Some Unusual Indications Of Splenectomy

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Citation

F Parray, N Wani, K Wani, M Wani, R Wani. *Some Unusual Indications Of Splenectomy*. The Internet Journal of Surgery. 2005 Volume 7 Number 2.

Abstract

While dealing with various hematological disorders which were subjected to therapeutic splenectomy in our department of general surgery at a tertiary care centre Sher-i-Kashmir Institute of Medical Sciences over a period of more than 20 years, we came across a rare group of diseases in our set up from time to time. This rare group included 5 patients with 1. Hairy cell leukemia -1, 2 Malignant histiocytoma -1, 3 Gaucher's disease -1, 4 Tropical splenomegaly -1, and 5 Splenic hemianginoma-1. All these patients were referred to our department from the department of clinical hematology of the same institute for splenectomy for various types of cytopenias. Response to splenectomy was excellent in malignant histiocytosis, splenic hemiangioma and some of the parameters in hairy cell leukemia and Gaucher's disease, while as an unsatisfactory response was observed in tropical splenomegaly sydrome and in some of the parameters of hairy cell leukemia and Gaucher's disease.

INTRODUCTION

Splenectomies have been performed as early as 2,000 years ago, as per references in ancient Greek and Roman literature. The first total splenectomy for disease is attributed to Adriano-Zaccarello in 1549, although it has been suggested that the excised viscus was an ovarian cyst, clark (1676) performed splenectomy in dogs with survival. Nicholas Mathias is credited with first total splenectomy for trauma in 1678. Mayo in his retrospective study of 500 splenectomies done mainly for anemia and leukemia concluded that 80% of patients had excellent results and lived comfortably but the mortality in those days was 10%[2]. The landmark report of king and Schumacher (1952) first postulated that the spleen may play a role in resisting infection particularly in infancy[3], but their view point was opposed by gross (1953) and Laski and Mac Millan (1959). The data given by Gross was reported in Gofstein and Gellis (1956) series of 206 cases where they concluded that there was some co-relation between splenectomy and sepsis irrespective of the age at operation.[4]

However, with the advent of safe anesthesia, modern surgical technique, broad spectrum antibiotics and specific vaccines, the morbidity and mortality following splenectomy is definitely on a decline, hence more and more studies were conducted in lost 35 years in an attempt to determine the true morbidity, mortality and long term infection following splenectomy[5,6,7,8].

Now-a-days, role of splenectomy for various hematological disorders has definitely earned a place as a therapeutic measure especially in disorders like idiopathic thrombocytopenic purpura and hereditary spherocytosis but in diseases like sickle cell anemia, the outcome cannot be predicted with certainity[9]. Still, it is too early or impossible to estimate the risk of overwhelming post splenectomy sepsis after partial splenectomy. The people who do this procedure follow common practice guidelines that advocate immunoprophylaxis against pneumococcus, meningococcus and H-influenza as well as antibiotic prophylaxis for the child undergoing total splenectomy, although the efficiency of these measures has not been proven[10]. Given the low rate of overwhelming post-splenectomy sepsis with proper use of immunization and antibiotic prophylaxis, any risk reduction for partial splenectomy compared to total splenectomy would require a prohibitively large clinical trial.[11]

MATERIAL & METHODS

Our Study included patients with various hematological disorders subjected to splenectomy from July 1982 to July 2005 at Sher-i-Kashmir institute of Medical Sciences, Soura Srinagar in the department of general surgery. Medical records of all the retrospective cases were reviewed. Preoperative management of all cases included a detailed hematological and coagulation studies. Preoperative antibiotics like ampicillin and gentamycin or a third generation cephalosporin were used for prophylaxis. Corticosteroids (prednisolone) were given in the

preoperative period in patients whenever indicated. A total of 125 patients with various hematological disorders were subjected to splenectomy under general anesthesia. The patients diagnosed to have splenectomy for rare clinical entitities in our set up were grouped under "others" group, which were "5" in number. All patients were put on I/V fluids post-operatively for 24-48 hours. Steroids were continued if the patient had been receiving these drugs before splenectomy. In all patients hydrocortisone was used and tapered for 48-72 hours in the post-operative period. Antibiotics were routinely given in the post-operative period. A preset routine was followed for taking blood samples on day "0", day "3" and day "7" for assessment of hematological parameters which included (Hb%), total leukocyte count (TCC) and platelet count (PC) and again on follow up in 1st month, 3rd month and 6th month. The mean of all these reading were used for comparison with preoperative parameters to assess the response as described below:

Response was defined as complete response (CR). Partial response(PR) and no response (NR) for thrombocytopenia neutropenia and anemia as explained under.[8,12,13,14,15].

WANI AND PARRAY CRITERIA

RESPONSE IN A THROMBOCYTOPENIC PATIENT.

Complete (CR) – 100,000 U/L or above.
Partial (PR) – 50,000-100,000 U/L
No Response (NR) -< 50,000 U/L

RESPONSE IN A NEUTROPENIC PATIENT

Complete (CR) – 4,000/cm^3 or above. Partial (PR) – 2,000 -4,000/cm^3 No Response (NR) - < 2,000/cm^3

RESPONSE IN AN ANEMIC PATIENT

Complete (CR) – Hb 10g% or above.
Partial (PR) – Hb 8-10g%
No Response (NR) –Hb <8g%

The follow up was complete in 92% cases; duration of follow up ranged from 1 week to 8 years with a mean of 10 months. The clinical diagnosis of all patients was confirmed after surgery by histopathological examination of spleen.

