

Littoral cell angioma mimicking hepatic tumor

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Abstract

Littoral cell angioma is a rare vascular tumor of the spleen that was described by Falk *et al.* in 1991. Because of the limited number, untypical imaging manifestations, and lack of knowledge on this tumor type, these tumors are often misdiagnosed. In most cases, the tumor presents with multiple small hypoattenuating nodules in the spleen with delayed enhancement. However, solitary littoral cell angiomas have not been well described. We present the CT features of an unusual littoral cell angioma mimicking hepatic tumor.

Keywords: Vascular tumor, spleen, diagnostic imaging, pathology, liver

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Littoral cell angioma is a rare vascular tumor of the spleen that was described by Falk *et al.* in 1991 (1). Because of the limited number (approximately 80 patients accessioned from November 1991 to February 2012 in English language journals), untypical imaging manifestations, and lack of knowledge, these tumors are often misdiagnosed. In most cases, the tumor presents with multiple small hypoattenuating nodules in the spleen showing delayed enhancement. However, solitary littoral cell angioma has not been well described. We present the CT features of an unusual littoral cell angioma mimicking hepatic tumor.

Case report

A 53-year-old woman presented with a 10-year history of intermittent abdominal pain, swelling and continuous vomiting. The patient denied presence of fever, nausea, and weight loss. There were no significant findings at physical examination. An abdominal ultrasound exam revealed a 10.4 × 10.0 cm mass of heterogeneous echogenicity in the left upper abdomen. Axial unenhanced CT scan (Fig. 1a) confirmed an ill-defined heterogeneous mass 9.1 cm in diameter that filled the left abdominal quadrant, located between the left lobe of the liver and the spleen. The CT attenuation of the mass was around 26–53 HU on non-enhanced scan. After intravenous contrast medium injection the mass enhanced gradually and heterogeneously (Fig. 1b–e), but was still hypodense relative to the spleen. There was no accompanying lymphadenopathy or evidence of malignant process elsewhere in the abdomen. Because the origin of the mass was unknown and a primary malignancy

could not be excluded, the patient underwent an explorative laparotomy. During the operation, a well-encapsulated mass was found under the left hepatic lobe and the pedicle arising from the spleen. Resection of the mass and the spleen was performed.

Gross examination of the spleen showed splenomegaly (580 g, 15 × 10 × 9 cm) with an ill-defined brownish nodule (5 × 8 × 8 cm). Histologic examination (Fig. 2) revealed that the lesion was composed of variably sized vascular channels lined with flat and tall endothelial cells, with papillary fronds extending into the vascular channels. No atypical cells or mitosis were present. The endothelial lining cells of the vascular channels were positive for CD31 and CD68, factor VIII and negative for CD34, CD21, and CD8. These findings were considered diagnostic of benign splenic littoral cell angioma.

Discussion

The most common primary tumors of the spleen are benign and originate from the vascular endothelium. Littoral cell angioma is a rare vascular tumor first described in 1991 by Falk *et al.* (1). This neoplasm arises from littoral cells that originate from the splenic red pulp sinuses, which are unique among vascular lining cells in that they express histiocytic antigens and the usual endothelial antigens.

There is no gender or age predilection, although most cases of littoral cell angioma occur in adults (median age, 48–49 years) (1). Patients are usually asymptomatic. Some patients presents with abdominal pain, weakness, weight loss, splenomegaly (2), or laboratory evidence of

