

Significance of Splenectomy in Patients with Haematological Disorders

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A B S T R A C T

Introduction: Normal spleen serves as a major role in destruction abnormal blood cells, thereby causing anemia, leukopenia and thrombocytopenia. This is the reason; splenectomy is chosen management for a variety of haematological disorders in special conditions. Aim: In this study, we tried to project the benefits of splenectomy in certain conditions and to study on epidemiology related to splenectomy.

Material and Methods: In this study period, we have observed a total of 32 patients with haematologic disorders, who required definite splenectomy to treat the disease. Open splenectomy was performed on all patients. After splenectomy, studied population were followed post operatively for one year and their outcome was assessed.

Results: Out of 32 patients, 11 (34.3%) had ITP, 10 (31.2%) had Hemolytic anemia, 4 (6.2%) had thalassemia and acquired haemolytic anemia each, 2 (12.5%) had non hodgkins lymphoma and 1 (3.1%) patient had thrombocytopenic purpura. Among 32 patients, 9 (28.1%) presented Overwhelming Post splenectomy infection syndrome (OPSI), 8 (25%) had haemorrhage, 6 (18.75%) presented with wound dehiscence and ascites each. Mortality among patients with OPSI was observed as 44.4%.

Conclusion: Along with prompt diagnosis and treatment for OPSI, clinicians should focus on educating postsplenectomized patients about their asplenic or hyposplenic status.

Keywords: Haematological Disorders, Splenectomy

INTRODUCTION

Spleen is a central lymphoid organ, plays a vital role in body's immune system and as well haematological functions.^{1,2} Important functions of the spleen are: clearance of microorganisms and particulate antigen from blood stream. Synthesis of immunoglobulin G (IgG), Properdin and tuftsin; removal of RBCs, extramedullary haematopoiesis in certain diseases.

In 1826 first splenectomy was performed by Quittenbaum. Until King and Schumacher drew attention to the risk of overwhelming post splenectomy infection (OPSI) in 1952, spleen was considered unnecessary for life.³

These are several reasons for splenectomy, broadly divided into absolute and relative indications. Absolute Indications include splenic trauma, splenic abscess, splenic cysts, neoplasm and aneurysm of splenic artery. Relative indications include Splenism, Symptomatic splenomegaly and destruction of abnormal blood cells in the spleen.⁴

Indications of splenectomy for haematological conditions includes autoimmune thrombocytopenia, haemolytic anemia,

felty's syndrome, thrombocytopenic purpura, hereditary spherocytosis, thalassemia, erythrocyte enzyme deficiencies, sickle cell disease, primary hypersplenism, hodgkins disease.⁵ Normal spleen serves as a major role in destruction abnormal blood cells, thereby causing anemia, leukopenia and thrombocytopenia. This is the reason; splenectomy is chosen management for a variety of haematological disorders in special conditions.⁶

The main reasons for splenectomy are; 1. treating disease by avoiding destruction of RBC's. 2. to prevent hypersplenism and splenomegaly, 3. to stage hogkins disease, 4. when blood and bone marrow lab investigations are in conclusive.⁷

Complications related to splenectomy includes difficult to recover from infections (sepsis, pneumonia, meningitis, malaria, other gram negative and parasitic diseases), OPSI, haemorrhage, DIC, Arteriovenous thromboembolism, pulmonary hypertension, enhanced atherosclerosis, acute pancreatitis.

In this study, we tried to project the benefits of splenectomy in certain conditions and to study on epidemiology related to splenectomy.

MATERIAL AND METHODS

This is a prospective study undertaken at department of General Surgery, Government General Hospital, Anantapuram from November 2014 to July 2018. In this study period, we have observed a total of 32 patients with haematologic disorders, who required definite splenectomy to treat the disease. Informed consent has been taken from all the patients include this study.

Patients who have been chronically suffering from haematological diseases and under treatment were assessed clinically. Studied population clinical condition and their treatment plan were assessed by clinical features, radiological and lab investigations. Open splenectomy was performed on all patients. Preoperatively, vaccination is provided and antibiotics were administered according to antibiotic prophylaxis. Perioperatively extensive search for accessory spleen was done. Post operative care given.

After splenectomy, studied population were followed post operatively for one year and their outcome was assessed. Results were analyzed and tabulated. A descriptive statistics was done for qualitative analysis in the form of number, percentages.

