

Congenital abnormalities of the spleen are rarely encountered. However, when present, they may be difficult to interpret due to unfamiliarity. Occasionally, they can mimic serious pathology and should be familiar to pathologists to exclude malignancies. Lesions illustrated in this chapter include accessory spleen, which when present in unusual locations may be a diagnostic problem; intrapancreatic spleen, which is a rare occurrence and probably a variant of

accessory spleen; splenosis, which is marked by the presence of multiple, small, fragments of functional splenic tissue within the abdominal cavity; and splenogonadal fusion, a rare lesion that may involve unusual clinical presentations, including left-sided inguinal hernia. Finally, surface grooves present in the spleen are illustrated and their embryologic origin is identified. Both histologic and gross images are presented.

2.1 Accessory Spleen

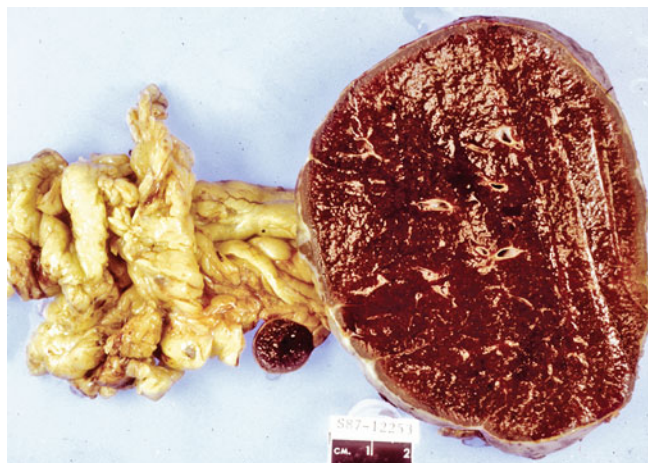


Fig. 2.1 Accessory spleen. A section of spleen with attached pancreas and omentum. In the mass of tissue arising from the hilum, there is a small nodule (approximately 2 cm) of dark red purple tissue. This is an accessory spleen. They are seen in approximately 10 % of the population and are most commonly seen in the splenic hilar region. (Image courtesy of D. Farhi, Atlanta, GA, USA.)



Fig. 2.3 Gross image of a sectioned accessory spleen. In this case, the accessory spleen was approximately 5 cm in diameter. Note that the cut surface has a deep red color, similar to the spleen. (Image courtesy of L. Morgenstern, Los Angeles, CA, USA.)