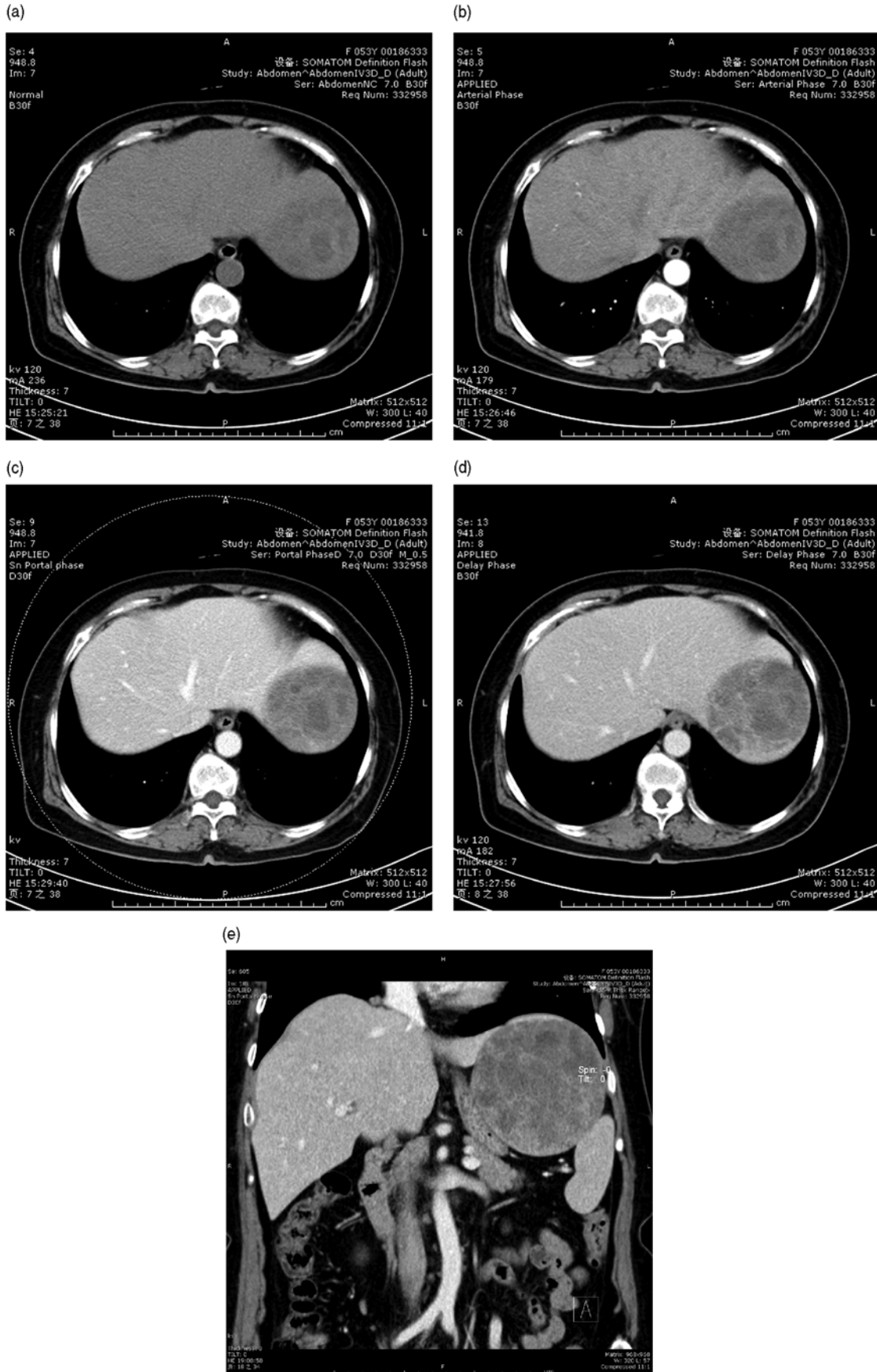


Fig. 1 (a) Axial unenhanced CT image demonstrates an ill-margined solitary mass that fills the left abdominal quadrant. (b, c) The mass enhanced gradually and heterogeneously during the arterial and portal phase. (d) At delay phase the mass was still hypodense relative to the spleen. (e) On the coronal image of portal phase, the mass was located between the left lobe of the liver and the spleen, and it appeared closely related to both organs

