CHAPTER 86
SPLEEN

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Introduction

The spleen, once thought to be a nonessential organ, has important functions in bacterial clearance, antibody formation, phagocystosis, and haematopoiesis. Aside from trauma, surgical intervention is mainly limited to haematologic diseases that affect the function of the spleen.

Demographics

The most common functional disease of the spleen is found in sickle cell disease and thalassaemic children in Africa. Splenomegaly is present in 21% of sickle cell disease patients in Nigeria.[1] Lebanese sickle cell disease children have a high prevalence (28.9%) of persistent splenomegaly.[2]

Aetiology/Pathophysiology

The spleen develops alongside the pancreas from mesenchyme of the dorsal pancreatic bud and the dorsal mesogastrium. The splenic primordium is first recognised at the 8–10 mm stage as a mesenchymal bulge in the left dorsal mesogastrium between the stomach and pancreas. By the fourth month of foetal life, the spleen produces both red and white cells, and it functions as a site of extramedullary haematopoiesis until approximately 1–2 months of age. This function gradually declines, as the spleen is rarely the site of clinically significant haematopoiesis in childhood.[3,4]

Anatomy and Function

Blood enters the spleen through segmental arteries that originate from the splenic artery and branch into trabecular arteries that eventually enter the white pulp. The white pulp consists of lymphocytes and macrophages arranged in germinal centers around a central artery. The white pulp is thought to function as an immune screening area that processes foreign antigen and antigen-antibody complexes. Approximately 20% of splenic volume is white pulp, with the remainder contributing to red pulp. The red pulp acts as a phagocytic filtration system that digests defective blood elements, foreign material, and bacteria. Encapsulated bacteria can be removed from the bloodstream by the spleen because of its variable antigenicity. The spleen functions in red blood cell maintenance, immune function, and as a reservoir.[3,4]

The spleen destroys red blood cells at the end of their life-span and repairs other damaged red cells. It selectively removes abnormal cells, such as spherocytes. As blood passes through the spleen, cellular and nuclear inclusion bodies are removed (i.e., Howell-Jolly bodies, Heinz bodies, and Pappenheimer bodies). After splenectomy, these inclusion bodies can be noted on peripheral blood smears.

The spleen's primary immune function is related to antigen processing of T cells. It also performs nonspecific immune functions by removing particulate matter from the bloodstream, primarily via macrophages. The spleen contains opsonins, which further activate the complement system and facilitate the destruction of organisms. The spleen functions as a biologic filter, which can assist in the clearance of bacteria. The spleen is a significant reservoir for platelets and factor VIII in the human body. The proportion of platelets within the spleen increases with splenomegaly and can occasionally lead to thrombocytopaenia.

Clinical Presentation

Aside from infections, injury, and trauma, splenic disorders can be classified into anatomic and functional abnormalities. Anatomic abnormalities are rarely the cause for surgical intervention, but may be associated with other surgical problems. Consequently, it is important for the paediatric surgeon to be aware of these conditions.

Congenital asplenia is usually associated with congenital heart disease, bilateral "right-sidedness" with bilateral three-lobed lungs, right-sided stomach, and a central liver. Patients with these disorders may have intestinal malrotation and are prone to overwhelming infections. Polysplenia, in which the spleen is divided into multiple splenic masses, may be associated with biliary atresia, preduodenal portal vein, situs inversus, cardiac defects, and malrotation.

Accessory Spleen

Accessory spleen is the most common anatomic abnormality, occurring in 13–30% of all children. Of those patients with accessory spleens, 80% have one, 11% have two, and 3% have three or more accessory spleens.[3,5,6] Nearly 75% of accessory spleens are located near the splenic hilum; the next location in terms of percentages is by the tail of the pancreas. Accessory spleens may also occur along the splenic artery, omentum, mesentery and retroperitoneum. When performing a splenectomy, it is important to routinely examine for accessory spleens because, if missed, they can account for recurrence of idiopathic thrombocytopaenic purpura (ITP) or hereditary spherocytosis.

Wandering Spleen

Wandering spleen is an anatomic abnormality characterised by the lack of ligamentous attachments to the diaphragm, colon, and retroperitoneum. The problem is related to a failure of development of the ligaments from the spleen to the dorsal mesentery. Children may present with episodic pain and abdominal mass from torsion and infarction of the spleen. Splenopexy is the preferred treatment; however, patients may require splenectomy for infarction. Among the variety of techniques of splenopexy are an extraperitoneal pocket, absorbable mesh bag or basket, or suture splenopexy. Laparoscopic techniques have also been described.

