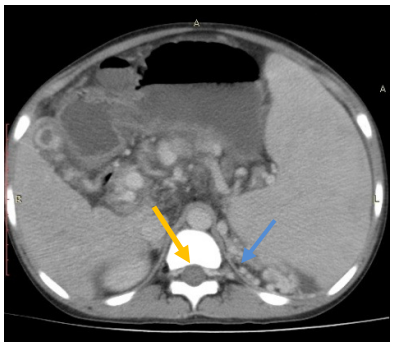
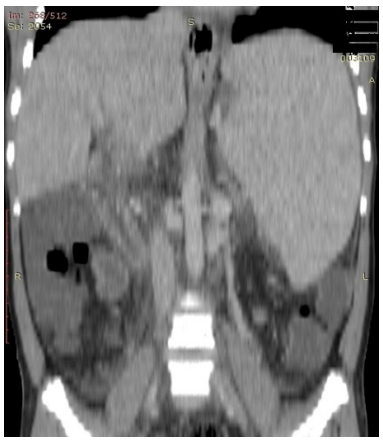


Figure 2 – CECT abdomen coronal section showing massive splenomegaly.



Discussion:

Non cirrhotic portal fibrosis remains a disease of unknown aetiology. Even so, there are many theories proposed for aetiopathogenesis of the disease. It appears to be more common in lower socioeconomic status group. One of the main theories 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.[4] In this case it may have been a possible aetiology as he had acute gastroenteritis.

The other theory put forward especially in India is chronic arsenic poisoning due to contaminated drinking water.[5]In the study done by Datta et al, the liver biopsy of people with chronic arsenic poisoning showed periportal fibrosis. The study also revealed portal collaterals. In our case, chronic arsenic poisoning may have been a possibility due to the similar abdominal CT findings. Even so, it is less likely considering the age of the patient and occupation, as chronic arsenic poisoning occurs more commonly in farmers who are exposed to pesticides.[6]

It other theory being proposed is that this may be immunologically mediated disease. As IgG, IgM, IgE and IgA are significantly elevated in this condition.[7] Some studies find cytotoxic lymphocytes to be significantly decreased in Banti's syndrome.[8] 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.[9]

www.ijbamr.com P ISSN: 2250-284X , E ISSN : 2250-2858

The patients usually present with gastrointestinal haemorrhage or splenomegaly. They usually have anemia. Out of the many causes of portal hypertension, a massive spleen is most commonly seen in Banti's syndrome. They may have features of hypersplenism. Ascites is only a rare finding in non cirrhotic portal fibrosis.[1]Our case had similar presentation. Our patient's initial presentation was upper gastrointestinal bleed. On current admission he presented with massive splenomegaly with pancytopenia. But he also had ascites along with the above manifestations. Features like spider nevi, palmar erythema and gynecomastia, which are features of chronic liver cell failure were absent as is the case with Banti's syndrome.[1]

The liver function tests are normal in non cirrhotic portal fibrosis. Pancytopenia is found in majority of individuals. Anemia may be microcytic hypochromic or normocytic normochromic anemia. Hypersplenism usually causes leukopenia and thrombocytopenia.[3]Similar picture was present in our case. Patients with non cirrhotic portal hypertension have thrombosed portal vein occasionally accompanied with collateral circulation.[10]Our patient's CT abdomen also revealed thrombosis of portal radicals with collateral circulation with no abnormality in liver.

The management of this disease is aimed at controlling the active variceal bleed. Endoscopic variceal ligation or sclerotherapy must be done immediately to control the variceal bleed, as was done in this case. But if there is no active variceal bleed drugs such as beta blockers can be administered prophylactically. Recurrent bleed may require shunt surgeries. Splenic embolization may be attempted for hypersplenism.[11] Non cirrhotic portal fibrosis has very good prognosis. The mortality from variceal bleed in this condition is lower than in cirrhosis.[11]

Conclusion:

This case highlights that though this disease usually occurs in 3rd or 4th decade, it must also be kept as a differential diagnosis in younger age groups too. This condition is often underdiagnosed. 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.

References:

1. Sarin SK, Kumar A, Chawla YK, Baijal SS, Dhiman RK, Jafri W, et al. Noncirrhotic portal fibrosis/idiopathic portal hypertension: APASL recommendations for diagnosis and treatment. Hepatol Int. 2007 Sep;1(3):398–413.

2. Bhargava DK, Dasarathy S, Sundaram KR, Ahuja RK. Efficacy of endoscopic sclerotherapy on long-term management of oesophageal varices: a comparative study of results in patients with cirrhosis of the liver, non-cirrhotic portal fibrosis (NCPF) and extrahepatic portal venous obstruction (EHO). J Gastroenterol Hepatol. 1991 Oct;6(5):471–5.

3. Dhiman RK, Chawla Y, Vasishta RK, Kakkar N, Dilawari JB, Trehan MS, et al. Non-cirrhotic portal fibrosis (idiopathic portal hypertension): experience with 151 patients and a review of the literature. J Gastroenterol Hepatol. 2002 Jan;17(1):6–16.

4. Marra F, Grandaliano G, Valente AJ, Abboud HE. Thrombin stimulates proliferation of liver fat-storing cells and expression of monocyte chemotactic protein-1: potential role in liver injury. Hepatol Baltim Md. 1995 Sep;22(3):780–7.

5. Datta DV, Mitra SK, Chhuttani PN, Chakravarti RN. Chronic oral arsenic intoxication as a

possible aetiological factor in idiopathic portal hypertension (non-cirrhotic portal fibrosis) in India. Gut. 1979 May;20(5):378–84.

6. Guha Mazumder DN. Chronic arsenic toxicity & human health. Indian J Med Res. 2008 Oct;128(4):436–47.

7. Guha Mazumder DN, Ghose S, Das K, Ghosh A, Ghose KK, Nag S. Immunological studies in cirrhotic& non-cirrhotic portal fibrosis. Indian J Med Res. 1986 Jul;84:59–61.

8. Nayyar AK, Sharma BK, Sarin SK, Malhotra P, Broor SL, Sachdev G. Characterization of peripheral blood lymphocytes in patients with non-cirrhotic portal fibrosis: a comparison with cirrhotics and healthy controls. J Gastroenterol Hepatol. 1990 Oct;5(5):554–9.

9. Yamaguchi N, Tokushige K, Haruta I, Yamauchi K, Hayashi N. Analysis of adhesion molecules in patients with idiopathic portal hypertension. J Gastroenterol Hepatol. 1999 Apr;14(4):364–9.

10.Dooley JS, Lok A, Burroughs AK, Heathcote J, editors. Sherlock's diseases of the liver and biliary system. 12th ed. Chichester, West Sussex ; Hoboken, NJ: Wiley-Blackwell; 2011.

11. Sarin SK, Kapoor D. Non-cirrhotic portal fibrosis: current concepts and management. J Gastroenterol Hepatol. 2002 May;17(5):526–34.