

# Rare Case of Massive Splenomegaly: Case Report

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## ABSTRACT

A rare case of massive splenomegaly in 7 year old boy. Case report review on management and pathogenesis. The aim of this article is to present a case report of Gaucher disease which was diagnosed in patient with hepatosplenomegaly. A 7-year-old boy who was referred to our department due to the presence of a large mass in the left upper abdomen; over 4 years it had steadily increased in size causing dull pain and early satiety. Ultrasound and CECT scanning of abdomen and pelvis were performed, it as massive splenomegaly with hypersplenism.

**Keywords:** Massive splenomegaly, splenectomy, vaccination, gauchers disease.

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## INTRODUCTION

Splenomegaly is a common finding in a wide spectrum of diseases. Massive splenomegaly, however, always indicates underlying pathology. Massive splenomegaly is usually defined as a spleen extending well into the left lower quadrant or pelvis or which has crossed the midline of the abdomen. Massive spleens weigh at least 500 to 1000 g<sup>[1]</sup>. The most common disorders associated with splenomegaly are infective like malaria; hemolytic, storage disorders like gaucher's disease and Niemann pick disease, hereditary spherocytosis, sickle cell anemia, portal hypertension with or without hypersplenism<sup>[12]</sup>.

## CASE REPORT

7 year old boy who was referred to our department with massive splenomegaly and hypersplenism since 4 years. The child had a history of malaria 4 years back and he was treated. On examination no lymphadenopathy and jaundice. Anemia was present.

### Local examination

On examination and investigation he was confirmed to have massive splenomegaly (19cms) with hypersplenism

and without ascites and no other relevant findings.

### Investigations

Laboratory data revealed pancytopenia (Hb=8.4 gm/dl, WBC=4000/mm<sup>3</sup> with 48% PMN and 49% lymphocytes, Platelet = 1,20,000/mm<sup>3</sup>) and peripheral blood film showed bicytopenia with hypochromic microcytic and normocytic hypochromic anemia. His coagulation profile was normal, as were his liver function tests.

Bone marrow biopsy showed a hypocellular marrow with marked increase in foamy histiocytes and lipid laden macrophages called Gaucher cells which are highly characteristic of Gaucher's disease. With a high possibility of Gaucher's disease (GD) the serum level of  $\beta$ -glucosidase was done and was found to be absent in this patient; this confirmed our diagnosis of Gaucher's disease<sup>[10]</sup>.

- X ray erect abdomen- normal
- X ray chest- normal
- USG Abdomen- Mild hepatomegaly and gross splenomegaly with altered echotexture



Figure:1 Showing gross distension of abdomen



Figure:2 showing anterior surface of spleen

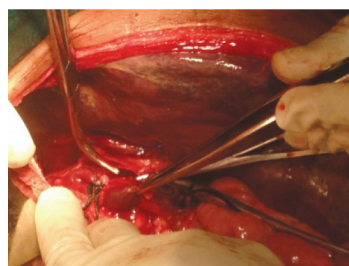


Figure:3 showing enlarged splenic vein

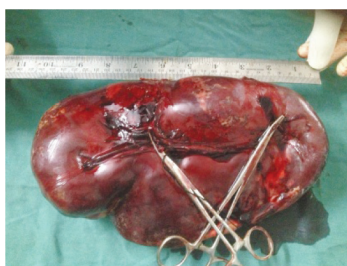


Figure:4 showing spleen size of 24cms



Figure:5 CT image of spleen(coronal view)

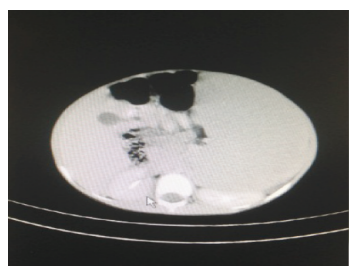


Figure:6 CT image of spleen(axial view)



Figure:7 Drain kept in left hypochondrium



Figure:8 post operative abdominal incision

- CECT Abdomen and Pelvis- Liver 14cms-increase in size, and portal vein normal. Spleen 19cms gross splenomegaly with few peripheral infarcts displacing the bowel loops to right side of the abdomen and compressing left kidney. Gall bladder, pancreas, right kidney urinary bladder, prostate and lung bases are normal[Fig:5,6].

#### Differential Diagnosis

In a patient with massive splenomegaly, differential diagnoses include malaria; hemolytic, storage disorders like gaucher's disease and Niemann pick disease, hereditary spherocytosis, sickle cell anemia, portal hypertension with or without hypersplenism.

#### Management

In view of splenomegaly with hypersplenism like severe pancytopenia he underwent open splenectomy.

Prior to splenectomy he was given pneumococcal, hemophilus, and meningococcal vaccine and was put on antibiotic prophylaxis to prevent OPSI<sup>[11]</sup>. Post splenectomy, the patient was kept under observation. During his post-operative period in the hospital serial monitoring of patient's blood counts was done; leucocytosis and thrombocytosis were observed. At the time of discharge his Hb was 14.8gm%, total leucocyte count was 15,700/mm<sup>3</sup> and platelet counts were 2,24,000/mm<sup>3</sup>.

- Spleen weighed 3 kgs, with Surface of whitish pale slightly depressed firmer areas; It also has Dark-red and grey areas due to infarcts, hypersplenism and enlarged vasculature[Fig:4].

## DISCUSSION

Gaucher disease is an autosomal recessive lysosomal glycolipid storage disorder characterized by deficiency of the enzyme acid beta-glucosidase<sup>[5,6]</sup>. This deficiency causes the accumulation of glucosylceramide in the lysosomes of macrophages leading to clinical manifestations of hepatosplenomegaly, anemia, thrombocytopenia, and bone disease. Bone manifestations include fractures, infarctions, and vertebral collapse. There are 3 phenotypes of gaucher disease: nonneuronopathic disease (known as type 1 disease), which is the most prevalent, accounting for more than 90% of cases; acute neuronopathic disease (type 2), which is rare and found mostly in infants; and chronic neuronopathic disease (type 3), which is characterized by progressive neurologic involvement. One third of patients are diagnosed after age 20 years. splenomegaly is one of the most common presenting signs. Analysis of a registry of patients with gaucher disease showed that the spleen was enlarged 5 to 75 times normal (median, 15.2 times normal) and frequently was massive at time of presentation. Enzyme replacement therapy with recombinant glucocerebrosidases<sup>[7]</sup> is the preferred treatment for many symptomatic patients with type 1 disease.<sup>[5-8]</sup>

## CONCLUSION

Massive splenomegaly is mostly due to malaria, hemolytic, storage disorders like Gaucher's disease and Niemann pick disease, hereditary spherocytosis, sickle cell anemia, portal hypertension with or without hypersplenism, and lymphomas. Treatment depends on the underlying disease.

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