Indications for Surgery in Non-Traumatic Spleen Disease

Danilo Coco¹ ², Silvana Leanza³

¹Ospedali Riuniti Marche Nord, Pesaro, Italy; ²Ospedale Augusto Murri - Fermo, Fermo, Italy; ³Carlo Urbani Hospital, Jesi, Italy

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 in bound 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 Schilttler 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 lymphangiomata, hemangiomata 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, etc., 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 intrahepatic 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 (OPSIs) 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.

References

1. Engwerda CR, Beattie L, Amante FH. The importance of the spleen in malaria. Trends Parasitol. 2005; 21:75-80. https://doi.org/10.1016/j.pt.2004.11.008 PMid:15664530
2. Vallabhaneni S, Scott H, Carter J, Treseler P, Machtinger EL. The importance of the spleen in non-invasive procedures. therefore, as a serious surgical procedure. Splenectomy should be undertaken only after the depletion of the clinic and non-invasive therapeutics. https://doi.org/10.1097/JSP.0b013e318277b009 PMid:19062140
3. Pozo AL, Godfrey EM, Bowles KM. Splenomegaly: investigation, diagnosis and management. Blood Rev. 2009; 23:105-111. https://doi.org/10.1016/j.bloodre.2008.10.001 PMid:19062140
4. Gomes CA, Junior CS, Coccolini F, Montori G, Soares AA, Junior CP, Filho FVM, Mendonça PRH, Gomes FC. Splenectomy in non-traumatic diseases. Australian Medical Journal. 2018; 11(5):295-304. https://doi.org/10.21767/AMJ.2018.3306
5. Weledji EP. Benefits and risks of splenectomy. Int J Surg. 2014; 12(2):112-9. https://doi.org/10.1016/j.ijsu.2013.11.017 PMid:24362830
6. Rodeghiero F, Ruggeri M. Short and long-term risks of splenectomy for benign haematological disorders: should we revisit the indications? Br J Haematol. 2012; 158(suppl 1):29. https://doi.org/10.1111/j.1365-2142.2012.09146.x PMid:22571181
7. Hadary A, Dashkovsky I, Ravaport A, Cozakov JC. Non-traumatic rupture of spleen: can splenectomy be applied selectively? The Israel Medical Association Journal. 2008; 10(12):889-91.
8. Browning MG, Bullen N, Nokes T. The envolving indications for splenectomy. Br J Haematol. 2017; 177:321-4. https://doi.org/10.1111/bjh.14060 PMid:27018168
9. Sreekar H, Saraf V, Pangi AC. A retrospective study of 75 cases of splenic abscess. Indian J Surg. 2011; 73:398-402. https://doi.org/10.1007/s12262-011-0370-y PMid:23204694 PMcid:PMC3236272
10. Schiltzler LA, Dillagasperina VW. Cistos esplênicos nãoparasitários. Rev Col Bras Cir. 2010; 37:442-6. https://doi.org/10.1590/S0100-69912010000600011
11. Silva WT, Gunasekera M. Spontaneous splenic rupture during the recovery phase of dengue fever. BMC Res Notes. 2015; 8:1-4. https://doi.org/10.1186/s13104-015-1234-5 PMid:26136216 PMcid:PMC489041
12. Kaza RK, Azar S, Al-Hawary MM. Primary and secondary neoplasms of the spleen. Cancer Imaging. 2010; 10:173-82. https://doi.org/10.1107/1470-7330.2010.0028 PMid:20713317 PMcid:PMC2943678
13. Chejra RK, Satish KA, Arya SV and Bajwa JS. Initial Presentation of Chronic Myeloid Leukaemia. Open Access J Surg. 2017; 7(2):001-003.