Splenogonadal Fusion

During embryologic development, early fusion between the spleen and the left gonad can occur, causing what is known as splenogonadal fusion. This developmental abnormality is often found with limb or anorectal abnormalities, indicative of some sort of embryonic event occurring between weeks 5 and 8 of development. Three types of fusion have been described: continuous, discontinuous, and combined. In the first type, a continuous cord consisting of splenic or fibrous tissue connects the spleen to the left gonad, typically originating from the upper pole of the spleen. The discontinuous type is characterised by ectopic splenic tissue fused to the left gonadal mesonephric structures, which are discrete from the spleen itself. The combined form is characterised by an extension of functioning splenic tissue without any actual connection to gonadal tissue. Male patients with this disorder may have an undescended testicle or may be presumed to have an inguinal hernia.

Splenic tissue may be mistaken for a tumour attached to the testicle and in most cases may be safely dissected off the testicle, thereby avoiding an unnecessary orchiectomy.

Splenic Cysts

Benign splenic cysts are rare. When they are asymptomatic, they are typically found incidentally during imaging for another reason. Splenic cysts can be classified by appearance as uni- or multilocular, or by origin: congenital, infectious (parasitic), or posttraumatic. Congenital cysts are typically unilocular with a squamous (epithelial) or endothelial cell lining filled with clear fluid. Infectious cysts are typically parasitic, originating from echinococcus (hytadid cysts). Echinoccocal cysts should be handled carefully during removal, and total splenectomy may be necessary in order to prevent rupture of the cyst. Posttraumatic cysts result from a resolving haematoma. Liquefaction of the haematoma forms a pseudocyst with a fibrous lining. Treatment for congenital or posttraumatic cysts is indicated only if they become symptomatic, which usually occurs at a size greater than 8 cm. Patients may present with pain, rupture, abscess of a previously uninfected cyst, or gastric compression. Treatment may consist of percutaneous drainage or sclerosis of the cyst cavity, but both congenital and posttraumatic cysts have a high rate of recurrence. Partial or total splenectomy may be necessary for primary treatment, or in treatment of recurrent symptomatic cysts.

Splenic Abscess

Splenic abscess is a rare occurrence in children, and usually is a result of secondarily infected haematoma, congenital or posttraumatic cysts, or an area of infarct. Patients with splenic abscess typically present with bacteraemia, fever, and left upper quadrant pleuritic pain, and they may be toxic. A computed tomography (CT) scan is usually diagnostic, and ultrasound may be useful as well. Unilocular abscesses can be drained percutaneously and combined with antibiotic therapy. Multilocular abscesses may respond to antibiotics alone. Patients with hemoglobinopathy and splenic infarct typically have infection with *Salmonella* species. Staphylococcus and streptococcus are more common in posttraumatic infections. Fungal infection may occur in immunosuppressed patients who have negative blood cultures. Empiric antibiotics should be started as soon as the diagnosis is suspected and can be tailored to blood culture and abscess drainage cultures as the results become available.

Hyperactive Malarial Splenomegaly

Hyperactive malarial splenomegaly (HMS) is thought to represent an immunological dysfunction of the spleen due to recurrent episodes of malaria. Specific criteria for the diagnosis are gross splenomegaly, high levels of antimalarial antibodies, IgM in serum at least two standard deviations above the local mean, and clinical and immunological response to antimalarial treatment. Detection of circulating malaria parasites by polymerase chain reaction (PCR) may represent a useful diagnostic tool.[7] In hospitals that do not have access to these laboratory tests, the diagnosis is made by exclusion or response to treatment.

Functional Abnormalities

Abnormalities in splenic function can be categorised broadly as hypersplenism or hyposplenism. Hypersplenism refers to the inappropriate sequestration of blood elements within the spleen and may be a primary or secondary occurrence. Primary hypersplenism is generally caused by hereditary spherocytosis, elliptocytosis, and idiopathic thrombocytopenic purpura. Secondary hypersplenism is most commonly caused by portal hypertension, but any cause of splenomegaly can result in hypersplenism. Treatment of secondary hypersplenism may consist of splenic artery embolisation or ligation, although if these options are not available, splenectomy may be necessary.

