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Review Article

Indications for Surgery in Non-Traumatic Spleen Disease

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Abstract

The spleen is the largest lymphatic organ that acts as a site for filtration of foreign particles from the blood, erythropoiesis and hematopoiesis. Splenectomy represents the first line of treatment for spontaneous splenic rupture, abscesses, cysts, tumours. It is also used to control hereditary, autoimmune, and myeloproliferative disorders alternatively. Numerous diseases have been indicated for surgery in non-traumatic spleen diseases such as non-traumatic spleen rupture, immune thrombocytopenic purpura (ITP), haemolytic anaemias, Felty's syndrome, Hodgkin's and non-Hodgkin's lymphoma among others. This result because the spleen is the most affected lymphoid organ following its overactivity that occurs during sequestration of dead or disrupted RBCs and lymphocytes. Abdominal pain is one of the major manifestations of splenomegaly, and can also designate other associated complications such as liver cirrhosis or bacterial endocarditis. As a secondary lymphoid organ, the spleen is more often an organ for lymphomas. Although splenectomy is a curative alternative in a few diseases, it is a complementary means of treating several other diseases. Splenectomy is a salvage therapy used when other therapeutic alternatives fail. Despite its indication in numerous diseases, controversies are still inbound of its use.

Introduction

As the largest of the lymphatic organs, the spleen also helps in the filtration of foreign matter from the blood and serves as a major site of erythropoiesis and hematopoiesis [1]. Weighing between 75 and 250g in healthy adults, its size decreases with age [2], [3]. The spleen also acts as a storage site for iron, erythrocytes and platelets; and produces antibodies that remove bacteria [4], [5], [6]. Non-traumatic spleen rupture is a rare condition and can occur in a pathological spleen due to a variety of diseases [7].

Indications for Surgery in Non-traumatic Spleen Disease

The most widespread indications for surgery in non-traumatic spleen disease includes conditions such as; immune thrombocytopenic purpura (ITP), haemolytic anaemias, malaria, thalassemia, splenic abscesses, congestive splenomegaly, splenic cysts, Felty's syndrome, Hodgkin's and non-Hodgkin's lymphoma, leukaemias, myelofibrosis e.t.c. [5], [6], [8]. Sreekar *et al.,* and Schlittler and Dallagasperina, in their study, reports that splenic abscess is a rare and potentially fatal disease found especially in men, with *Escherichia coli*, *Klebsiella pneumonia, Staphylococcus aureus* and *Salmonella typhi* as causative factors [9], [10]. They also noted neoplasia, splenic infarcts, diabetes mellitus and immunosuppressive conditions as possible risk factors associated with the disease. Another condition that has drawn attention for surgery is a splenic cyst. Splenic cysts, which is asymptomatic with greater incidence in women are benign and without solid components [11]. The aetiologies of splenic cysts are numerous and include congenital, post-traumatic pseudocysts, peliosis and cystic neoplasias such as lymphangioma, hemangioma and lymphoma [11], [12].

Surgery has also been indicated in spontaneous splenic rupture (SSR) or non-traumatic rupture of the spleen, a rare, lethal, but potentially

treatable condition [13]. With predisposing factors such as leukaemias, malaria, lymphomas, liver cirrhosis, rheumatoid arthritis, pancreatitis, e.t.c, SSR accounts for more than 20 percent mortality rate [14], [15]. SSR diagnosis is important in subjects with haematological malignancies accompanied by unexpected abdominal pain, hypotension and shock [13]. The autosomal recessive disorders (Thalassemias) where one or more globin chains are reduced, results in defective erythropoiesis, haemolysis, and consequent hypersplenism following overactivity of the spleen. The disease courses with a hypercoagulable state, thrombosis with the risk of thromboembolic complications [16]. Haemangioma is the most common, asymptomatic benign neoplasm of the spleen that is also indicated for surgery [17]. Splenic metastases which are rare, have 0.9-1.86 percent prevalence in breast cancer cases [18], [19].

Malaria is an important disease especially in tropical regions of the world caused by protozoa of the genus *Plasmodium spp.* (*falciparum, vivax, malariae, ovale* and *knowlesi*). This disease affects the fundamental function of the spleen, as the spleen removes dead cells or cells infested by parasites and returning intact erythrocytes to the blood. It has also been observed that asplenic and hypoplastic individuals are more susceptible to fatal progress of the disease. This disease can cause spontaneous splenic rupture and in rare cases, splenic infarction, which are indications for splenectomy [20], [21]. Another close relative in terms of anaemia, is sickle cell disease (SCD), a genetic disease of haemoglobin leading to tissue damage and anaemia.

