Outcomes of Splenectomy for Hematologic Diseases -

A Single Center Experience

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ABSTRACT

Splenectomy is a standard, effective and approved approach to the treatment of patients with failed medical treatment who develop recurrent, refractory or chronic diseases. The leading indications for elective splenectomy are benign and malignant hematologic diseases. We present here the findings of a retrospective analysis of the splenectomies performed in a single center for the treatment of hematologic diseases, and the associated outcomes.

A retrospective examination was made of 64 patients who underwent splenectomy for the treatment of hematologic diseases at our clinic between 2010 and 2018. The patients were assessed for gender, age, hematologic disease, spleen size, presence of hepatomegaly, presence of accessory spleen, type of surgery (laparoscopic or open), wound site infection, preoperative and postoperative platelet counts, intraoperative and/or postoperative blood replacement, and length of hospital stay.

The study sample comprised 23 (36%) male and 41 (64%) female patients, with a mean age of 40.4 years. Of the patients, 51 underwent laparoscopic splenectomy and 13 underwent splenectomy with laparotomy. Of the total, 57 (89%) patients responded fully to the splenectomy, six (9%) patients recorded a partial response and one (2%) patient had no response. Following the splenectomy procedure, one patient developed wound site infection and two patients died due to sepsis.

Splenectomy should be considered a good treatment option in hematologic splenic diseases that are resistant to medical treatment, being also associated with low mortality and morbidity.

Keywords: Spleen, Splenectomy, Hematologic Splenic Diseases, ITP

Introduction

The spleen is a lymphoid organ that plays a role in both cellular and humoral immunity. Splenectomy is a common treatment approach in the presence of hematologic splenic diseases. Benign and malignant hematologic disorders are the major indications for elective splenectomy (1). Splenectomy is a standard, effective and approved treatment approach for patients with failed medical treatment and with recurrent, refractory or chronic diseases (2). Splenectomy can also be successful in reversing hypersplenism myeloproliferative in disorders. Splenectomy has limited indications and benefits in the treatment of leukemia and lymphoma.

ITP (idiopathic thrombocytopenic purpura), as one of the most common hematologic indications for splenectomy, is an autoimmune disorder that is characterized by a decrease in the number of platelets, increased megakaryocytes in the bone marrow and a shortened platelet lifespan due to the antiplatelet factors of immunoglobulin G (IgG) class (3). The platelet membrane proteins become antigenic and cause the immune system to produce autoantibodies that once produced, either bind to the platelets, causing their destruction by phagocytosis, complement activation and lysis, or bind to megakaryocytes, resulting in reduced platelet production and thrombocytopenia (4). ITP treatment begins with immunosuppression, administered initially via IV or oral steroid therapy. In patients who do not respond to treatment, IVIG (intravenous immunoglobulin), anti-Rh immunoglobulin "anti-D" or a combination therapy can be administered (5, 6). Splenectomy should be performed as a second-line treatment in unresponsive patients (6, 7).

Studies have reported 60-90% success rates with splenectomy (8, 9). The presence of an accessory spleen must be investigated to increase treatment success, being present in 18-28% of the normal population (10). Accordingly, the presence of an accessory spleen should be considered in patients scheduled for splenectomy for hematologic reasons. The morbidity and mortality reported for splenectomy are 8-52% and 1.7%, respectively (9, 11, 12, 13). We present here the outcomes of patients who underwent splenectomy at our clinic for diagnostic or therapeutic purposes due to hematologic diseases.

East J Med 26(2): 261-264, 2021 DOI: 10.5505/ejm.2021.34735

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	Number of Patients	Splenomegaly	Hepatomegaly	Accessory spleen	Partial Response to Surgery	Non-response to Surgery
ITP	53 (82%)	1	17	10	5	1
HS	4 (6%)	4	3	1		
LYM	3 (5%)	3	2	0		
AHA	3 (5%)	3	2	0		
TTP	1 (2%)	0	0	0	1	

Table 1. Distribution by Patient Diagnoses

ITP (Immune thrombocytopenic purpura), HS (hereditary spherocytosis), LYM (lymphoma) AHA (autoimmune hemolytic anemia), TTP (thrombotic thrombocytopenic purpura)

Materials and Method

A total of 64 patients who underwent splenectomy for hematologic reasons in the Dursun Odabaş Medical Center General Surgery Clinic of the Yüzüncü Yıl University Medical Faculty between 2010 and 2018 were analyzed retrospectively. Patients undergoing splenectomy due to benign/malignant masses of the spleen and trauma-induced splenic injuries were excluded from the study. The patients were assessed for gender, age, hematologic disease, spleen size, presence of hepatomegaly, presence of accessory spleen, type of surgery (laparoscopic or open), intraoperative and/or postoperative blood replacement, preoperative and postoperative platelet counts, wound site infection and length of hospital stay. A spleen size of ≥ 13 cm was considered as splenomegaly and a liver size of ≥ 16 cm as hepatomegaly. A platelet count of $\geq 120,000/\text{mm}^3$ recorded during post-splenectomy follow-ups with no additional treatment was considered complete response, and 50,000-100,000/ mm³ as partial response. No response to treatment, in turn, was considered as failure to achieve a postsplenectomy platelet count doubled the preoperative value or >50,000/mm³. The data were examined and recorded retrospectively.

