Splenomegaly

A Combined Clinical and Radiologic Approach to the Differential Diagnosis

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KEYWORDS

• Spleen • Splenomegaly • CT

KEY POINTS

- Splenomegaly is commonly encountered at clinical imaging, whether due to a known condition, an incidental finding, or an undiagnosed condition leading to symptomatic evaluation.
- The differential diagnosis for splenomegaly is broad but can be grouped into discrete categories based on pathophysiology.
- The differential diagnosis for splenomegaly can be narrowed by incorporating all relevant clinical and radiologic data.

INTRODUCTION

An enlarged spleen is a common imaging finding. Splenomegaly may be an incidental imaging finding, an expected finding based on a known clinical entity, or related to the underlying cause of a patient's symptoms or clinical presentation.[1] The broad physiologic functions of the spleen make it susceptible to involvement by a variety of pathophysiologic processes. Understanding the pathophysiology of splenomegaly allows for the formation of a limited differential diagnosis. By incorporating all relevant clinical and radiologic data, the differential diagnosis may be narrowed even further. This article provides an overview of splenomegaly, including a general categorization method of splenomegaly based on the pathophysiology and associated imaging findings.

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ANATOMY AND FUNCTION

A mesodermal derivative in the dorsal mesogastrium, the spleen migrates to the left upper quadrant during embryogenesis, where it connects to the stomach and the left kidney via the gastrosplenic and splenorenal ligaments, respectively. The celiac artery supplies the spleen via the splenic artery, which enters the spleen along with lymphatics and nerves at the splenic hilum. The splenic artery supplies end arteries that branch into small arterioles. The lack of collateral flow between the end arteries makes the spleen susceptible to infarction from branch occlusion. Small central end arterioles are surrounded by red pulp, consisting of pulp sinuses and pulp cords, and white pulp, which consists of B-cell follicles, marginal zones surrounding follicles, and T-cell lymphoid tissue surrounding the arterioles. A fraction of blood goes from the arterioles to capillaries, through the splenic veins, and out the spleen, but the larger percentage of blood enters the macrophage-lined sinuses and cords. The cords are blind-ending structures with very small openings through which red blood cells (RBCs) must traverse to re-enter the sinusoids and systemic circulation. RBCs that are damaged cannot squeeze through these slits[2] and are then destroyed and recycled. Venous drainage of the spleen is via the splenic vein, which drains into the portal vein. If the splenic vein is obstructed, collateral perigastric veins or a splenic vein to renal vein shunt can form.

The unique structure of the spleen allows for its participation in the body's adaptation to hostile circumstances. As a reticuloendothelial organ with hematopoietic capabilities, the normal physiologic roles of the spleen include erythrocyte quality control via removal of abnormal RBCs, antibody synthesis, and removal of bacteria and blood cells coated by antibodies. Additionally, the spleen is able to undergo hematopoiesis as a normal physiologic response during gestation.[2] These normal physiologic roles allow the spleen to assist in clearing bacteria and particulates from the blood, generating immune responses, and generating blood cells if the marrow is unable to meet the body's needs.

Interpreting the normal spleen size depends on the interpreter's desired position on the receiver operating characteristic curve. There is a normative distribution and wide variety of normal splenic volumes. Although volumetric assessment of the spleen is most accurate, this has generally not yet been practical. In attempting to determine a single maximum splenic length to accurately diagnose splenomegaly, Bezerra and colleagues[3] suggest that a 9.8-cm craniocaudal dimension corresponds with a volume of greater than 314.5 cm^3, which has previously been suggested as the upper volume limit of the normal spleen.[4] Other studies have suggested that a maximal diameter of 13 cm or greater on imaging is abnormal.[5,6] Given the variation in gender-specific normal spleen sizes and volumes (**Fig. 1**), the authors believe that a maximal craniocaudal dimension of 13 cm is a reasonable marker of splenomegaly, with automated volumetric assessment most optimal in the (near) future.

PATHOPHYSIOLOGY

Pathologic splenomegaly can best be considered as the result of any combination of 3 general pathophysiologic mechanisms: (1) hyperplasia or hypertrophy of normal splenic components, (2) passive congestion, and (3) infiltrative diseases. Alternative categorization systems based on the type of pathologic condition affecting the spleen have also been proposed,[1] with a general classification system, including (1) hematologic, (2) infectious, (3) congestive, (4) inflammatory, (5) neoplastic, and (6) infiltrative