Hyposplenism occurs with splenectomy or sickle cell disease with splenic infarcts and eventual involution, and it has also been seen with ulcerative colitis. These patients are at risk of infection by encapsulated organisms. Hyposplenism may be detected by Howell-Jolly bodies present in erythrocytes on peripheral blood smear, and patients may present with overwhelming sepsis. Early treatment with broad-spectrum antibiotics is essential in the face of sepsis if this diagnosis is suspected.

Hereditary Spherocytosis

Hereditary spherocytosis is an autosomal dominant hereditary disorder that is thought to be due to a spectrin deficiency. Spectrin is a cytoskeletal protein. The erythrocytes take on an abnormal spherical shape, have decreased cell wall flexibility, and are prematurely sequestered in the spleen. Index cases in a family may present with anaemia, jaundice, and splenomegaly; indirect hyperbilirubinaemia may occur in infants. The estimated incidence of spherocytosis in Algiers was reported as 1 in 1,000, with approximately the same features among Algerians as in people of European heritage.[8] It is very rare in sub-Saharan Africans and African Americans, with few literature reports.[9,10]

Diagnosis is made by demonstrating increased osmotic fragility of erythrocytes. Patients should also undergo a Coombs' test to rule out other immune causes of haemolytic anaemia. An aplastic crisis can occur in affected children from a parvovirus B19 infection, which results in suppression of erythrocyte production and severe anaemia due to ongoing erythrocyte destruction. Splenectomy is generally recommended for moderate to severe anaemia. Pigmented gallstones are a common occurrence in patients with hemolysis, so a preoperative ultrasound should be performed so that a concomitant cholecystectomy can be planned, if necessary. Hereditary elliptocytosis is a similar disorder, but most patients are asymptomatic and rarely require splenectomy.

Idiopathic Thrombocytopaenic Purpura

ITP is an autoimmune disorder of unknown aetiology. Antiplatelet autoantibodies, typically IgG, bind to platelet-associated antigens and cause destruction of platelets via phagocytosis within the spleen. Acute ITP occurs most commonly in children younger than 10 years of age after an acute viral illness. Symptoms are related to the severe thrombocytopaenia that results. The majority of patients have an acute form of ITP, which can generally be treated conservatively with restricted activity, corticosteroids, and intravenous immunoglobulin (IVIG). Medical treatment is indicated for platelet counts below 40,000/mm^3 or in older children with a higher risk of intracranial haemorrhage. Ten to twenty percent of patients with acute ITP will have persistence of their symptoms for more than 6 months, which is considered to represent chronic ITP. For patients who fail medical therapy, splenectomy may be an option, but failure to respond to either steroids or IVIG is also associated with a 70% chance of failure rate with splenectomy.[3,11] ITP in children is usually a self-limited disorder, and splenectomy should be considered only for chronic cases.

Sickle Cell Disease

Sickle cell disease occurs when valine is substituted for glutamic acid on the β-chain of hemoglobin A, resulting in hemoglobin S. The red cells are prone to become rigid and sickle in a hypoxic and acidic environment, such as in the spleen. The sickled cells block capillaries and can cause infarction distal to the obstruction, which can result in painful crises and, with regard to the spleen, areas of infarction. The spleen initially becomes large; then, with progressive infarction, slowly atrophies and produces a functional autosplenectomy. Persistent gross splenomegaly is found in patients with anaemic crises.[12] Acute splenic sequestration crisis (ASSC) can occur and is the most common indication for splenectomy. Classic presentation of patients with ASSC is a rapidly enlarging spleen with severe anaemia or hypotension and associated hypersplenism and thrombocytopaenia.[1] ASSC may be so severe that it causes circulatory collapse; it carries a high mortality rate.

Thalassaemia

Thalassaemia is a group of haemoglobinopathies related to abnormal production of α or β chains of haemoglobin that may be classified as of major (homozygous), minor (heterozygous), or intermediate form. Significant erythrocyte sequestration may occur within the spleen, and

splenectomy has been utilised in an attempt to decrease transfusion requirements. Partial splenectomy may have benefit to these patients because postsplenectomy sepsis occurs in approximately 10% of these patients who have undergone total splenectomy.

Leukaemia and Lymphomas

The spleen is often involved in both leukaemia and lymphoma, and may lead to splenomegaly without hypersplenism. Splenectomy is typically not indicated in these patients and plays no therapeutic role. In chronic myelogenous leukaemia, patients may have pain, mass-related symptoms, and secondary hypersplenism, and splenectomy may be performed for palliation of symptoms. In acute myelogenous leukaemia (AML) and acute lymphocytic leukaemia (ALL), the degree of splenomegaly is much less. In these cases, splenectomy is rarely of benefit and may be detrimental in those in which the spleen has become an important site of extramedullary haematopoiesis.