Results

The study sample consisted of 23 (36%) male and 41 (64%) female patients, with a mean age of 40.4 years. Of the patients, 51 underwent laparoscopic splenectomy and 13 underwent splenectomy with laparotomy. As shown in table 1; Immune thrombocytopenic purpura (ITP) was the most common hematologic disease indicating splenectomy, accounting for 53 (82%) patients. Other diagnoses

were hereditary spherocytosis in four (6%) patients, lymphoma in three (5%) patients, autoimmune hemolytic anemia (AHA) in three (5%) patients and thrombotic thrombocytopenic purpura (TTP) in one (2%) patient. All surgeries were elective, and 13 patients had open surgery, while 51 underwent laparoscopic surgery. When evaluated for spleen size, the spleen was normal in 53 (82%) patients. Among the patients with splenomegaly, one (2%) had ITP, four (6%) had hereditary spherocytosis (HS), three (5%) had leukemia/lymphoma and three (5%) had autoimmune hemolytic anemia. Hepatomegaly was detected in 24 (38%) patients, and among these patients, 17 (26%) had ITP, three (5%) had HS, two (3%) had lymphoma and two (3%) had AHA. An accessory spleen was detected in 11 (17%) patients, and one patient developed wound site infection after splenectomy. An evaluation of postoperative platelet counts revealed 57 (89%) patients with complete response, six (9%) patients with partial response, and one (2%) patient with no response to splenectomy. There was an accessory spleen in one of the six patients with partial response and in the patient with no response to treatment. These two patients had previously been treated with splenectomy in another center. The mean length of hospital stay was 3.4 days. Of the total, two patients developed sepsis and stayed in the intensive care unit (ICU) for 13 and 25 days, respectively, but both died due to sepsis.

Discussion

The spleen is an organ with important functions that protects the human body from infections. These functions include, but are not limited to, the filtration of circulating microorganisms, the synthesis of specific IgM antibodies and the removal of abnormal blood cells from circulation. Under certain pathological conditions, the spleen cannot function fully, and can even cause the destruction of hematopoietic elements. Splenectomy is performed for therapeutic purposes for the diagnosis and treatment of hematologic diseases.

A significant proportion of patients undergoing splenectomy due to hematologic diseases are ITP patients who do not respond well to medical treatment. ITP is the most common of all autoimmune hematologic diseases, for which splenectomy has become the second-line treatment approach following developments in medical treatments. The rate of spontaneous recovery is 80% in children diagnosed with chronic ITP, while there is a very low rate of spontaneous recovery in the adult patient group (14, 15, 16). The proportion of ITP patients who fail to achieve remission following medical treatment is 25%, and a complete response rate of 60-90% has been reported in this group of patients after surgery (8, 9, 17). The hematologic diseases treated with splenectomy in the present study were ITP in 53 (82%), hereditary spherocytosis in four, lymphoma in three, autoimmune hemolytic anemia in three and thrombotic thrombocytopenic purpura (TTP) in one patient.

Regarding spleen size, the spleen was normal in 53 patients. When patients with splenomegaly were analyzed based on their diagnosis, one (2%) had ITP, four (6%) had thrombotic thrombocytopenic purpura (TTP), three (5%) had leukemia/lymphoma and three (5%) had autoimmune hemolytic anemia.

The present study included patients who underwent open and laparoscopic splenectomies. The patients had a shorter hospital stay, better palpation of pain and superior cosmetic outcomes with laparoscopic splenectomy.

There have been studies reporting the presence of an accessory spleen in 18-28% of the normal population (10, 18), and is the most common cause of recurrence in the surgical treatment of ITP. Thus, the presence of an accessory spleen should be investigated in ITP patients scheduled for splenectomy, and must be removed if detected. Although an accessory spleen can be found in a considerable proportion of the population, it is a significant factor that reveals the efficacy of surgical response in this group of patients (19). In the present study, 11 (17%) patients had an accessory spleen, but none developed recurrence. preoperative radiological methods Both and intraoperative explorations are important for the detection of an accessory spleen, and accordingly, for the prevention of recurrence.

Complication rates vary in patients undergoing splenectomy due to hematologic diseases. The most

postoperative complications include common bleeding, thromboembolism, atelectasis and wound site infection (3, 10). The risk of development of post-splenectomy infection is increased following splenectomies performed due to hematologic diseases (20). Vaccination is recommended within two weeks of an emergency splenectomy, although it is not known to be particularly protective after splenectomy. If not previously administered, a meningococcal vaccine and an annual influenza vaccine should be administered (21). In the present study, one patient developed wound site infection, while two patients were treated in the intensive care unit due to sepsis, however one died 13 days and the other died 25 days later due to sepsis.

An evaluation of post-splenectomy platelet counts revealed 57 (89%) patients with complete response, six (9%) with partial response and one (2%) with no response to splenectomy.

In conclusion, splenectomy can be performed safely for the treatment of resistant hematologic diseases. In order to reduce postoperative recurrence, the presence of an accessory spleen should be investigated, and must be removed if detected. Patients should be vaccinated against encapsulated bacteria to reduce infection rates. We believe splenectomy to be a good treatment option for the treatment of hematologic splenic diseases that are resistant to medical treatment, being associated with low mortality and morbidity.

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East J Med Volume:26, Number:2, April-June/2021