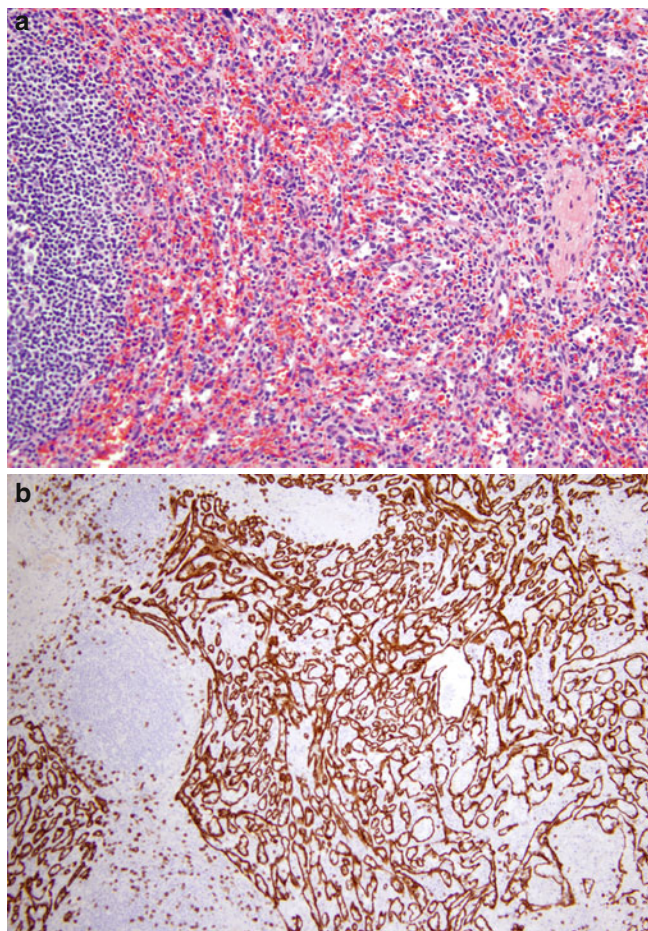


Fig. 2.2 Accessory spleen. This pathologic specimen was submitted as an “enlarged lymph node” with a suspicion of metastatic carcinoma. It was seen adjacent to the pancreas and, as illustrated by histology (a) and immunohistochemical stains for CD8 (b), is an accessory spleen. CD8 staining highlights the distinctive splenic sinus architecture of the spleen and can be useful in identifying spleen tissue at unusual sites

2.2 Intrapancreatic Spleen

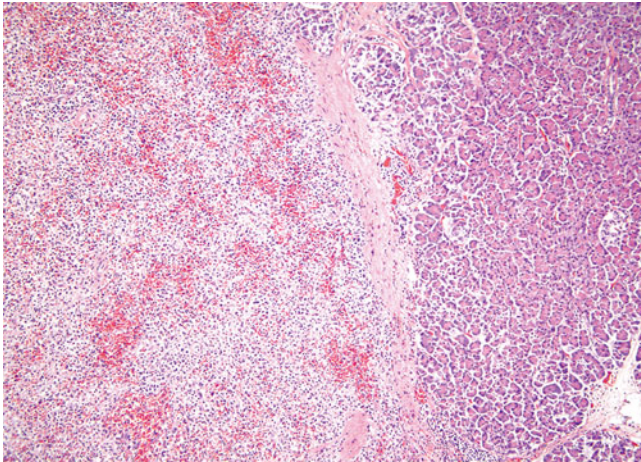


Fig. 2.4 Intrapancreatic spleen is a rare finding associated with embryologic abnormality. Small fragments of spleen tissue are entrapped within a portion, usually distal, of the pancreas (normal pancreatic tissue; *right*). Although these can vary in size, they are usually quite small. In this case, the elements are predominant red pulp components, suggesting that this was a relatively early embryologic event. (Image courtesy of R. Mills, Salt Lake City, UT, USA.)

2.3 Polysplenia



Fig. 2.5 Polysplenia. Gross image of polysplenia. In this case, the primordial separate lobes of splenic tissue do not fuse to make a single unified organ. Rather, they are partly fused or remain separated by fibrous bands. Although this anatomic abnormality can be seen associated with normal function, it is often associated with other, severe congenital abnormalities. (Image courtesy of D. Farhi, Atlanta, GA, USA.)



Fig. 2.6 Polysplenia. Another example of a small gross specimen from a fetus with polysplenia. Separated nodules of spleen tissue are present. Each nodule would have its own artery and vein, which eventually merge into the common splenic artery or vein. (Image courtesy of D. Farhi, Atlanta, GA, USA.)

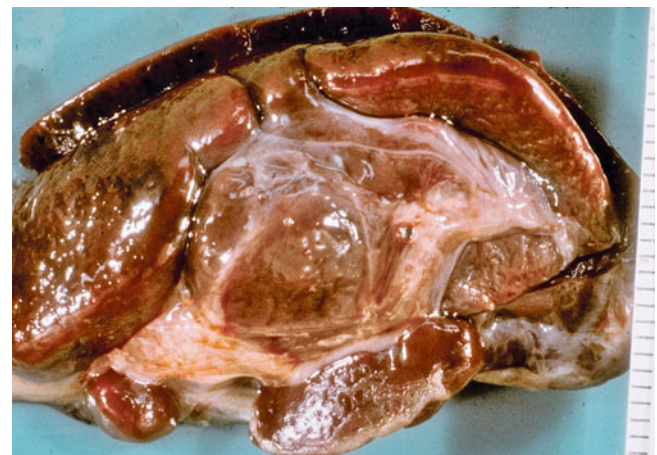


Fig. 2.7 Polysplenia. Still another example of polysplenia. Note that the lobes of the spleen have not completely fused. (Image courtesy of D. Farhi, Atlanta, GA, USA.)

