CASE REPORT

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Banti's Syndrome: An Unusual Cause Of Pancytopenia

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Abstract

Banti's syndrome is also known as Idiopathic Portal Hypertension (IPH) or Non-cirrhotic Portal Fibrosis (NCPF). NCPF/IPH is a disease of uncertain aetiology characterised by periportal fibrosis and involvement of small and medium branches of the portal vein, resulting in the development of portal hypertension. It is a rare cause of pancytopenia and is usually a diagnosis of exclusion. Here we present a case of a middle aged female with Banti's syndrome.

Key words: Pancytopenia, hepatosplenomegaly, non-cirrhotic portal fibrosis.

Case report

A 41-year-old female resident of Uttaranchal presented with dull, aching, upper abdominal pain since 15 days which was not associated with food intake, nausea or vomiting.

On general physical examination, patient was thin built with severe pallor, vitals were stable except tachycardia. On systemic examination, there was an ejection systolic murmur in the mitral area along with mild hepatosplenomegaly. (A soft, non-tender liver was palpable 1 cm below right costal margin with a liver span of 17 cm and a non-tender palpable spleen of a similar consistency which was 5 cm below left costal margin).

Routine blood tests revealed a pancytopenia (Haemoglobin - 4.3 gm%, total leucocyte count: 1.2 x 10^9/l, platelet count: 1.2 x 10^6/l), liver function was normal except raised gamma glutamyl transferase (GGT-251IU/L).Ultrasound whole abdomen was also consistent with clinical findings. In view of low haemoglobin, patient was transfused 2 units of packed red blood cells (PRBCs).

With the above findings, our initial differentials were chronic infections like chronic malaria, Kala azar, viral aetiology, tuberculosis; chronic liver disease; lymphoma, sarcoidosis, and vasculitis. However, malarial antigen, peripheral smear for malaria parasite, *Leishmania donovani* bodies, viral markers for hepatitis B, C and retrovirus turned out to be negative. Work-up for chronic liver disease, autoimmune hepatitis work-up, vasculitis work-up was also normal. Angiotensin converting enzyme (ACE) and total and ionized serum calcium levels were within normal limits. CECT chest and abdomen revealed additional finding of a dilated portal vein (Fig. 1) and was not suggestive of tuberculosis or sarcoidosis. In view of CECT finding, opinion from gastroenterology team was taken. They suggested for USG Doppler splenoportal axis which revealed a normal flow direction in splenoportal axis with prominent superior mesenteric and splenic vein. Peak systolic portal velocity was low (14 cm/sec). UGIE was also done which showed only diffuse gastritis with no variceal formation.

Besides UGIE, a bone marrow examination was done which revealed a hypercellular marrow, negative for immune-histochemical staining for lymphoma. Thus, it did not any give any clue of the diagnosis. Meanwhile patient's blood parameters did not improve much and the pancytopenia continued except an improvement in haemoglobin (8.4 gm%) following blood transfusions. Therefore, patient was planned for a liver biopsy for a conclusive diagnosis. Biopsy revealed dilated central vein, hepatic venules and aberrant portal venous channels with no amyloid deposits (Fig. 2). With all these findings a diagnosis of Banti's syndrome also known as Non-Cirrhotic Portal fibrosis (NCPF) was made.

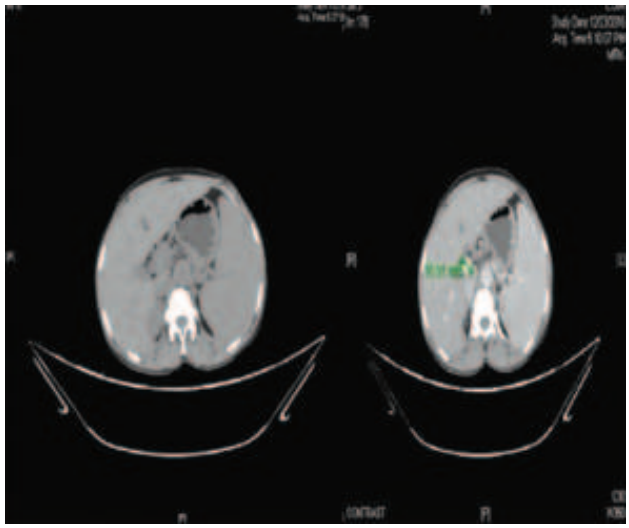

Fig. 1:

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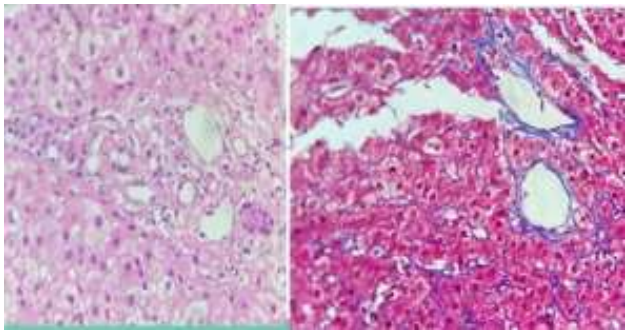

Fig. 2:

Patient was discharged on propronolol 20 mg to reduce portal pressures and was kept on regular follow-up. Patient's blood counts remained on lower side; however, there was no episode of bleeding and hence splenectomy was not considered.

Discussion

Banti's syndrome is a rare condition also known as non-cirrhotic portal fibrosis (NCPF) or idiopathic portal hypertension (IPH). A study conducted between 1980 - 1990 in India by Sarin *et al* revealed that only 98 such cases had been reported and over time incidence has further declined[1]. However, there are no national registry data of patients with NCPF and there has been no nationwide study to determine the trend in the incidence of NCPF.

As per the Asia-Pacific Association for the Study of the Liver (APASL), the definition of NCPF is a disease of uncertain aetiology characterised by periportal fibrosis and involvement of small and medium branches of the portal vein, resulting in the development of portal hypertension[1].

Complications of portal hypertension dominate the clinical features present in patients with NCPF, the most common being variceal bleeding[2,3,4,5]. Unlike cirrhotic patients, prognosis of variceal bleeding in NCPF is usually good due to the preserved liver function.

Ascites is also reported in 50% cases and it usually develops in the context of precipitating factors such as variceal bleeding or infections. Besides this, portal vein thrombosis can also be seen (13 - 46%)[6]. Jaundice, hepatic encephalopathy, awareness of lump in left upper quadrant are rare presentations.

In our case, patient presented with dull aching upper abdominal pain and patient had no awareness of her lump. On examination, there was severe pallor with hepatosplenomegaly. On routine tests, there was pancytopenia with a normal liver function (except raised GGT) and normal coagulation profile.

In 2008, APASL defined the diagnostic criteria of Banti's syndrome on needle liver biopsy are as follows[1]:

1. Absence of regenerative nodules with features of possible or definite cirrhosis in an adequate-sized liver biopsy.
2. Features usually seen (however, these may not be seen in all): Small portal vein obliteration; aberrant vasculature; portal tract fibrosis, rounded or streaky; absence of significant hepatocellular injury.

In our case, liver biopsy was done which showed dilated central vein, hepatic venules and aberrant portal venous channels with no amyloid deposits. UGIE was also done which showed only diffuse gastritis with no variceal formation. The key management issues in patients with Banti's syndrome are gastrointestinal haemorrhage and hypersplenism. Data on management and prophylaxis of variceal bleeding in NCPF patients are scarce with a remarkable lack of randomised controlled trials. Nevertheless, expert opinion recommends following the guidelines of prophylaxis and management of portal hypertension in cirrhotic patients[2]. 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 anaemia requiring repeated transfusions or repeated splenic infarcts[1]. Our patient was discharged on beta-blockers to reduce portal pressures and is under regular follow-up. Patient has had no episode of bleeding till now, and thus splenectomy is not being considered.

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