RESULTS

In this study a total of 32 patients were underwent open splenectomy, those patients were assessed. Out of 32 splenectomised patients, 11 (34.3%) patients were observed in the age group of 31-40 years predominantly, 7 (21.8%) patients were in the age group of 21-30 and 41-50 years each, 4 (12.5%) patients were in the age group of 51-60 years and

Age in years	No. of patients	Percentage
0-20	3	9.3
21-30	7	21.8
31-40	11	34.3
41-50	7	21.6
51-60	4	12.5
>60	0	0
Total	32	100

Table-1: Distribution of splenectomised patients according to age

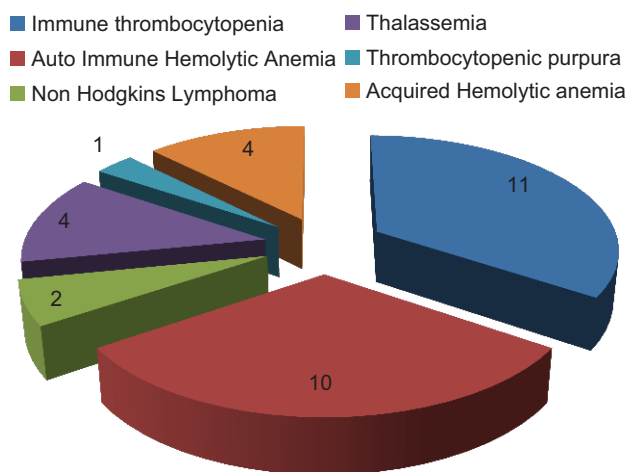


Figure-1: Various causes for splenectomy

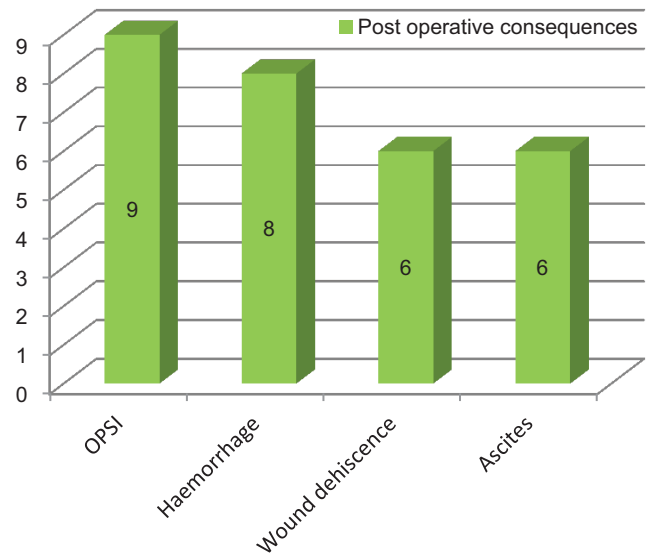


Figure-2: Prevalence of post operative consequences

remaining 3 cases were observed ≤ 20 years age group. We haven't done splenectomy on patients aged above 60 years in this study period (table-1).

On evaluating the cause of splenectomy, we have observed most common etiology was Immune thrombocytopenic purpura (ITP) followed by autoimmune haemolytic anemia. Out of 32 patients, 11 (34.3%) had ITP, 10 (31.2%) had Hemolytic anemia, 4 (6.2%) had thalassemia and acquired haemolytic anemia each, 2 (12.5%) had non hodgkins lymphoma and 1 (3.1%) patient had thrombocytopenic purpura (fig-1).

After splenectomy patients were followed and observed for post operative consequences. Among 32 patients, 9 (28.1%) presented Overwhelming Post splenectomy infection syndrome (OPSI), 8 (25%) had haemorrhage, 6 (18.75%) presented with wound dehiscence and ascites each. A good outcome is achieved in patients with ITP. A significant proportion of patients were improved haematologically after splenectomy in Hemolytic anemia, Thalassemia (fig-2).

5 (15.6%) out of 32 patients expired. Among them 4 (80%) were presented with OPSI as post operative complication. Mortality among patients with OPSI was observed as 44.4%.

DISCUSSION

Splenectomy is a useful procedure in many cases; potential benefits should outweigh the risks. Spleen is one of the most injured intraperitoneal organs, which may require splenectomy or rarely splenorhaphy.

