Splenic Infarct: An Outlandish Presentation of Banti's Disease - A Case Report

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ABSTRACT

Banti's disease is a rare disease of unknown aetiology acquiring various names in different parts of the world. The hallmark of the disease is a chronic enlargement of the spleen, signs of hypersplenism, and portal hypertension without any known liver pathology and biopsy-proven healthy liver tissue. A 16-year girl presented with complaints of abdominal pain and fever. After extensive work-up, she was diagnosed as a case of Banti's disease/non-cirrhotic portal hypertension (NCPH), presenting with splenic infarction. This presentation of NCPH is unique, and it has not been mentioned in the literature. The patient was managed surgically *via* splenectomy.

Key Words: *Splenic infarct, Banti's disease, Non-cirrhotic portal hypertension, Hypersplenism*.

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INTRODUCTION

Banti's disease was named by Guido Banti in 1894.[1] Mikkelsen *et al*. classified the condition known as hepatoportal sclerosis in 1965 after they reported 36 patients with splenomegaly and non-cirrhotic portal hypertension (NCPH).[2]

Banti's disease is denoted by numerous names across the world. In India, it is referred to as non-cirrhotic portal fibrosis, in Japan as idiopathic portal hypertension (IPH), in the United States as hepatoportal sclerosis, and in Europe as Banti's disease.[3]

Herein, the authors present a case of Banti's disease in a young female presenting with abdominal pain and fever.

CASE REPORT

A 16-year female, resident of Sindh, presented with excruciating abdominal pain and fever of 102-103˚F for a month. The abdomen was distended with a palpable, tender spleen, crossing the midline. Ultrasound of the abdomen showed a massively enlarged spleen with infarctions and moderate ascites. These findings were further confirmed by contrast-enhanced computed tomography (CECT) abdomen, with the addition of splenic vein thrombosis (Figure 1). She was worked-up meticulously at the age of 7 years, with upper gastrointestinal (UGI) endoscopy and bone marrow biopsy, which were unremarkable.

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She has had multiple blood transfusions since then, and the last one was a month ago. A liver biopsy revealed few portal tracts, with no granuloma or malignancy. The patient was labelled as a case of NCPH/Banti's disease after ruling out possible causes (Table I). Splenectomy was performed after proper vaccination. Post-splenectomy, the platelet count returned to normal (Table II). She is on regular follow-up and is leading a normal life.

DISCUSSION

Banti's disease is a chronic, congestive enlargement of the spleen that destroys blood cells. It manifests as signs of hypersplenism such as recurrent infections, bruisability, easy fatigability, and anaemia.[4]

It is a diagnosis of exclusion when all other known causes of splenomegaly and portal hypertension are ruled out, and liver biopsy reveals no pathology.[5]

There are a few cases of NCPH reported before including a case report of a 20-year female reported with easy fatigability and menorrhagia and another case report of a 29-year male having haematemesis and melena.[6,7] In this patient, there was no such presentation. The presentation with splenic infarcts has not yet been reported.

Splenic infarctions result from disrupted blood supply to the spleen. These are most commonly caused by haematological disorders and thromboembolism.[8] Haemoglobinopathies, including sickle cell disease and thalassaemia major, have a fair contribution to the cases presenting with splenic infarcts. A 23-year male, having no prior medical history, presented with dull upper abdominal pain, and was later found to have an infarcted spleen secondary to sickle-thalassemia.[9] Another case report was of a 16-year girl, diagnosed as a case of β-thalassemia, who presented with splenic infarct.[10]

Table I: Pertinent diagnostic work-up of the patient.

Lab parameters	Results	Reference ranges
Total bilirubin	1.0	0.2-1.3 g/dL
SGOT	12	8-45 U/L
SGPT	5	5-40 U/L
Alkaline phosphatase	45	35-130 U/L
Serum albumin	3	3.5-5.2 g/dL
Prothrombin time	10.0	10.5 sec
INR	1.14	–
HBsAg	Negative	–
Anti- HBc	Not detected	–
Anti- HCV Ab	Not detected	–
FBS	92	<100 g/dL
ANA + ASMA + AMA	Negative	–
Serum ceruloplasmin	0.31	0.21-0.55 g/dL
Serum ferritin	969	10-310 µg/dL
HPLC	HbA- 97% HbA2- 2% HbF- 1%	–
α1-anti trypsin	Negative	–
MP + MP-ICT	Not detected	–
Serum IgM	0.94	0.5-3 g/dL
Protein-C	61%	70-140%
Protein-S	77%	56-121%
Blood C/S	No growth	–

SGOT: Serum glutamic-oxaloacetic transaminase, SGPT: Serum glutamic pyruvic transaminase, INR: International normalisation ratio, HBsAg: Hepatitis B surface antigen, Anti-HBc: Anti hepatitis B core antibody, Anti-HCV Ab: Anti hepatitis C virus antibody, FBS: Fasting blood sugar, ANA: Anti-nuclear antibody, ASMA: Anti smooth muscle antibody, AMA: Anti mitochondrial antibody, HPLC: High-performance liquid chromatography, HbA: Haemoglobin A (adult haemoglobin), HbA2: Haemoglobin A2, HbF: Haemoglobin F (foetal haemoglobin), MP: Malarial parasite, MP-ICT: Malarial parasite-immunochromatographic test, IgM: Immunoglobulin M, Blood C/S: Blood culture and sensitivity.

Table II: Pre- and post-splenectomy full blood counts.

CBC	Preoperative	Postoperative	After 6 months
Hb (g/dL)	7.7	10.6	11.8
TLC (×10³/µ)	3.2	5.2	5.4
PLT (×10³/µ)	83	282	560

CBC: Complete blood count, Hb: Haemoglobin, TLC: Total leucocyte count, PLT: Platelets.

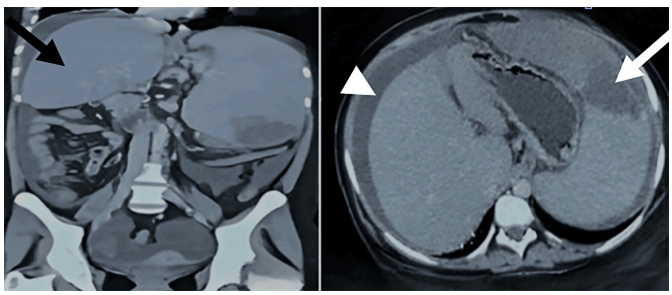

Figure 1: Contrast-enhanced CT of abdomen. Left-coronal section - healthy liver tissue (black arrow). Right-transverse section-wedged shaped area of infarction in the spleen (white arrow) and ascites (white arrow head).

The present patient was comprehensively worked up to determine the cause of the infarction. After ruling out all potential causes, it was labelled as a case of NCPH / Banti's disease. The thrombophilia profile revealed a slightly low level of protein C, which was likely acquired due to consumption coagulopathy. She has been following up regularly and living a focused life.

Banti's disease is an infrequent illness without any established cause. It has various clinical manifestations, and splenic infarction is somewhat exceptional. This case report gives an insight to the variable presentation of cases of NCPH.

PATIENT'S CONSENT:

Written informed consent for patient information and images to be published was provided by the patient.

COMPETING INTEREST:

The authors declared no conflict of interest.

AUTHORS' CONTRIBUTION:

RJ: Intellectual contribution, case diagnosis, and proofreading.
MM: Manuscript drafting and proofreading.
TBK: Collected and contributed the data.
HA: Final approval.
All authors approved the final version of the manuscript to be published.

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