Partial Splenectomy for Gaucher's Disease: A Case Report

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ABSTRACT

Massive splenomegaly leading to hypersplenism is one of the main clinical manifestations of Gaucher's disease, an autosomal recessive disorder caused by deficiency of beta glucocerebrosidase. Traditionally, splenectomy has been the treatment of choice, but this may be associated with a high incidence of post splenectomy sepsis. To obviate this risk, partial splenectomy is recommended as the treatment of choice. This is the case report of 1 1/2 year old girl with Gaucher's disease complicated by massive splenomegaly and hypersplenism treated successfully by partial splenectomy.

Gaucher's disease is an autosomal recessive disorder caused by deficiency of beta-glucocerebrosidase resulting in an accumulation of glucocerebroside in the reticuloendothelial system. The disease was first described by Phillipe Gaucher in 1882,[1] but it is in 1965 when Brady and co-workers[2] actually demonstrated the deficiency in Gaucher's disease. As a result of the accumulation of glucerebroside in the reticuloendothelial system, the principal clinical manifestation of Gaucher's disease is Hepatosplenomegaly. Traditionally, splenectomy is indicated in these patients to alleviate the pressure symptoms from massive splenomegaly and the associated pancytopenia,[3,4] but total splenectomy is known to be associated with overwhelming post splenectomy infection (OPSI) especially in children.[5,6] To overcome this, partial splenectomy is recommended. Partial splenectomy is feasible and has been shown to offer protection against sepsis,[7,8] and in Gaucher's disease it also affords improvement in the mechanical and haematological problems.[9,10]

We report a successful partial splenectomy in a Saudi girl with Gaucher's disease.

THE CASE

A 1 1/2 year old female patient presented with abdominal distension which was noticed by her mother few days prior to her presentation. This patient's older brother has been diagnosed as having Gaucher's disease several months earlier and had undergone a total splenectomy. On examination, the patient was found to be pale but not jaundiced. Abdominal examination revealed hepatomegaly of 4cm below the costal margin and the spleen was massively enlarged about 5 finger breaths below the left costal margin. Laboratory studies revealed a haemoglobin of 7 gram/dl, a haematocrit of 25%, white blood cell count of 8300 x 10^9/1 and platelet count of 74,000 x 10^9/1. Bone marrow aspiration was diagnostic of Gaucher's disease. She was then referred to paediatric surgery. The patient was operated on and an estimated 90% partial splenectomy was done. The operation was performed through a left upper quadrant transverse incision. The spleen was fully mobilised by sharp dissection, and all vessels in the splenic hilum were divided between ligatures, with the exception of the single vessel to the upper pole of the spleen. The spleen was divided at the line of vascular demarcation with electro-cautery. The resected specimen weighed 1 kilogram (Fig 1). Bleeding was controlled with electrocautery. The raw surface of the spleen was covered with

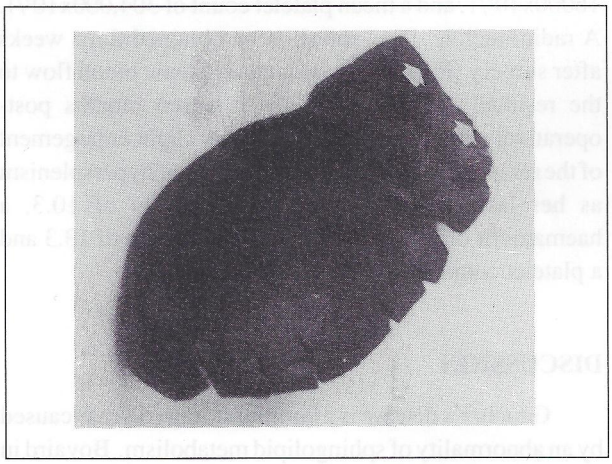

Figure 1. *The resected spleen, weighing 1 kg. The arrows indicate the resection margin.*

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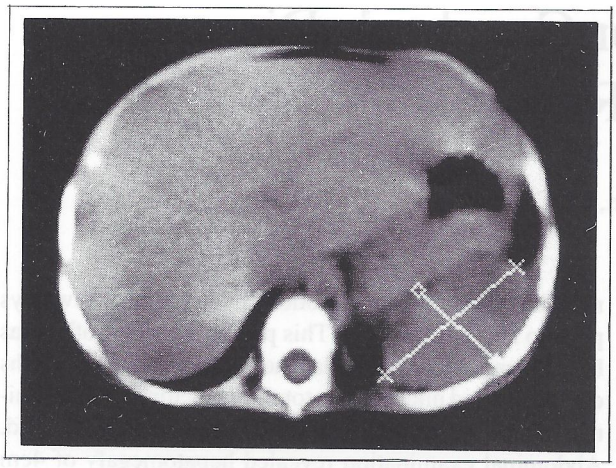