After splenectomy, liver performs immunological function and 30% of people who have accessory spleen will start functioning when the main spleen is removed. In both cases, immunological function is insufficient, higher levels of specific antibody and an intact complement system are probably required.⁸

Autoimmune haemolytic anemia is of two types: warm or cold acting agglutinin syndromes. It can cause mild to moderate splenomegaly, runs a self limiting course; here spleen functions as production of antibodies and site for erythrocyte destruction. Warm is the most common type about 70%. Autoimmune hemolytic anemia may be

idiopathic or may develop in the course of other disorders, such as malignant lymphomas, chronic lymphatic leukemia, lupus erythematosus and others. Splenectomy is indicated when medical therapy is ineffective.⁹

Thalassemia is an inherited blood disorder in which body makes abnormal form of haemoglobin. It is of two major degrees depending on the severity and abnormal production of globin chains: Thalassemia major and minor. Beta thalassemia is characterized by deficient synthesis of the beta chain, whereas alpha thalassemia results from deficient synthesis of the alpha chain. Thalassemia can be managed by medical therapies and blood transfusions. Patients with thalassemia major require transfusions at regular intervals; the disease usually results in early death. Splenectomy is indicated among transfusion dependent patients; usually reserved for symptomatic splenomegaly and recurrent splenic infarction. The post splenectomy infection rate in these patients is rather high.¹⁰

Idiopathic thrombocytopenic purpura is the most common hematologic indication for splenectomy.¹¹ During splenectomy, surgeon should focus on accessory spleens and missing splenic tissue; if not properly checked, and then it may cause recurrence. The platelet count is characteristically less than $50,000/\text{mm}^3$. The spleen is usually only mildly enlarged. Splenectomy is usually performed only in cases in which corticosteroid therapy or IgG and plasmapheresis fails to achieve remission.¹² ITP may occur as an isolated condition or in association with such other disorders as chronic lymphatic leukemia or systemic lupus erythematosus, in which it may be the presenting manifestation.¹¹

Thrombotic Thrombocytopenic Purpura (TTP) is a rare blood disorder. In TTP, blood clots form in small blood vessels throughout the body. The clots can limit or block the flow of oxygen-rich blood to the body's organs, such as the brain, kidneys, and heart. The etiology is unclear, although it has been postulated that the disease is autoimmune in origin. The first line therapy is plasmapheresis, which is usually effective. When plasmapheresis is ineffective, splenectomy is indicated.

Hodgkin's disease (lymphogranulomatosis) and non-Hodgkin's lymphomas are grouped together under the heading "malignant lymphomas". They develop primarily in lymphatic tissue, mostly in lymph nodes. Only in less than 1% of all cases do they occur primarily in the spleen.¹³

On evaluating the cause of splenectomy, we have observed most common etiology was Immune thrombocytopenic purpura (ITP) followed by autoimmune haemolytic anemia. Out of 32 patients, 11 (34.3%) had ITP, 10 (31.2%) had Hemolytic anemia, 4 (6.2%) had thalassemia and acquired haemolytic anemia each, 2 (12.5%) had non hodgkins lymphoma and 1 (3.1%) patient had thrombocytopenic purpura.

Wilhelm MC et al¹⁴ did a study on incidence of Splenectomy among haematological disorders in a two series: Series I – 1946 to 1962 and series II – 1963 to 1982. In series II there is a fall in the annual incidence of splenectomy for hereditary spherocytosis, idiopathic hypersplenism, and myeloproliferative disorders when compared to series I. For ITP, incidence of splenectomies was 1.1 per year in

series I and 3.6 per year in series II. The total number of splenectomies for hairy cell leukemia and Felty's syndrome increased from zero in Series I to 12 and 17, respectively, in Series II, whereas the number of miscellaneous reasons dropped from 29 (1.7 per year) in Series I to 15 (0.75 per year) in Series II. Staging laparotomy currently is rarely done for non-Hodgkin's lymphomas. The mortality rate in Series I was 6.3% compared with 4.0% in Series II. No deaths occurred in Series II after 1979. Indications for splenectomy in Series II were for diagnostic purposes in 3.2%, therapeutic in 56.5%, staging in 39.5%, and restaging in 0.8%. Accessory spleens were found in 49 (12.5%) in Series II.

Norman oneil Machado et al¹⁵ stated that out of 150 patients, 96 (64%) had SCD and 34 (22.6%) had β -thalassaemia; the rest comprised patients with refractory idiopathic thrombocytopenic purpura (ITP) $n = 12$, hereditary spherocytosis (HS) $n = 6$, and auto-immune haemolytic anaemia (AHA) $n = 2$.

A good outcome is achieved in patients with ITP. A significant proportion of patients were improved haematologically after splenectomy in Hemolytic anemia, Thalassemia in the present study.

