Splenomegaly

When sufficiently enlarged, the spleen causes a dragging sensation in the left upper quadrant and, through pressure on the stomach, discomfort after eating. In addition, enlargement can cause a syndrome known as *hypersplenism*, which is characterized by anemia, leukopenia, thrombocytopenia, alone or in combination. The probable cause of the cytopenias is increased sequestration of formed elements and the consequent enhanced phagocytosis by the splenic macrophages.

Table 13-12 lists major disorders associated with splenomegaly. Splenomegaly in virtually all the conditions mentioned has been discussed elsewhere. A few disorders are left to consider.

Nonspecific Acute Splenitis

Enlargement of the spleen occurs in any blood-borne infection. The nonspecific splenic reaction in these infections is caused both by the microbiologic agents themselves and by cytokines that are released as part of the immune response.

Table 13-12 Disorders Associated with Splenomegaly

I. Infections
Nonspecific splenitis of various blood-borne infections (particularly infectious endocarditis)
Infectious mononucleosis
Tuberculosis
Typhoid fever
Brucellosis
Cytomegalovirus
Syphilis
Malaria
Histoplasmosis
Toxoplasmosis
Kala-azar
Trypanosomiasis
Schistosomiasis
Leishmaniasis
Echinococcosis
II. Congestive States Related to Portal Hypertension
Cirrhosis of the liver
Portal or splenic vein thrombosis
Cardiac failure
III. Lymphohematogenous Disorders
Hodgkin lymphoma
Non-Hodgkin lymphomas and lymphocytic leukemias
Multiple myeloma
Myeloproliferative disorders
Hemolytic anemias
IV. Immunologic-Inflammatory Conditions
Rheumatoid arthritis
Systemic lupus erythematosus
V. Storage Diseases
Gaucher disease
Niemann-Pick disease
Mucopolysaccharidoses
VI. Miscellaneous Disorders
Amyloidosis
Primary neoplasms and cysts
Secondary neoplasms

MORPHOLOGY

The spleen is enlarged (200 to 400 gm) and soft. Microscopically, the major feature is acute congestion of the red pulp, which may encroach on and virtually efface the lymphoid follicles. Neutrophils, plasma cells, and occasionally eosinophils are usually present throughout the white and red pulp. At times the white pulp follicles may undergo necrosis, particularly when the causative agent is a hemolytic streptococcus. Rarely, abscess formation occurs.

Congestive Splenomegaly

Chronic venous outflow obstruction causes a form of splenic enlargement referred to as *congestive splenomegaly*. Venous obstruction may be caused by intrahepatic disorders that retard portal venous drainage, or arise from extrahepatic disorders that directly impinge upon the portal or splenic veins. All of these disorders ultimately lead to portal or splenic vein hypertension. *Systemic, or central, venous congestion* is encountered in cardiac decompensation involving the right side of the heart, as can occur in tricuspid or pulmonic valvular disease, chronic cor pulmonale, or following left-sided heart failure. Systemic congestion is associated with only moderately enlarged spleens that rarely exceed 500 gm in weight.

Cirrhosis of the liver is the main cause of massive congestive splenomegaly. The "pipe-stem" hepatic fibrosis of schistosomiasis causes particularly severe congestive splenomegaly, while the diffuse fibrous scarring of alcoholic cirrhosis and pigment cirrhosis also evokes profound enlargements. Other forms of cirrhosis are less commonly implicated.

Congestive splenomegaly is also caused by obstruction of the extrahepatic portal vein or splenic vein. This can stem from *spontaneous portal vein thrombosis*, which is usually associated with some intrahepatic obstructive disease, or inflammation of the portal vein *(pylephlebitis)*, such as follows intraperitoneal infections. Thrombosis of the splenic vein can be caused by infiltrating tumors arising in neighboring organs, such as carcinomas of the stomach or pancreas.

MORPHOLOGY

Long-standing splenic congestion produces marked enlargement (1000 to 5000 gm). The organ is firm, and the capsule is usually thickened and fibrous. Microscopically, the red pulp is congested early in the course but becomes increasingly fibrotic and cellular with time. The elevated portal venous pressure stimulates the deposition of collagen in the basement membrane of the sinusoids, which appear dilated because of the rigidity of their walls. The resultant slowing of blood flow from the cords to the sinusoids prolongs the exposure of the blood cells to macrophages, resulting in excessive destruction (hypersplenism).