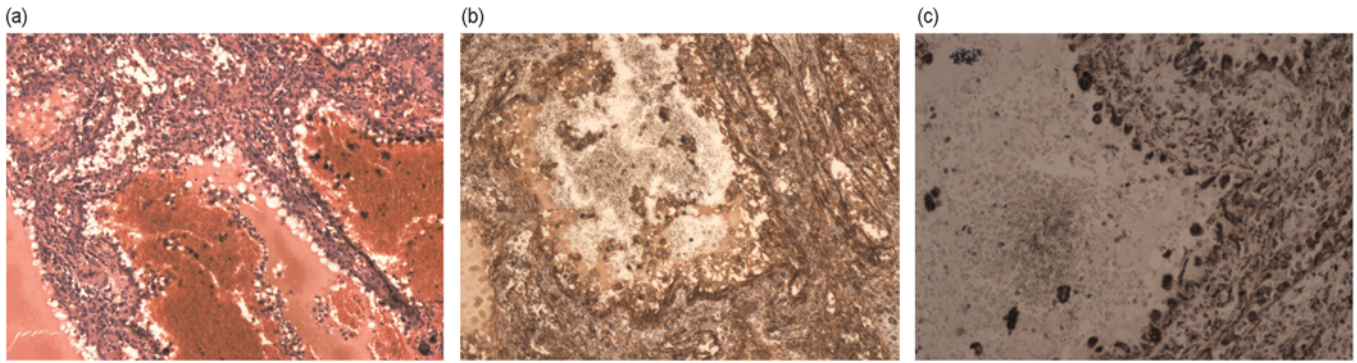


Fig. 2 (a) Variably sized vascular channels lined with flat and tall endothelial cells (HE, $\times 100$). The endothelial lining cells of these vessels were positive for CD31 (b) ($\times 100$) and CD68 (c) ($\times 200$)

hypersplenism (3). Massive splenomegaly because of littoral cell angioma can be mistaken as a hepatic tumor (as in our case). Despite several reported cases of malignant variants (4), most cases of littoral cell angiomas are benign. It may occur with concomitant visceral malignancies, such as non-small cell lung cancer, transitional cell carcinomas (5), colon cancer, rectal cancer, renal and pancreatic adenocarcinomas (6). Association with melanomas and lymphomas (7) has also been reported. Thus an evaluation and surveillance for concomitant malignancy is recommended.

There are two types of radiologic presentation of littoral cell angiomas. The most frequent type is seen as a tumor including multiple nodules involving the entire spleen (8). The ultrasonographic findings are non-specific, with descriptions of LCA varying from a cystic hypoechoic mass to a homogeneously hyperechoic mass in the literature. The lesions are invisible on unenhanced CT scans and appear as multiple, well-marginated, and hypoattenuating on early portal venous phase after contrast material administration. They enhanced homogeneously and become isodense to the normal splenic parenchyma in the late portal venous phase (8). Various splenic pathologies can present as multiple low-attenuating lesions including hemangiomas, lymphoma, metastases, lymphangiomas, sarcoidosis, and disseminated fungal disease (8).

The second type is rare and presents as a solitary lesion (9). Although there are cases that associate littoral cell angiomas with hepatocellular carcinomas (10, 11), to our knowledge, the LCA in our case is first one presenting as a solitary, large, and solid mass mimicking a hepatic tumor. The heterogeneous mass was located in between the hepatic left lobe and the spleen and showed gradual heterogeneous enhancement. Several differential diagnoses exist. In addition to littoral cell angioma, other splenic tumors should also be considered, such as hemangioma, hamartoma, angiosarcoma, hemangioendothelioma, extramedullary hematopoiesis, and inflammatory pseudotumor (9). Despite sophisticated imaging techniques, it is not often possible to differentiate these primary splenic vascular neoplasms from hepatic tumors.

As described by Falk *et al.* (1), the findings by gross pathological examination usually consist of multiple sponge-like, blood-filled, red-brown nodules. These lesions

are well-demarcated and compress the adjacent splenic parenchyma but do not exhibit a capsule. Histopathologically, there are anastomosing vascular channels of variable sizes replacing the splenic red pulp. These channels are lined with flat and tall endothelial cells and have irregular lumina, often displaying papillary projections and cyst-like spaces. The cells usually sloughed off into the vascular lumina and may show macrophage-like morphology and exhibit hemophagocytosis. Typically, few mitotic figures and no cytologic nuclear atypia are seen. Immunohistochemical staining is specific, as the tumor demonstrates immunoreactivity with endothelial markers (factor VIII, CD31) and histiocytic markers (CD68, lysozyme, CD21), which demonstrate the tumor's dual histiocytic/endothelial differentiation. They are normally negative for CD8, CD34, and S-100 antigens.

In conclusion, littoral cell angiomas are usually benign and do not mandate immediate resection unless the lesion is symptomatic. However, the benign form needs to be differentiated from the malignant variant, the littoral cell angiosarcoma. Splenectomy is curative as well as useful in providing an exact diagnosis. Given its apparent association with malignancy and immunologically mediated diseases, careful clinical work-up before surgery and postoperative surveillance are recommended.

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