One important complication of sickle cell disease is splenic sequestration, where red blood cells become entrapped in the spleen, causing the spleen to enlarge, pooling and resulting in the final destruction of red blood cells [22]. The spleen is also implicated in hereditary spherocytosis in that it is the site of sequestration and phagocytosis of non-deformable red cell, leading to anaemia [23]. This disease is characterised by pallor resulting from the anaemia, jaundice from the hyperbilirubinemia and splenomegaly. While autoimmune haemolytic anaemia is a disorder caused by autoantibodies directed against red blood cells, idiopathic thrombocytopenic purpura (ITP) with no specific cause is characterised by thrombocytopenia and microangiopathic haemolytic anaemia, diagnosed by the definite presence of schizocytes in the peripheral blood smear film. Majority of these conditions may cause spleen enlargement. Spleen enlargement (splenomegaly) can also be caused by cirrhosis of the liver, lymphoma, and acquired immunodeficiency syndrome (AIDS) and venous thrombosis. Splenomegaly is characterised by pain in the upper left quadrant referred to the shoulder and sensation of early satiety [24]. Surgery is indicated in cases of severe thrombocytopenia associated with spontaneous bleeding, post-transplant splenic sequestration, or abdominal pain due to repeated splenic infarction [25]. Chronic venous congestion of the spleen, most often caused by sinusoidal intra-hepatic cirrhosis or splenic artery aneurysm, which can produce chronic venous obstruction by direct compression of the splenic vein can also cause congestive splenomegaly (CS) [5], [26]. Splenectomy is indicated as the treatment of choice for patients with severe residual thrombocytopenia, with venous thrombosis as the underlying cause of CS. However, in splenic artery aneurysm, a satisfactory result has been obtained with a patient treated with aneurysm resection and/or complementary splenectomy [5], [26].

As a secondary lymphoid organ, the spleen is generally involved by lymphomas [27], [28]. While the primary splenic lymphomas (PSL) originate inside the organ, the splenic tissue is compromised by diffuse dissemination of Hodgkin's Lymphoma and Non-Hodgkin's Lymphoma in the secondary splenic lymphomas (Silva and Gunasekera, 2015). Among the diverse haematological malignancies, lymphoid and myeloid leukaemias are well-known. Persons with these diseases may develop considerable splenomegaly and then splenic sequestration, which is responsible for the worsening of anaemia and pre-existing thrombocytopenia in bone marrow failure, observed especially in leukaemias [29], [30]. Felty’s syndrome (FS) is a severe form of rheumatoid arthritis (RA) with longstanding, severe and erosive arthropathy. FS causes splenomegaly and neutropenia, which result from increased neutrophil sequestration, peripheral neutrophil destruction, and bone marrow failure to produce neutrophils. Splenomegaly which is manifested by abdominal pain from splenic infarcts, can indicate other complications such as liver cirrhosis or other infections, like bacterial endocarditis [9].

Surgery is indicated in the majority of these non-traumatic diseases affecting the spleen. This is because; the spleen is an important lymphoid organ, whose role in erythropoiesis, immunity cannot be denied, as it is often enlarged (splenomegaly) following excessive activity from sequestration of old and damaged RBCs and lymphocytes. This overactivity affects the blood supply to the spleen, increasing its risk of infection.

Complications/Laparoscopy

The major complications of surgery which may involve removing the spleen (splenectomy) include haemorrhage, thromboembolism, subphrenic abscess, thoracic infection and fulminate sepsis. The greater mortality rates are majorly due to haemorrhage, bacterial infections and myelofibrosis [4]. The risk of devastating post-splenectomy infection (OPSI) is both more common and is characterised by

hypotension, altered consciousness or cardio-circulatory shock.

Conclusion

Splenectomy, despite being indicated in several conditions, is still controversial and remains open to further studies. Although splenectomy is a curative alternative in a small number of diseases, it is a complementary treatment in numerous other clinical disorders. While its purpose and its effects on the host's homeostasis are not fully understood. Unlike its indication in trauma, it is compromised by chronic disease and with the use of corticosteroids; immunity is reduced in these patients. Clotting disorders, changes in platelet function and associated diseases are common. This exposes these individuals to serious risks and complications, greater morbidity and mortality rates compared to other intra-abdominal surgical procedures. Therefore, as a serious surgical procedure, splenectomy should be undertaken only after the depletion of the clinic and non-invasive therapeutics.

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