Gaucher's Disease

Gaucher's disease is an autosomal recessive metabolic disorder caused by a deficiency of the enzyme β-glucocerebrosidase, which causes the body to accumulate glucosylceramide in the macrophages of the spleen, liver, lungs, and bone marrow. Patients may have massive splenomegaly that may cause hypersplenism. Splenectomy has been performed in these patients to improve erythrocyte, leukocyte, and platelet counts and to alleviate the hypersplenism. Partial splenectomy may help improve symptoms temporarily, but recurrence seems to be the rule.

Splenosis

Splenic trauma may result in fragmentation of the spleen with subsequent growth of splenic tissue within the abdominal cavity. This event is called splenosis. The splenic implants do not function in the same capacity as the entire spleen would, and specifically they do not clear encapsulated organisms. Consequently, patients with splenosis should be treated with immunisations and antibiotics as if they were asplenic.

Investigations

The spleen and splenic disorders can be visualised with many different imaging modalities. Ultrasound is often the easiest and safest way to visualise the spleen and can be useful for initial evaluation of abdominal masses. Cysts are well visualised with ultrasound. Splenic trauma, lacerations, or haematomas are not reliably diagnosed with ultrasound, and CT may be necessary. With limited resources, ultrasound would likely be the most beneficial imaging study for the paediatric surgeon and can be easily done at the bedside.

Management

Nonoperative management is the treatment of choice for a child with a blunt splenic injury who is hemodynamically stable. However, in areas of Africa where CT scans are not available, surgical management with splenorrhaphy or splenectomy predominates.[13,14] When elective splenectomy is planned, patients should receive three immunisations two weeks prior to surgery to provide adequate time for immunologic response. The immunisations include *Streptococcus pneumonia*, meningococcus, and *Haemophilus influenza B*. Preoperative transfusion may be necessary in sickle cell disease to improve anaemia and to reduce the hemoglobin S level to less than 30% to avoid complications related to bleeding or heart failure.

Open Splenectomy

The operative approach to splenectomy has recently become more variable, depending on the reason for surgery and the surgeon's operative experience.

1. The traditional open splenectomy is approached with the patient in the supine or right decubitus position with a midline, transverse, or left subcostal incision.

2. The short gastric vessels are divided (often with suture ligation to avoid the potential catastrophic event of a tie slipping off with haemorrhage) and the spleen mobilised by dividing the diaphragmatic, colic, and renal attachments. In cases of severe splenomegaly, it may be necessary to open the gastrocolic ligament laterally to expose and ligate the splenic artery at the superior aspect of the pancreas in order to decrease blood loss with further mobilisation.

3. The spleen, when fully mobilised, is lifted up into the wound to allow safe division of the splenic artery and vein at the hilum. The tail of the pancreas may extend into the splenic hilum. Care should be taken when dividing the hilar vessels to avoid injury to the pancreas.

4. Once the spleen has been removed, the splenic bed is inspected for hemostasis. A nasogastric tube is often left in the stomach to provide gastric decompression in an attempt to avoid bleeding from the short gastric vessels.

5. A careful search is made for any accessory spleens, especially if the indication for surgery is hemolytic anaemia or ITP.

Laparoscopic Splenectomy

The laparoscopic approach to splenectomy began in 1991 and has become the preferred method for many institutions for elective cases. Benefits include decreased pain, shorter hospital length of stay, and improved cosmesis. There is, however, a steep learning curve with this procedure, and the procedure can be quite challenging when splenomegaly is present.

1. A lateral approach is typically used, with the patient's left side up at approximately 45°. The table is rotated to the patient's left for trocar placement, then back to the right to achieve a right lateral decubitus position. Four ports are often used: three in the midline or just off to the right (upper, mid, and umbilicus) and one in the left midabdomen. Two grasping instruments are usually used in the upper ports to provide traction and elevate the spleen.

