A 36-year-old man presented with a 4-week history of intermittent right upper quadrant pain, radiating to the back. No abnormalities were found on physical examination and blood tests. Abdominal ultrasonography showed gallstones and multiple hyperechoic masses in an enlarged spleen. Computed tomography (Fig. 1) demonstrated multiple hypodense lesions up to 3 cm in diameter, replacing 40% of otherwise normal splenic parenchyma. No other abnormalities were noted. The referring hematologist and the radiologist raised concerns of this being a primary splenic lymphoma. Laparoscopic cholecystectomy and splenectomy were scheduled.

At laparoscopy, dense right upper quadrant adhesions, abundant intra-abdominal fat and inadequate visualization of the anatomy of the area prompted conversion to an open procedure. The gallbladder and spleen were removed without complication. Pathological examination of the gallbladder showed cholelithiasis and chronic cholecystitis. Grossly, the spleen weighed 500 g and measured $22 \times 11 \times 5$ cm. The red pulp contained tan, spongy nodules, ranging in size from 0.5 to 3 cm (Fig. 2). The intervening splenic parenchyma was unremarkable.

Microscopically, the lesions were composed of a mesh of ectatic anastomosing vascular channels lined by splenic sinus cells (Fig. 3). The cells were positive for both endothelial (Factor VIII, CD31 and CD34) and histiocytic (CD68) markers on im-
munohistochemical staining. Cyto-
logic atypia and mitotic figures were
absent. The diagnosis was littoral cell
angioma.

Primary splenic tumours are most
often benign proliferations of vascular
endothelial tissue. Littoral cell an-
gioma is an unusual primary splenic
vascular neoplasm first described in
1991. It originates from lining cells
of the red-pulp sinuses (littoral cells),
exhibiting endothelial and histiocytic
features. Although usually found in-
cidentally, littoral cell angioma may
present with splenomegaly, thrombo-
cytopenia or anemia. Diagnosis is
difficult because of the nonspecific
nature of clinical, laboratory and radi-
ologic features; splenectomy is neces-
sary to exclude lymphoma and other
malignant neoplasms. There are no
reports of local recurrence or distant
metastases with typical benign littoral
cell angioma, but a malignant variant
(littoral cell angiosarcoma) has been
described. An association with vis-
ceral organ cancers (colorectal, renal,
pancreatic) has also been noted.

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FIG. 3. Hematoxylin–eosin stain, original magnification × 200.