

# Answer

## Sclerosing Angiomatoid Nodular Transformation of the Spleen

The removed spleen measured  $9 \times 7 \times 5$  cm and weighed 133 g. Grossly, the lesion was a single, well-circumscribed, round-to-bosseled mass that measured  $4 \times 3.5$  cm (Figure 2). At its periphery, multiple dark brown nodules were interspersed with stellate whitish fibrotic stroma. On microscopic examination, multiple angiomatoid nodules were seen submerged in a fibrotic stroma, with sharp demarcation of the lesion from the adjacent splenic parenchyma (Figure 2). The angiomatoid nodules were composed of complex disorganized vasculature with 3 distinct types of vessels that showed immunostain reactivity to CD34, CD8, and CD31. The lesion was consistent with sclerosing angiomatoid nodular transformation (SANT).

Sclerosing angiomatoid nodular transformation of the spleen is an entity recently described by Martel et al<sup>1</sup> and earlier under the term *capillary cord hemangiomas* by Krishnan et al.<sup>2</sup> It is usually asymptomatic, seen in women in middle age, and diagnosed on imaging studies performed on patients for other reasons, usually in a background of malignancy. Patients may present to the surgeon with abdominal pain, raised erythrocyte sedimentation rate, fever, and anemia, requiring an extensive evaluation.

Hemangioma is the most common vascular tumor of the spleen and is distinguished from SANT by nuclear imaging. Histologically, it is composed of a single type of blood vessel similar to littoral cell angioma and lacks the distinctive angiomatoid nodular features seen in SANT.<sup>3</sup> Epithelioid hemangioendothelioma is a rare splenic tumor of borderline malignant potential. It may be differentiated from SANT by the presence of cytological atypia and a low mitotic index.<sup>4</sup>

Splenic hamartomas contain structurally disorganized red pulp tissue and are differentiated from SANT by the absence of angiomatoid nodular pattern. Metastases to the spleen may also present as multiple nodules owing to exaggerated stromal response and are easily differentiated on histology. Kaposi sarcoma is usually seen in immunocompromised patients with human immunodeficiency virus or after transplant.

Splenectomy is uniformly curative in all patients with SANT. No recurrence has been reported to date.

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