2.4 Splenosis



Fig. 2.8 Splenosis. Gross image of splenosis. In this case, there are nodular, dark red tissue fragments attached to segments of bowel. This most often occurs after trauma, when the spleen is ruptured. Small fragments of splenic tissue are seeded throughout the abdominal cavity. Each establishes its own blood supply and functions as a tiny but complete spleen. In cases of splenectomy and pathologic processes, such as hemolytic anemias, these small displaced splenic fragments may enlarge greatly. (Image courtesy of D. Farhi, Atlanta, GA, USA.)

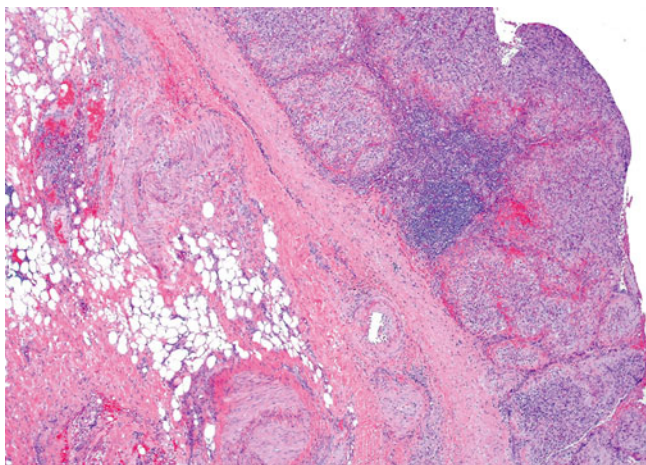


Fig. 2.9 Splenosis. Microscopic image of splenosis. The tissue present is composed of fibroadipose tissue from the peritoneum. At the intraperitoneal surface (right and upper) are slightly fibrous lymphoid areas. These represent fragments of spleen tissue that were likely "transplanted" to this site following abdominal trauma

2.5 Splenogonadal Fusion



Fig. 2.10 Spleno-ovarian fusion. Gross image of spleno-ovarian (eg, splenogonadal) fusion. In this case, there is a physical attachment between the stretched and elongated splenic fragment, which has tethered a portion of the left ovary. In some cases, the fusion may only consist of a thin fibrous band, which may have small entrapped fragments of splenic parenchyma



Fig. 2.11 Spleno-gonadal fusion. Ectopic splenic tissue in a sectioned testicle (lower). The splenic tissue has the usual deep red color, with normal, somewhat compressed testicular tissue in the upper portion of the sample

2.6 Other Findings



Fig. 2.12 Surface grooves. This spleen is enlarged by a low-grade B-cell lymphoma. However, deep grooves are seen in the medial surface (*upper left*), the upper pole (*far right*), and in the lateral/dorsal surface (*lower, middle*). These grooves are a normal variant, and are residual separations from the original embryologic splenic lobes that are incompletely fused in the adult. They have no specific physiologic consequences. (Image courtesy of W. Greaves, Houston, TX, USA.)