CASE DISCUSSION AND RESULTS

In "Others" group "5" patients with various hematological disorders subjected to splenectomy are included which are:

Hairy cell leukemia – 1 patient.
Malignant histiocytosis – 1 patient
Gaucher's disease – 1 patient
Tropical splenomegaly – 1 patient
splenic hemiangioma – 1 patient

CASE 1

Hairy cell leukemia (HCL) is a disease characterized by pancytopenia, circulating mononuclear cells with prominent cytoplasive projections and moderate to massive splenomegaly without significant lymphadenopathy.[16]

Patient with HCL was 45 years old male patient who presented with features of hypersplenism i.e., pancytopenia. The pre-operative platelet count was 32,000 U/L; Hb 8.5g/dl and leukocyte count was 2,500/cm^3. the patient did not receive any preoperative medication, however, received whole blood transfusion and 2 fresh frozen plasma units. Patient was subjected to elective splenectomy; no significant per-operative complication was encountered. Patient received 1 unit of platelet concentrate and 1 unit of whole blood in the pre-operative period. No post-operative complication was encountered. The response to splenectomy was excellent in 2 parameters. Mean post-operative platelet count was 2,50,000 U/l i.e., complete response and mean leukocyte count was 4,500/cm (complete response). However, the anemia showed only a partial response i.e., mean post operative hemoglobin was 9.8g/dl.

Golomb and vardiman [17](1983) observed a complete remission in 27 out of 65 patients in all the three parameters and a partial remission in 38 out of 65. since, our study includes only one patient, so we don't have sufficient data for comparison. The weight of the spleen was 4200 gms which is within the range [17]as observed by others. No accessory spleen was found.

CASE 2

Malignant Histiocytosis: the disease is also known as " histiocytic medullary reticulosis". The disease is widely believed to represent a malignant tumor of mature and immature histiocytes with diffuse invasion of lymph nodes and the bone marrow. It derives its name from the infiltration of the medullary zone of the lymph nodes by the histocytes which by no means is a diagnostic feature. The major clinical features include 1. lymphadenopathy 2. hepatosplenomegaly 3 anemia or pancytopenia 4 .fever and sometimes 5. skin infiltrates .[18]

Our patient with malignant histiocytosis was a 10 year old female child who presented with splenomegaly, lymphadenopathy, fever, leucopenia and border line anemia

(Hb=log/dl) the patient received 10 units of fresh frozen plasma and 1 unit of whole blood in the pre-operative period and was subjected to elective splenectomy. No post-operative transfusion was given ;post operative period was uneventful.

Post-operative hematological response was excellent; mean leukocyte count was 6,000/cm^3 i.e., complete response and even Hb improved to 11.5g/dl. the spleen weighed 2060gm. no accessory spleen was found.

CASE 3

Gaucher's disease: is a disorder of lipid metabolism that may result in massive splenomegaly and hypersplenism. Genetically transmitted as autosomal recessive trait, the disease is mostly commonly found in Ashkenazie jews, caused by a deficiency of B-gluco-cerebrocidase, an enzyme responsible for breaking down certain lipid complexes. Gaucher's disease ultimately leads to retention of glucocerebosides in macrophages, especially those of spleen, liver, bone marrow and lungs.[19,20]

In our study, the patient with Gaucher's disease was the youngest i.e., 4 years old male child. The patient's platelet count was 1,55000 u/l.

Hemoglobin was 8.6g /dl and leukocyte count was 3,500/cm^3. The patient received 150 ml of whole blood in the preoperative period; was subjected to elective splenectomy. No per operative complication was encountered as in other studies[21], however, patient did not show CR in leukocyte count. The mean post operative count remained 3800/cm^3, however, Hb% improved and got stabilized at 10.8g/dl (CR).

Splenic weight was 850gms and no accessory spleen was found. splenic weight observed in other studies in Gaucher's disease is quite higher (mean 4270gm)[21]. Even though, Gaucher's disease with gross splenomegaly and severe depression of blood count, is recognized as a poor risk for splenectomy, but our experience has been encouraging because the patient is living a normal life after operation consistent with experience of Dawson et al (1987) study[22].

CASE 4

Tropical splenomegaly syndrome (TSS): the existence of massive splenomegaly of uncertain etiology in different parts of the world is being referred by the non-specific term (TSS) tropical splenomegaly syndrome) ; until precise etiology in different population is ascertained (pitney, 1968). Tropical splenomegaly syndrome is found only where malaria is endemic in the adult life, although pitney (1968) emphasized that it may be fully developed by the age of 8 years. Mechanical symptoms due to massive enlargement of spleen are common. There is often a history of repeated attacks of fever and malaise but typically patients are afebrile when splenomegaly is established[23].

Our patient with TSS was a 48 years old female who presented with massive splenomegaly, anemia and thrombocytopenia. The pre-operative platelet count was 50,000 U/l and Hb was 8.0 g/dl. Patient received 1 unit of platelet concentrate and fresh frozen plasma in the per-operative and post operative period. Elective splenectomy was done. No per operative or post operative complication encountered. The response to splenectomy was not good. The mean platelet count was 90,000U/l i.e., partial response only while as the mean post-operative Hb% was 7.9g/dl i.e., no response. Weight of the spleen was 6050 gm, no accessory spleen was found.

CASE 5

The patient was a 47 year old male, presented to us with splenomogaly and anemia. The patient was subjected to elective splenectomy, who improved post-operatively in an excellent way. The mean post operative Hb stabilized at 13.6g/dl. No per-operative or post operative complication was encountered. The splenic weight was 225 gms, no accessory spleen was found. To conclude, we are of the opinion that splenectomy still continues to be a gold standard in various cytopenias secondary to any common or rare disorder in patients where all other medical measures have failed provided there is an excellent co-operation and understanding between a hematologist and a surgeon.

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