Assessment of Hand-port-assisted Laparoscopic Splenectomy for Massive and Supramassive Splenomegaly

Thesis

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By

Karim Gamal Moustafa Saied

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Supervised by

Prof. Dr. Amr Abdallah Mohsen
Professor of General Surgery
Faculty of Medicine, Cairo University

Dr. Ahmed Abd Elfattah Nada
Assistant Professor of General Surgery
Faculty of Medicine, Cairo University

Faculty of Medicine
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Abstract

The presence of a massively enlarged spleen is commonly a deterrent to its removal by the laparoscopic approach. The causes are multiple and include difficult manipulation, encroachment on the field, extraction problems and bleeding tendency, particularly in portal hypertension patients.

The use of open technique for these patients denies them the known advantages of laparoscopy.

The introduction of a surgeon's hand in the abdomen solves these problems. This is a thesis presentation that shows the technique of hand-assisted laparoscopic splenectomy for such patients. It also demonstrates how it overcomes the aforementioned obstacles.

Key words:

Splenomegaly, Hand-port, Laparoscopic splenectomy, HALS.

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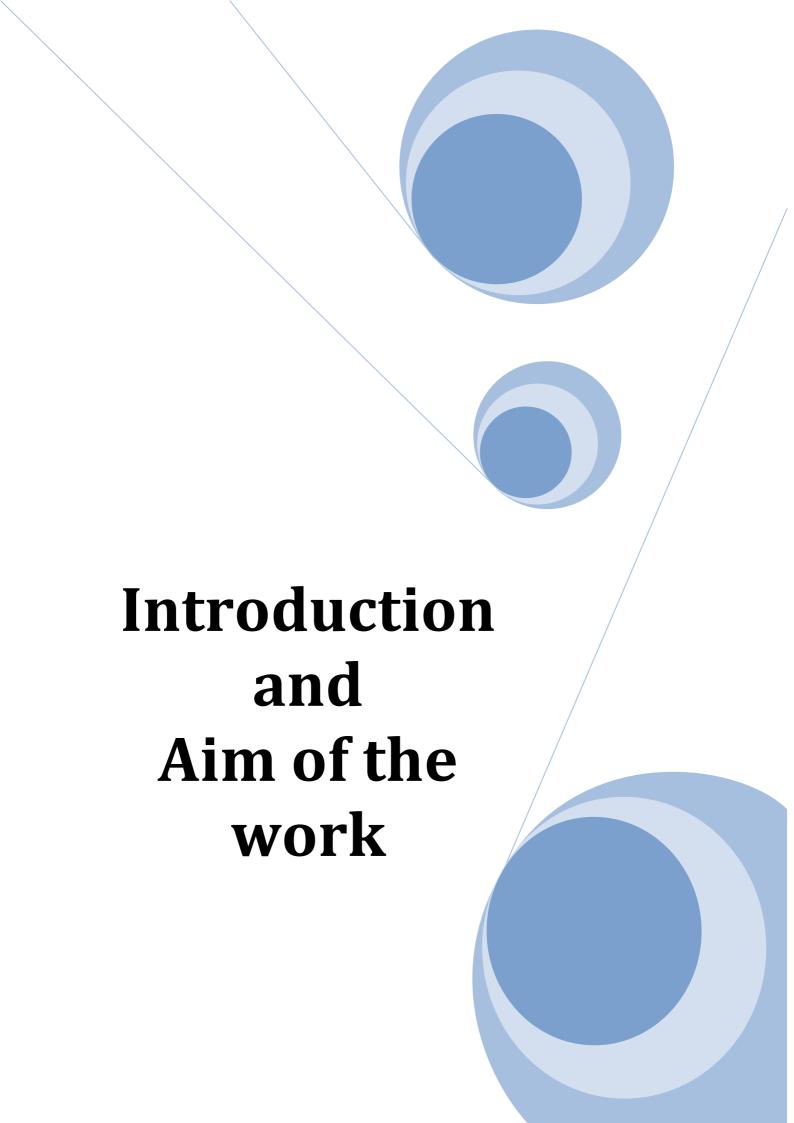
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List of abbreviations