14. Rebzulli P, Hostettler A, Schoepfer AM. Systematic review of atraumatic splenic rupture. Br J Surg. 2009; 96:1114-21. https://doi.org/10.1002/bjs.5737 PMid:19787754
15. Thapar PM, Philip R, Masurkar VG. Laparoscopic splenectomy for spontaneous rupture of the spleen. J Minim Access Surg. 2016; 12:75-8. https://doi.org/10.4103/0972-9941.158950 PMid:26917926 PMcid:PMC4746982
16. Weatherall DJ. The inherited diseases of hemoglobin are an emerging global health burden. Blood. 2010; 115:4331-6. https://doi.org/10.1182/blood-2010-01-251348 PMid:20233970 PMcid:PMC2881491
17. Despoina M, Dionysios D, Georgios A. Primary Angiosarcoma of the Spleen: An Oncological Enigma. Case Rep Oncol Med. 2014; 2014:93038. https://doi.org/10.1155/2014/93038 PMid:25105042 PMcid:PMC4101939
18. Moncólova J, Sloer ET, Solís YP. Laparoscopic Approach for Isolated Splenic Metastasis: Comprehensive Literature Review and Report of 6 Cases. Surg Laparosc Endosc Percutan Tech. 2013; 23:21-4. https://doi.org/10.4103/0972-9941.158950 PMid:23396144
19. Owusu-Ofori S, Hirst C. Splenectomy versus conservative management for acute sequestration crisis in people with sickle cell disease. Cochrane Database Syst Rev. 2013; 31(5):CD009425. https://doi.org/10.1002/14651858.CD009425.pub2
20. Browning MG, Bullen N, Nokes T. The envolving indications for splenectomy. Br J Haematol. 2017; 177:321-4. https://doi.org/10.1111/bjh.14060 PMid:27018168
21. Bulus H, Mahmoud H, Altun H. Outcomes of laparoscopic versus open splenectomy. J Korean Surg Soc. 2013; 84:38-42. https://doi.org/10.4147/kjss.2013.84.1.38 PMid:23332324 PMcid:PMC3039108
22. Mourea R, Sobreira ML, Jaldin RG. Aneurisma sacular de artéria escamosa: tratamiento endovascular ou cirúrgico convencional? J Vasc Bras. 2013; 12:230-3. https://doi.org/10.1590/jvb.2013.045
23. Bolton-Maggs PH, Langer JC, Iolascon A. Guidelines for the diagnosis and management of hereditary spherocytosis--2011 update. Br J Haematol. 2012; 158:37-49. https://doi.org/10.1111/j.1365-2142.2011.08921.x PMid:21905020
24. Booyer TD, Habib S. Big spleens and hypersplenism: fix it or forget it? Liver Int. 2015; 35:1492-8. https://doi.org/10.1111/liv.12702 PMid:25312770
25. Gangireddy VG, Kanneganti PC, Sridhar S. Management of thrombocytopenia in advanced liver disease. Can J Gastroenterol Hepatol. 2014; 28(10):558-64. https://doi.org/10.1155/2014/532191 PMid:25222481 PMcid:PMC4234356
26. Ingle SB, Hinge Ingle CR. Primary splenic lymphoma: Current diagnostic trends. World J Clin Cases. 2016; 4(12):385-389. https://doi.org/10.12998/wjcc.v4.i12.385 PMid:23055311 PMcid:PMC1556875
27. Bickenbach KA, Gonen M, Labow DM. Indications for and efficacy of splenectomy for haematological disorders. Br J Surg. 2013; 100:794-800. https://doi.org/10.1002/bjs.9067 PMid:23436838
28. Rialon KL, Speicher PJ, Ceppa EP. Outcomes following diagnostic trends. World J Clin Cases. 2016; 4(12):385-389. https://doi.org/10.12998/wjcc.v4.i12.385 PMid:23055311 PMcid:PMC1556875
29. Tefferi A. Primary myelofibrosis: 2013 update on diagnosis risk-stratification, and management. Am J Hematol. 2013; 88:141-50. https://doi.org/10.1002/ajh.23384 PMid:23439007
30. Tefferi A. Pathogenesis of myelofibrosis with myeloid metaplasia. J Clin Oncol. 2005; 23:8520-30. https://doi.org/10.1200/JCO.2004.09.9316 PMid:16293880