Idiopathic Portal Hypertension: A Case Report of an Adult Indian Rural Female of “Banti Syndrome”

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Abstract

Non-cirrhotic portal fibrosis (NCPF)/idiopathic portal fibrotic hypertension (IPH) is one of the major diseases that include a group of diseases caused by increased portal venous pressure which is thought to be due to intrahepatic or prehepatic lesions in the absence of liver cirrhosis. In the present case a 35-year-old female presented with abdominal fullness and pain in the upper abdomen for the past two months which was not associated with the consumption of water or food, nausea, or vomiting. When we investigated it was seen that she had severe anemia with pancytopenia with no malarial parasite on peripheral smear, liver renal and liver function tests were within the normal range. The diagnosis of Banti’s syndrome was made. However, the patient was then vitally stable and was started on propranolol and was advised for splenectomy due to refractory anemia. The present study provides easy and conventional diagnostic techniques for clinical diagnosis and proves the radiological technique.

Introduction

NCPF/IPH is one of the major diseases that include a group of diseases caused by increased portal venous pressure which is thought to be due to intrahepatic or prehepatic lesions in the absence of liver cirrhosis. In the Indian subcontinent, in most of the literature this is called NCPF, while in Japan and other Asian countries, it is called idiopathic portal hypertension. In other parts of the Western world, this problem is addressed differently, such as
portal sclerosis [1], noncirrhotic intrahepatic portal hypertension, and idiopathic noncirrhotic intrahepatic portal vein hypertension [2]. NCPF incidents are reported worldwide; However, their incidence is higher in developing countries than in developed countries. The major regional differences in these diseases are not clear but appear to predict the impact that may be explained by differences in socioeconomic status, living conditions, life expectancy, and ethnicity. NCPF and IPH are often found in poor social groups such as the rural population of India [3]. It's possible that NCPF's standing in India has declined recently. It has been suggested that neonatal omphalitis and/or recurrent diarrhea in infants or early childhood may be the culprits of these diseases. Changes in pregnancy practices in India have led to a decrease in the incidence of omphalitis.

**Case presentation**

A 35-year-old female resident of the rural area of Osmanabad, Maharashtra presented with abdominal fullness and pain in the upper abdomen for the past 2 months which was not associated with consumption of food or water, nausea, or vomiting. There was no history of fever, any past hospitalization, or any co-morbidities. On general physical examination, the patient was thin-built and had severe pallor, with another vital parameter normal except for tachycardia. On systemic examination, auscultatory findings were of an ejection systolic murmur in the mitral area along with massive splenomegaly, the spleen was non-tender surface was smooth which was approximately 26 cm below the left costal margin crossing the umbilicus- Hackett grade 4.

Routine blood investigations revealed pancytopenia (Hemoglobin - 5.3 gm%, total leucocyte count: 1.5 x 10⁹/l, platelet count: 1.4 x 10⁹/l), with no malarial parasite on peripheral smear, liver and renal function tests were within the normal. Activated partial thromboplastin time, prothrombin time, and international normalized ratio (INR) were normal. The urine routine examination was normal. Ultrasound entire abdomen was also consistent with clinical findings. Two units of packed red blood cells (PRBCs) were transfused due to severe anemia. With this presentation and findings, our initial differential diagnosis was of chronic parasitic infection like Kala-azar, HIV, tuberculosis, lymphoma, sarcoidosis, and vasculitis.

However, Leishmania donovani antibodies and retrovirus turned out to be negative. Markers for view of vasculitis were also normal. Total and ionized serum calcium and angiotensin-converting enzyme (ACE) as a marker of sarcoidosis were within normal limits. The CECT chest was normal but the CECT of the abdomen revealed a dilated portal vein [Figure: 1, 2 and 3] and there were no findings of tuberculosis or sarcoidosis. In sonographic Doppler scan of the spleno-portal axis showed a normal flow direction in the splenoportal axis with prominent superior mesenteric and splenic vein and there was a decrease in the peak systolic portal velocity measuring 12 cm/sec. There was no evidence of esophageal varices on UGIscopy. USG abdomen revealed mild hepatomegaly and normal liver echotexture.
Table 1: Clinical investigations

<table>
<thead>
<tr>
<th>Investigation</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Complete blood count</td>
<td>Haemoglobin - 5.3 gm%, total leucocyte count: 1.5 x 10^9 /l, platelet count: 1.4 x 10^6 /l, mean corpuscular volume 81.3, Retic count 2.5, no malarial parasite on peripheral smear.</td>
</tr>
<tr>
<td>Liver function tests</td>
<td>SGOT-40 IU/L, SGPT-30 IU/L, Total Bilirubin 1.0mg/dl, Serum Albumin 3.8 g/dl, PT/INR 13sec/ 1.1</td>
</tr>
<tr>
<td>HIV, HCV and HbsAg</td>
<td>Negative</td>
</tr>
<tr>
<td>C- ANCA, P- ANCA</td>
<td>Negative</td>
</tr>
<tr>
<td>Serum ACE</td>
<td>28 mcg/L</td>
</tr>
<tr>
<td>Serum Calcium</td>
<td>8.5 mg/L</td>
</tr>
<tr>
<td>UGIscopy</td>
<td>Within normal limits, no esophageal varices were seen.</td>
</tr>
<tr>
<td>USG abdomen</td>
<td>Mild hepatomegaly and normal liver echotexture, massive splenomegaly.</td>
</tr>
<tr>
<td>USG portal vein Doppler</td>
<td>Normal flow direction in splenoportal axis with prominent superior mesenteric and splenic veins. Decrease in the peak systolic portal velocity measuring 12 cm/sec..</td>
</tr>
<tr>
<td>Serum arsenic levels</td>
<td>Normal</td>
</tr>
<tr>
<td>ANA Blot</td>
<td>Negative</td>
</tr>
</tbody>
</table>
Figure 1: CECT abdomen (coronal view without contrast) showing significantly dilated and tortuous extrahepatic portal vein and Porto-splenic confluence, mildly enlarged liver with fatty infiltration and with over distended gall bladder, gross splenomegaly with homogenous texture, multiple collaterals in lienorenal, left subdiaphragmatic and peri gastric regions; with no evidence of any changes of cirrhosis.