Word	Abbreviation
Anti-immunoglobulin G	IgG
Central nervous system	CNS
Chronic lymphocytic leukemia	CLL
Chronic myelogenous leukemia	CML
Hand-assisted laparoscopic splenectomy	HALS
Immunogenic thrombocytopenic purpura	ITP
Intravenous immunoglobulin	IVIG
Laparoscopic splenectomy	LS
Low molecular weight heparin	LMWH
Magnetic resonance imaging	MRI
Open splenectomy	OS
Overwhelming postsplenectomy infection	OPSI
Thrombotic thrombocytopenic purpura	TTP



Splenomegaly should be defined in metric terms by preoperative imaging. The normal spleen weighs 150 to 200g and has a cranio-caudal length shorter than 12cm; thus, spleens weighing less than 600g or with a cranio-caudal length shorter than 17cm are considered nonmassive. Spleens weighing 600 to 1600g or with a cranio-caudal length of 17 to 22cm are classified as massive, and spleens weighing more than 1600 g or with a cranio-caudal length greater than 22cm are considered supramassive (Grahn et al., 2006).

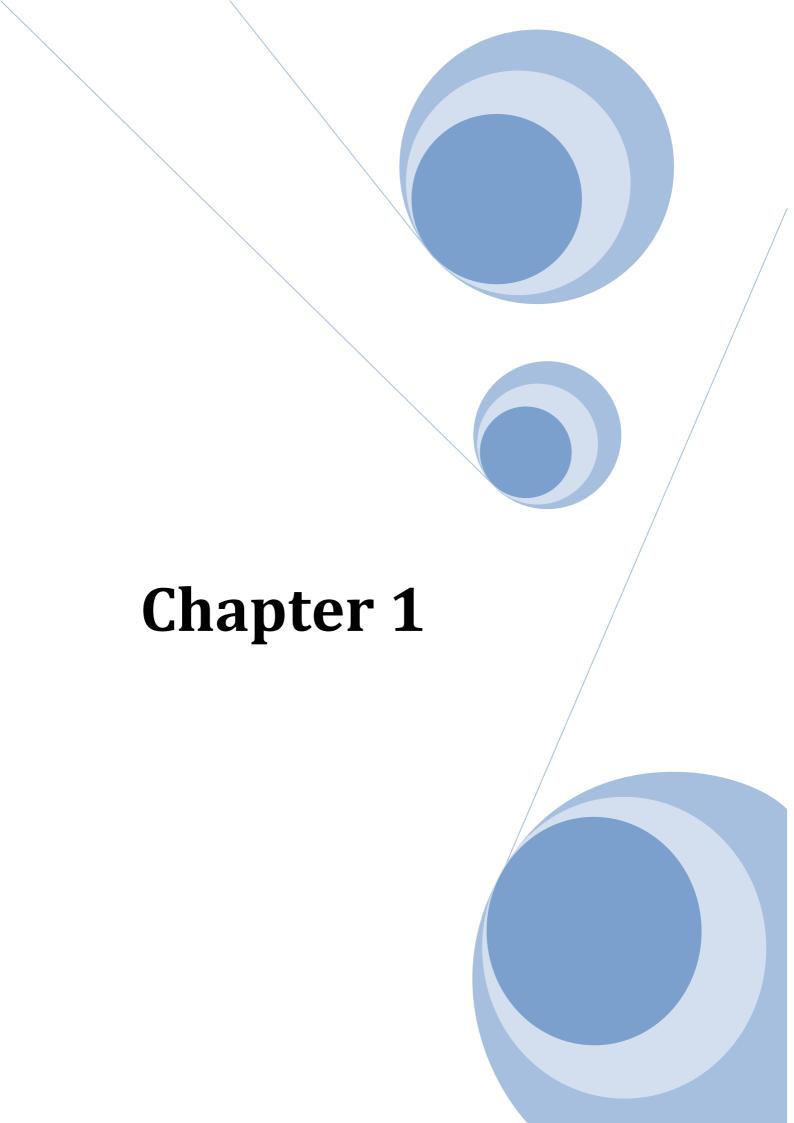
Laparoscopic splenectomy (LS) was first described by Delaitre et al. (1992). Since that time, several studies have demonstrated the advantages of the laparoscopic approach over open splenectomy (OS), including shorter hospital stay, decreased blood loss, faster recovery, and better quality of life outcomes. Success with normal sized and mildly enlarged spleens has led to increasing use of LS in patients with splenomegaly (Velanovich et al., 2000).