Complete response is defined as the achievement and maintenance of a platelets count of at least $100 \times 10^9/l$ 30 days or longer after splenectomy in the absence of bleeding and without additional ITP treatment, except tapering of perioperative glucocorticoids; partial response is defined as any platelet count between 30 and $100 \times 10^9/l$ 2 months after surgery in the absence of bleeding or any treatment; no response is defined as any platelet count $<30 \times 10^9/l$ or bleeding, while relapse of ITP is defined as the loss of complete response or response, or when medical treatment is required to maintain a safe platelet count.¹⁶

Seymour I et al¹⁷ observed over 80% of patients with idiopathic thrombocytopenic purpura and patients with hereditary spherocytosis achieved good results with splenectomy. Among patients with thrombotic thrombocytopenic purpura, thalassemia, myeloid metaplasia, and malignant hemopathy there is a significant improvement haematologically.

Arne Nordoy et al¹⁸ observed outcome of splenectomy in 179 patients for about 3-15 years after surgery. In about 75% patients and 100% patients of ITP and hereditary spherocytosis showed remission after splenectomy respectively. No haemolytic disease symptoms were observed in more than 50% of patients with acquired hemolytic anemia. 25% of Pancytopenia patients were in good condition for 10 years. The proliferative diseases including chronic lymphatic leukemia, lymphosarcoma and myelofibrosis may represent indications for surgery whereas chronic myelogenous leukemia seemed to be a contraindication for splenectomy. The hematological disturbances associated with congestive splenomegaly usually can be corrected by splenectomy.

Emad Gomaa et al¹⁹ did a study on comparison between laparoscopic splenectomy (LS) and open splenectomy (OS) procedure. 65% of patients who underwent LS were approached through anterior side. Surgical time was significantly longer in LS than OS group. There was significant correlation between surgical time and splenic size in both groups. Hospital stay after operation in the LS

group was much less than the OS group. Return of off-bed activities, bowel movements, oral intake and drain removal were longer but not significantly different compared to LS figures.

Demetrios moris et al²⁰ found LS surgery is a feasible and safe with a mortality rate ranging from 0% to less than 4% and the postoperative complications rate from 0% to 35.7%. The conversion rate was also very low (4%) and response (complete or partial) was achieved in more than 80% of patients.

At least 2 weeks prior to surgery, Triple vaccination (*Hemophilus influenza*, *Pneumococcus pneumoniae* and *Meningococcus*) is recommended^{21,22} Patients taking corticosteroids should be given steroids parenterally during the perioperative period^{22,23} Antibiotic prophylaxis should be commenced at the time of induction to anesthesia and continued postoperatively for at least 24 h.^{21,22,24} All patients should have compression stockings.^{21,22,25}

In this study, 5 (15.6%) out of 32 patients expired. Among them 4 (80%) were presented with OPSI as post operative complication. Mortality among patients with OPSI was observed as 44.4%.

The complications associated with splenectomy for haematological disorders reported in old publications ranged from 13% to 49%, with mortality rates of 6% to 27%.²⁶

An overwhelming post-splenectomy infection (OPSI) or post splenectomy sepsis syndrome is a rare but rapidly fatal infection. It is serious disease that can progress from a mild flu-like illness to fulminant sepsis in a short time period. Waghorn DJ²⁷ observed the mortality rate is 50-70% and most deaths occur within the first 24 hours. Marte G²⁸ did a study on Laparoscopic splenectomy, documented a single case of OPSI. Mortality due to OPSI can be reduced by prompt diagnosis and early management.²⁹

Splenectomy is ultimate therapy for few haematological disorders; more often thrombocytopenia related disorders can improve a lot. Search for accessory spleens should be considered. In warm antibody acquired hemolytic anemia, splenectomy is indicated once conventional medical therapy fails. Splenectomy is a therapeutic option for Idiopathic thrombocytopenic purpura (ITP). For Hereditary spherocytosis disorder, splenectomy is curative. Myeloproliferative disorders a group of slow growing blood cancers potentially improved by splenectomy, but preoperative management should be modified in this group. Splenectomy in patients with chronic leukemias and lymphomas are often palliative and facilitate chemotherapy.²⁹

CONCLUSION

Among patients with haematological disorders, ITP and Autoimmune haemolytic anemia were predominantly underwent splenectomy. A lasting remission was observed in haemolytic anemia cases. Mortality was high in patients with OPSI. Along with prompt diagnosis and treatment for OPSI, clinicians should focus on educating postsplenectomized patients about their asplenic or hyposplenic status. Clinicians should adhere to pre, peri and post operative care including vaccination against bacterial infections, administration of antibiotics, thromboprophylaxis, compression stockings

provision, usage of corticosteroids etc.

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