Figure 2: CECT abdomen shows spleen grossly enlarged around 28.48 cm along the maximum long axis.
With the consecutive PRBC transfusions her anemia settled. With all these above-discussed findings the patient was diagnosed with Banti’s syndrome which is also known as NCPF. However, the patient was then vitally stable and was started on propranolol and was advised for splenectomy.

Discussion

A persistent congestive enlargement of the spleen that causes hypersplenism and pancytopenia is described as Banti’s syndrome. As a result, patients often exhibit generalised weakness, exhaustion, decreased effort tolerance from anaemia, frequent infections from neutropenia, easy bruising from thrombocytopenia, fullness in the belly, and early satiety from splenomegaly. With time as the disease progresses, there is a possibility of hematemesis and melena, which is the cause for the worsening of anemia and making its symptoms more profound. Thereby the patient also sometimes can present with complications of portal hypertension, the most common being variceal bleeding [4]. The prognosis of variceal bleeding in NCPF is usually in comparison with cirrhotic patients as the liver function test parameters are not altered.

The etiology of Banti’s syndrome is multifactorial possibly is seen as follows:

• Infection: This is one of the possible etiologies which is proposed to be due to the recurrent bacterial infection from the gut causing repeated septic embolization in the portal circulation, this was documented in the literature by Boyer [5] and Wanless [6] as their studies suggested that IPH and NCPF are caused by the thrombosis of large intrahepatic portal veins.

• Xenobiotics: It is seen that chronic exposure to certain xenobiotics is thought to predispose individuals to the development of NCPF. Amongst all the suspicion of inorganic arsenic to be a major cause has been investigated to the greatest extent. It is seen that the drinking of arsenic-contaminated water is a suspected cause of NCPF, according
to data from Chandigarh, India, and also subsequently from Eastern parts of India [7].

- Immunological anomalies: There is also evidence supporting immunologic abnormalities to be one of the causes of this syndrome. The total T cells and cytotoxic T cells population was decreased, and tumour necrosis factor (TNF) activity is increased in this syndrome which turns out to be a cause involved in fibrosis, as there is evidence that increased soluble TNF receptors I and II found in this syndrome and this explains the fibrosis around the portal vein. Some studies also reveal that the TNF makes upregulation of vascular cell adhesion molecule-1 (VCAM-1), hence there is an increase in the VCAM-1 in these patients.

The important management of patients with Banti’s syndrome is gastrointestinal hemorrhage and most importantly hypersplenism. Literature on the management and prophylaxis of variceal bleeding in patients of NCPF is minimal due to a lack of research trials on this entity. It’s seen that beta-blockers are efficacious in primary prophylaxis even in people with non-cirrhotic portal hypertension [8].

Surgical management can also be considered in patients with symptomatic hypersplenism, recurrent hematemesis, or severe anemia requiring repeated transfusions or having repeated splenic infarcts. In patients with Non-cirrhotic portal hypertension (NCPH), surgery is often indicated for the prevention of variceal hemorrhage (i.e., secondary prophylaxis). [9, 10]. Endoscopic therapy alone may not address issues like growth retardation, hypersplenism, portal biliopathy, or recurrent pain from infarcts in the enlarged spleen. These conditions often improve following shunt surgery, particularly after splenectomy and proximal lienorenal shunt (LRS). [11, 12]

Surgical options for NCPH can be broadly categorized into portosystemic shunts, esophagogastric devascularization, and the more recently described Rex shunt. [13,14] Despite the availability of Transjugular Intrahepatic Portosystemic Shunt (TIPS) and liver transplantation, shunt surgery and devascularization will likely continue to play an important role in the management of NCPH patients.[15]

Our patient was discharged on beta-blockers to decrease the portal pressures and with regular timely follow-ups. But due to refractory anemia splenectomy was considered.

**Conclusion**

In conclusion, this case report highlights the nuanced clinical presentation of Banti syndrome in our patients. The challenges in diagnosis and management underscore the need for heightened awareness among healthcare providers. Further research and collaboration are crucial to enhancing our understanding and refining treatment strategies for this condition.
Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his/her consent for his/her images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published, and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflict of Interest: Nil

Financial Disclosure: None

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References

7. Dutta 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;20:378-84.