In another study, seven patients with massive spleens underwent successful laparoscopic resection; however, the safety of this approach has been disputed (Choy et al., 2004). Patel reported a 23% conversion rate, a 10 fold increase in morbidity and prolongation of the hospital stay with the laparoscopic approach in 27 patients with massive spleens (Patel et al., 2003).

Hand-assisted laparoscopic splenectomy (HALS) is a modification of LS and should be considered as a valid procedure to avoid conversion to open surgery. For massive and supramassive splenomegaly, HALS is recommended as a primary procedure because it shortens operative time and minimizes intraoperative blood loss (Habermalz et al., 2008).

Aim of the work

The aim of this work is to assess the outcome of using the hand port for laparoscopic excision of the massive and supramassive splenomegaly.



Chapter 1. General principles of splenectomy

Splenectomy is performed for numerous indications. Patients with benign hematologic disorders, especially, can benefit from this procedure. Formerly, open splenectomy represented the traditional method for normalizing platelet levels or for staging the extent of malignant disease (Gadenstatter et al., 2002).

Indications for splenectomy

The initial therapy for most hematologic disorders of the spleen is medical. In general, splenectomy is warranted after failure of medical therapy, as an adjunct to medical therapy, for diagnostic reasons, or in some cases as primary therapy for an underlying malignancy (Klingensmith et al., 2008).

Immunogenic thrombocytopenic purpura (ITP) and hemolytic anemias are the most common indications for elective splenectomy. Splenectomy may also be used for palliation as in chronic lymphocytic leukemia (CLL), hairy cell leukemia and Felty syndrome, primarily via the control of cytopenias. Patients with refractory cytopenias due to hypersplenism that require frequent transfusion or significantly limit the dose of chemotherapy may benefit from splenectomy. Patients with a massively enlarged spleen can develop early satiety, abdominal pain, and weight loss (Habermalz et al., 2008).

Furthermore, solid mass lesions in the spleen can be an indication for splenectomy, particularly if a malignant diagnosis is suspected. Splenectomy may be used to establish a diagnosis of lymphoma in the absence of more easily accessible tissue to be biopsed (Klingensmith et al., 2008).

Although splenic injury is increasingly managed conservatively, splenectomy is considered to be the definitive treatment for patients with traumatic splenic hemorrhage. In addition, splenic hemorrhage may rarely occur spontaneously in diseases such as infectious mononucleosis (Klingensmith et al., 2008)

Heamatological disorders

Immunogenic thrombocytopenic purpura (ITP) is an acquired disorder in which auto-antibodies are produced against a platelet glycoprotein. The spleen is the major site for the production of antiplatelet antibodies and the principal site for platelet destruction. Children usually present with acute ITP, often associated with a recent viral syndrome. In 90% of cases, the disease spontaneously remits within 6 to 12 months. Only refractory cases require splenectomy. Adults typically present with a more chronic form of ITP that is much less likely to spontaneously remit. Asymptomatic patients with platelet counts greater than 50,000/mm³ are candidates for only follow up. Symptomatic patients or those with counts less than 30,000/mm³ should be treated with oral glucocorticoids. Greater than 50% of patients respond to glucocorticoids. In refractory cases, or in patients with bleeding, intravenous immunoglobulin (IVIG) is used, although the effects are transient. Indications for splenectomy are failure to respond to medical therapy and intolerable side effects from steroid administration (Stasi and Provan, 2004).