2. The harmonic scalpel is typically used to divide the short gastric vessels and other splenic attachments.

3. The endostapler is used to ligate the hilar vessels, and the spleen is placed in an endocatch bag and removed via morselisation through the largest port site, typically in the left midabdomen.[3]

The rate of conversion to open procedure has been reported up to nearly 3% in several large series,[3] with splenomegaly and bleeding as the most common reasons. It is imperative to search for accessory spleens, especially if the indication for splenectomy is ITP or hemolytic anaemia. Studies in paediatric patients have not detected a significant difference in the detection rate of accessory spleens in either open or laparoscopic procedures.[3] Patients may recover somewhat faster with less pain after laparoscopic splenectomy, but there is a steep learning curve, and operating room time is generally longer for the laparoscopic procedure. This may not be a feasible option for hospitals with limited resources and laparoscopic capabilities.

Partial Splenectomy

The segmental blood supply of the spleen allows for partial splenectomy along the lines of demarcation that are apparent after vessel ligation. The procedure has been done both open and laparoscopically, and has been primarily used for hemolytic anaemia. It has also been used for trauma, splenic cysts, haemangiomas, and hamartomas. Approximately 25% of the normal splenic remnant is left after successful partial splenectomy. If the spleen is totally mobilised, the remnant should be fixed to adjacent structures or the retroperitoneum to prevent torsion. Patients should receive preoperative immunisations and prophylactic antibiotics postoperatively until splenic immune competence is noted.

Postoperative Complications

Postoperative complications are rare after splenectomy, but thrombosis may occur in the splenic, portal, or mesenteric veins. This should be considered in the evaluation of postsplenectomy abdominal pain. Splenosis may occur after splenectomy if spillage of splenic contents occurred during the operation. This may simulate intraabdominal tumour implants and present as small bowel obstruction, haemorrhage, or relapse of haematologic disorder.[15]

Postsplenectomy sepsis (PSS) is the most worrisome complication of splenectomy and is thought to be due to decreased immunoglobulin levels as well as decreased clearance of encapsulated organisms. The overall incidence of PSS is 4%, with a higher incidence in patients with thalassaemia (nearly 10%), and mortality approaches 50%.[4,16] Patients typically have a fulminant infection caused by encapsulated bacteria (pneumococcus, *Haemophilus influenza B*, gonococcus, and *Escherichia coli*). Empiric antibiotics should be initiated as soon as possible in a patient with fever and illness after splenectomy.

Ideally, patients should receive immunisations prior to splenectomy, and booster immunisations are recommended every 5 to 10 years. Prophylactic antibiotics are recommended for patients after splenectomy, and the duration varies from 2 years to a lifetime. The risk of PSS declines significantly after 2 years from splenectomy.[17] There have also been case reports of severe malarial infections and fatality in patients after splenectomy such that lifelong prophylaxis is recommended for patients living in endemic areas.[14]

Prevention

There are no preventive measures to take regarding this disease process. Once a decision for elective splenectomy has been made, obtaining the immunisations preoperatively would be of highest importance in terms of reducing the risk of PSS as well as the availability of prophylactic postoperative antibiotics. Preoperative transfusion may be necessary in sickle cell disease, as previously mentioned.

Ethical Issues

The availability of postoperative health care may be of paramount concern in countries with decreased health resources. This extends to the availability of immunisations as well as antibiotics. Prophylactic antibiotic use after splenectomy may be problematic when both financial and pharmaceutical resources are scarce for the patients' families and their health care providers. Patients with symptomatic splenomegaly may have to wait until the severity of their symptoms outweighs the risks associated with asplenia.

Key Summary Points

1. The spleen is an important immunologic intraabdominal organ that functions in bacterial clearance, antibody formation, and phagocytosis.
2. Surgical splenectomy is mainly limited to haematologic diseases.
3. Both open and laparoscopic approaches are feasible for splenectomy, depending on the surgeon's experience and hospital resources.
4. Preoperative transfusion may be necessary in sickle cell disease to improve anaemia and to reduce the hemoglobin S level to less than 30% to avoid complications related to bleeding or heart failure.
5. Preoperative immunisations against *Streptococcus pneumonia*, meningococcus, and *Hemophilus influenza B* are of paramount importance to help reduce the risk of postsplenectomy sepsis.
6. Prophylactic antibiotics are recommended postsplenectomy for at least 2 years, and often for a lifetime, depending on the patient's clinical condition.
7. Postsplenectomy sepsis is the most feared complication, with an overall incidence of 1–4%; the risk decreases after 2 years from splenectomy.
8. Splenectomy may predispose to severe or even fatal malaria infestation, and lifelong prophylaxis is recommended for patients living in malaria endemic areas.

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