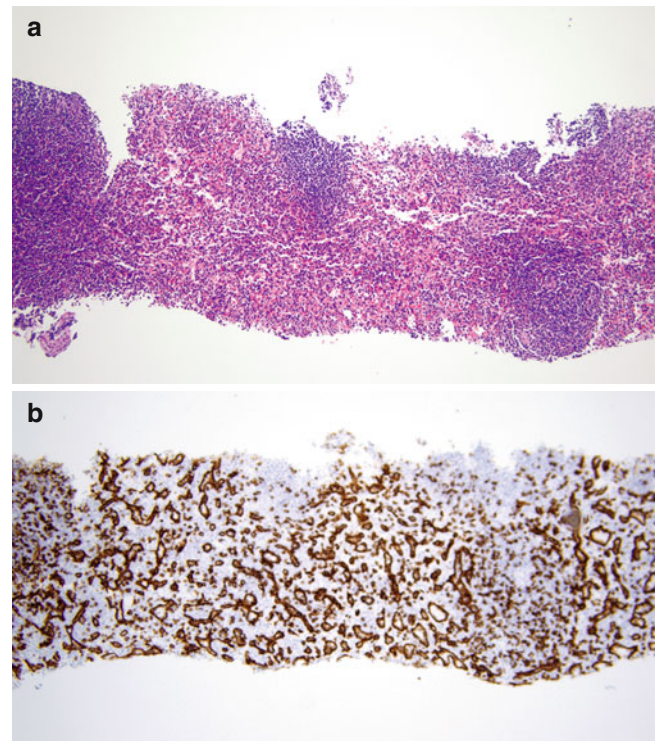


Fig. 2.14 Needle biopsy, lung. A biopsy of the left lower lobe of lung revealed this tissue. In this case, the lung was missed and a sample of normal spleen was taken (**a**). Normal splenic architecture is highlighted by CD8 staining (**b**), which is a useful stain for highlighting splenic red pulp architecture

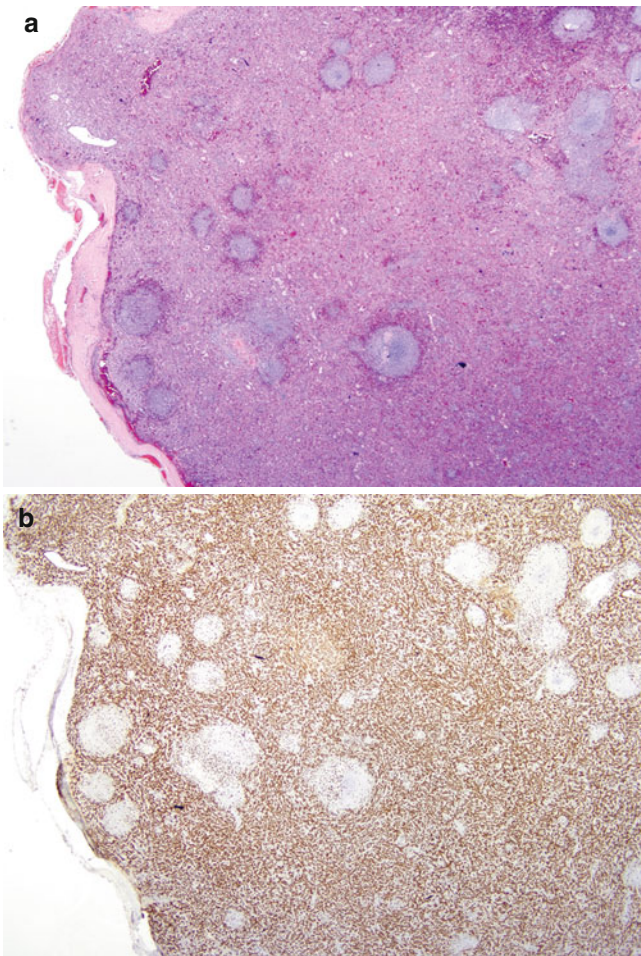


Fig. 2.13 Tissue taken from “left inguinal hernia.” In this case, the histology (**a**) and presence of a red pulp architecture staining by CD8 (**b**) show that the “inguinal hernia” was in fact caused by ectopic spleen tissue. In the descent of the testes from the mesonephros, small portions of spleen tissue may be transported. When these fragments are attached, it is splenogonadal fusion; when unattached, these fragments may be present as inguinal lumps, within the inguinal canal, simulating a hernia

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O'Malley, D.

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