Indications for more urgent splenectomy in patients with ITP include need for other emergency surgery or a life threatening bleed, including central nervous system (CNS) hemorrhage. The majority of patients who respond to splenectomy do so within the first 10 postoperative days. A systematic review study of 135 case series, found a complete remission in two thirds of patients. Either a complete or partial response was observed in 88% of patients. Complete response was defined as an achievement of the platelet count of at least 100×10^9 /L. A partial response included a platelet count response of at least 30×10^9 /L. Variables associated with a response to splenectomy include younger age and response to IVIG therapy. Mortality rates of 0.2% to 1% are similar to mortality due to medically treated severe ITP over 5 to 10 years (0.4% to 1.6%) and surgical complication rates range from 10% to 13% (Kojouri et al., 2004).

Patients with recurrence of symptoms after splenectomy or relapse after an initial response should be investigated for accessory splenic tissue. A peripheral smear and magnetic resonance imaging (MRI) or nuclear medicine studies with technetium (Tc)-99m-labeled heat-damaged red cells are indicated. If accessory splenic tissue is found,

re-exploration should be considered, although long-term response to removal of an accessory spleen is uncommon (Cines and Blanchette, 2002).

Thrombotic thrombocytopenic purpura (TTP) is characterized by the pentad of hemolytic anemia, consumptive thrombocytopenia, mental status changes, renal failure, and fever, although only unexplained hemolytic anemia and thrombocytopenia, are required to initiate therapy. Symptoms result from multi organ microvascular thrombosis. First-line therapy for TTP is medical, with plasmapheresis. A randomized, controlled trial done by Rock et al. (1991) with 102 patients demonstrated significantly improved initial response (47% vs. 25%) and 6 months survival (78% vs. 63%) for plasmapheresis as compared to plasma infusion. Glucocorticoids, in addition to plasmapheresis, are prescribed in rare cases of relapse. Splenectomy is indicated in patients who do not respond to medical therapy or those with chronically relapsing disease. A recent 20 years retrospective study of 33 patients, found that those who were continuously dependent on plasmapheresis and underwent splenectomy had a postoperative relapse rate of 0.07 per patient in one year period. In those patients who were not continuously dependent on plasmapheresis, splenectomy reduced the relapse rate from 0.74 to 0.10 per patient in one year period (Klingensmith et al., 2008).

Hemolytic anemias constitute a group of disorders in which splenectomy is almost universally curative. Hereditary spherocytosis is an autosomal dominant disorder in which there is a defect in spectrin, a red blood cell (RBC) membrane protein. This defect leads to small, spherical, relatively rigid erythrocytes that are unable to deform adequately to traverse the splenic microcirculation. This results in their sequestration and destruction in the splenic red pulp. In addition to anemia, patients may have jaundice from the hemolytic process and splenomegaly from RBC destruction in the spleen. Splenectomy is indicated in nearly all cases, but should be delayed to age 6 years in children to minimize the risk of overwhelming postsplenectomy infection (OPSI), but an exception to this rule is if the child is transfusion dependent. Prior to splenectomy, patients should have an abdominal ultrasound, and if gallstones are present, which are usually pigment stones from hemolysis, a cholecystectomy can be performed concomitantly (Klingensmith et al., 2008).