Figure 2. *CT scan demonstrating the splenic remnant*

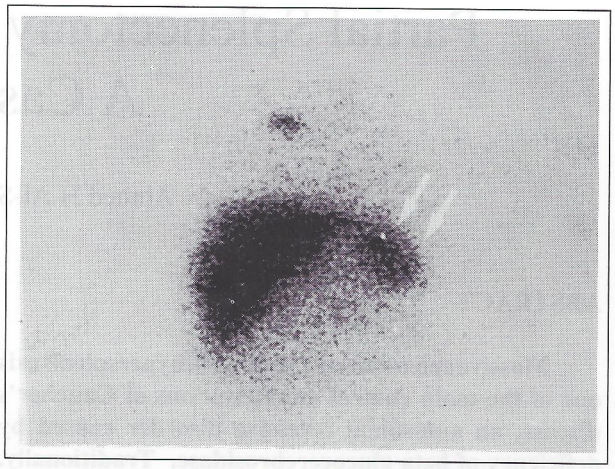

Figure 3. *Radionuclide scan after partial splenectomy demonstrating patency of the blood supply and function of the splenic remnant.*

surgical (oxidised cellulose) and few stitches were applied to the edges to hold the surgical in place. The remaining spleen was returned to the splenic bed, and few stitches were used to fix it in its place so as to prevent wandering of the spleen or torsion of its vessels. The patient was treated with perioperative antibiotics and received pneumococcal vaccination before operation. Post-operatively the patient did well and was discharged home on the seventh post operative day. Two weeks post operatively, a CT scan of the abdomen was obtained. This demonstrated an intact small size spleen (Fig 2). Laboratory studies revealed a mean haemoglobin of 11.5 g/dl, a mean Haematocrit of 37.4%, a mean white blood cell count of $16000 \times 10^9/1$, and a mean platelet count of $304{,}000 \times 10^9/1$. A radio-nuclide liver-spleen scan obtained three weeks after surgery (Fig 3) indicated intact splenic blood flow to the residual spleen. Now she is seven months post-operatively and clinically there is only slight enlargement of the remaining spleen with no evidence of hypersplenism as her last CBC showed a haemoglobin of 10.3, a haematocrit of 33.1, a white blood cell count of 13.3 and a platelet count of $187{,}000 \times 10^9/1$.

DISCUSSION

Gaucher's disease is a familial storage disease caused by an abnormality of sphingolipid metabolism. Bovaird in 1990 demonstrated the familial nature of the disease.[11] Brady and co-workers in 1965[2] demonstrated the underlying metabolic defect as a deficiency of B-glucocerebrosidase. This will result in an accumulation of gluco-cerebroside in the reticuloendothelial system leading to hepatosplenomegaly and bone pain. The spleen in these patients can be massively enlarged leading to pressure symptoms and invariably to hypersplenism.

The most common complication of Gaucher's disease amenable to surgical therapy is the massive splenomegaly that develops in the majoriy of patients, and traditionally the surgical treatment in these patients was total splenectomy.[3,4] This aims at alleviating the pressure symptoms caused by the massively enlarged spleen as well as correction of the associated pancytopenia, but splenectomy has been demonstrated to result in an increased risk for sepsis.[5,6] Post splenectomy sepsis is now a well recognized sequelae of splenectomy which can occur at any age, and remains a life long risk, although the risk is greatest in childhood. This was clearly demonstrated by King and Shumaker report in 1952.[5]

To obviate the dangers inherent in total splenectomy and due to improvement in surgical techniques in recent years, alternative means of splenic preservation became feasible. These have already been used successfully in cases of splenic trauma,[12] splenic cysts,[13,14] splenic abscess,[15] hodgkin's disease,[16] thalassaemia major,[17] and gaucher's disease.[9,10,18] Partial splenectomy can be performed because of the segmental nature of its blood supply[19] and infact the first successful partial splenectomy date back to 1869, when Pean reported the first successful partial splenectomy in a human for a treatment of a splenic cyst.[20] In Gaucher's disease, partial splenectomy or properly called near total splenectomy has been shown to be beneficial,[9,10] although long term follow-up is not available. There is a marked decrease in the splenic size which alleviate the pressure symptoms and improvement in the haematological parameters, and since some of these patients have a normal life expectancy,[10] partial splenectomy should protect them from the danger of life-threatening sepsis. It will also spare them the necessity of taking long term antibiotic prophylaxis.

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