

World's Heaviest Spleen Removed

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ABSTRACT

A 33 yr old man with hepatosplenomegaly, presented with abdominal pain and distension. After establishing the diagnosis of type I Gaucher's disease, splenectomy was done which weighed a massive 14.96 kilograms. The heaviest spleen reported till date. A case report of splenectomy for massive splenomegaly with comparison of other reports is presented. Splenectomy in gauchers's disease have been reported in the past for hypersplenism and splenomegaly. Splenomegaly and hypersplenism causes poor quality of life with abdominal distension and discomfort, recurrent blood transfusions, increased risk of bleeding, pancytopenia. The main indication in our case was massive splenomegaly extending well below the umbilicus and reaching upto the opposite iliac fossa (Hackett's grade V). Such massive sized spleens have never been reported in the past as patients present before reaching this level. Performing splenectomy in this case was in itself an arduous task. Our patient was operated by conventional midline laparotomy and recovered well after surgery.

Key words: Gaucher's disease, splenomegaly, splenectomy, abdominal distension.

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Conflict of interest: No

Case report is Original: YES

Whether case report publishes any where? NO

INTRODUCTION

Gaucher's Disease was first described as a clinical syndrome by Dr. Phillippe Gaucher as an inborn error of metabolism that results from the deficiency of lysosomal enzyme glucocerebrosidase with subsequent collection of glucocerebroside within the lysosome of cells mainly in liver, spleen, bone marrow and brain [1,2].

Splenectomy has been beneficial in solving the hematological and mechanical complications of this disease. The massive splenomegaly can lead to abdominal pain and obstructive signs which are readily relieved by splenectomy [3].

In most cases splenectomy for patients with massive splenomegaly relieves symptoms whether due to the huge spleen itself or to the associated hypersplenic cytopenias [4].

CASE REPORT

A 33 yr old gentleman from Iraq presented to us with grade V splenomegaly (Hackett's grading) (figure 1) and hepatomegaly, abdominal pain and weakness on admission. Ultrasound and CT abdomen revealed Hepatomegaly with massive splenomegaly. His vitals were stable. His complete blood count showed profound anaemia with Haemoglobin level of 6gm% and pancytopenia. Bone marrow biopsy showed collections of plump distended macrophages having single eccentric nuclei and fibrillary cytoplasm (crumpled paper appearance) at many places resembling Gauchers cells. Finally a diagnosis of type 1 Gaucher's disease was established.

In view of features of Hypersplenism and the massive size of the spleen causing abdominal pain and deteriorating quality of life, Splenectomy was done. As a part of presplenectomy protocol, the patient was immunized against H. influenza, Meningococcus and Pneumococci. Splenectomy was done through a formal laparotomy approach by a midline incision due to massive splenomegaly (figure 2). The post operative specimen weighed 14.96 kilograms. The post operative course was uneventful and the patient was discharged after 7 days of surgery.

It is till date the heaviest spleen removed. The case has been reported in The Limca Book of records.

Figure 1 – preoperative picture of the abdomen showing surface marking of the massive spleen (grade V of Hackett's grading)



Figure 2 – post operative picture of the gigantic spleen removed.



DISCUSSION

Significant leukopenia and anemia do occur in gaucher's disease but are unusual prime indications for splenectomy. However, splenectomy does not alter the amount of glucocerebroside to be metabolized. Once the spleen is removed, accumulation of glucocerebroside in the liver and bone may actually increase. Therefore, splenectomy should

be deferred until hypersplenism or mechanical symptoms present severe problems in patient management as in our case [3].

Splenectomy for massive splenomegaly (>1500 g) provides palliation but is associated with a high rate of perioperative complications . Splenectomy for massive splenomegaly was performed most commonly for non-Hodgkin lymphoma (48%) and myeloid metaplasia (31%) in a study of 222 consecutive patients [5].

Of the various studies reported worldwide of splenectomy for splenomegaly, the ranges of the size of spleen removed have been 0.5 to 13.0 kgs [3,5]. Removal of 14.96 kgs of spleen (heaviest ever reported) from a patient of gaucher's disease (rare disorder) makes this report distinctive.

CONCLUSION

Splenomegaly of any origin should not be deferred and prompt action to elucidate its cause be initiated. If due to untreatable medical illness, all efforts must be made to improve the quality of life of the patient by total or partial splenectomy.

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