Acquired autoimmune hemolytic anemias are characterized as either warm or cold, depending on the temperature at which they interact with antibody. Warm autoimmune hemolytic anemia results from splenic sequestration and destruction of RBCs coated with autoantibodies that interact optimally with their antigens at 37°C. Anti-immunoglobulin G (IgG) antiserum causes agglutination of the patient's RBCs (positive direct Coombs test). Primary treatment is directed against the underlying disease, if this is unsuccessful, therapy is corticosteroids. Non-responders or patients requiring high steroid doses respond to splenectomy in 60% to 80% of cases. Cold autoimmune hemolytic anemia is characterized by fixation of C3 to IgM antibodies that bind RBCs with greater affinity at temperatures approaching 0°C and cause Raynaud-like symptoms combined with anemia. Hemolysis occurs either immediately by intravascular complement-mediated mechanisms or via removal of C3-coated RBCs by the liver. Treatment is usually successful with use of increased protective clothing and splenectomy has no therapeutic benefit (Habermalz et al., 2008).

.Sickle cell anemia is due to the homozygous inheritance of the S variant of the hemoglobin β chain. The disease is usually associated with autosplenectomy due to repeated vaso-occlusive crises, but splenectomy may be required for those patients with acute splenic sequestration crisis, evidence of hypersplenism, splenic abscess, and symptomatic splenomegaly. Thalassemias are hereditary anemias caused by a defect in hemoglobin synthesis. β-Thalassemia major is primarily treated with iron chelation therapy, but splenectomy may be required to treat symptomatic splenomegaly or pain from splenic infarcts (Klingensmith et al., 2008).

Polycythemia vera and essential thrombocytosis are chronic diseases of uncontrolled red blood cell and platelet production, respectively. These diseases are treated medically, but splenectomy is indicated to treat symptomatic splenomegaly or pain from splenic infarcts. Splenectomy can result in severe thrombocytosis, causing thrombosis or hemorrhage, which requires perioperative antiplatelet, anticoagulation, and myelosuppressive treatment. A case series study demonstrated success in surgical management of severe, symptomatic splenomegaly in patients with polycythemia vera with no long-term complications (Cunningham and Napolitano, 2004).

Malignancy

Myelofibrosis and myeloid metaplasia are incurable myeloproliferative disorders that usually present in patients older than 60 years. The condition is characterized by bone marrow fibrosis, leukoerythroblastosis, and extramedullary hematopoiesis, which can result in massive splenomegaly. Indications for splenectomy include symptomatic splenomegaly and transfusion-dependent anemias. Although the compressive symptoms are effectively palliated with splenectomy, the cytopenias frequently recur. In addition, these patients are at increased risk for postoperative hemorrhage and thrombotic complications after splenectomy (Klingensmith et al., 2008).

Chronic myelogenous leukemia (CML) is a myelodysplastic disorder characterized by the bcr-abl fusion oncogene, known as the Philadelphia chromosome. This oncogene results in a continuously active tyrosine kinase. First-line therapy is medical with imatinib mesylate, which targets the tyrosine kinase protein. Alternative chemotherapies are used in cases of intolerance or suboptimal response. Stem cell transplantation is used for cases of treatment failure in eligible patients. The Italian Cooperative Study Group prospectively randomized 189 patients in the early phase of CML to splenectomy plus chemotherapy or, chemotherapy alone. Splenectomy had no effect on the survival or the disease progression, but it did increase the rate of intravascular thrombosis. Thus, splenectomy is indicated only for palliation of symptomatic splenomegaly or hypersplenism that significantly limits therapy (Baccarani et al., 2006).

Chronic lymphocytic leukemia (CLL), a B-cell leukemia, is the most common of the chronic leukemias and is characterized by the accumulation of mature but nonfunctional lymphocytes. Primary therapy is medical, with splenectomy reserved for those patients with symptomatic splenomegaly and severe hypersplenism. Although the use of splenectomy in such cases is controversial, data from one retrospective study demonstrated that early splenectomy for hypersplenism or cytopenias in some patient subgroups was associated with improved survival. The non-Hodgkin lymphomas are a diverse group of disorders with a wide range of clinical behaviors, ranging from indolent to highly aggressive. As with other malignant processes, splenectomy is indicated for