Littoral Cell Angioma of the Spleen: a case report

YU-PENG LIU¹ SHE-MENG CHENG¹ JON-KWAY HUANG¹²

Department of Radiology¹, Mackay Memorial Hospital
Department of Radiology², Taipei Medical University

Littoral cell angioma (LCA) is a rare primary vascular tumor of the spleen that has been reported infrequently and variably in the radiology literature. We present a patient with LCA of the spleen who had only nonspecific low back pain and epigastralgia for several months. The imaging findings in unenhanced and contrast-enhanced CT, ultrasonography, as well as histologic findings are presented. The imaging findings and differential diagnoses of splenic tumors are discussed.

Key words: Spleen; Neoplasms

Littoral cell angioma (LCA) is a very rare vascular tumor of the spleen arising from splenic sinus lining cells (littoral cells). We present a 44-year-old female with pathologically proven LCA, studied by computed tomography (CT) and ultrasonography (US). In cases of splenomegaly with multiple splenic lesions, one should consider this possibility of LCA in the differential diagnoses.

CASE REPORT

A 44-year-old female presented with a 2-year history of asymptomatic splenomegaly. She complained of low back pain and epigastralgia in recent several months, but denied weight loss, fever, night sweats, or changes in bowel habits. Physical examination revealed splenomegaly, the laboratory data was unremarkable: hemoglobin (13.2 mg/dl); WBC count (4.10 × 10³ µl); hematocrit (39.7%); platelet count (141 × 10³ µl); and mean cell volume (90.9 fl).

Abdominal US showed splenomegaly, heterogeneous splenic echotexture with multiple hypoechoic lesions (Fig. 1). The abdominal CT images revealed an enlarged spleen measuring 12.5 × 11.0 × 4.5 cm in size. In the precontrast study, multiple low-attenuating nodular lesions were present throughout the spleen. The post-enhanced CT images at late portal venous phase revealed multiple well-defined, heterogeneously hypoattenuating lesions with partial areas of enhancement that measured several millimeters to 1.7 cm in diameter (Fig. 2, 3). Under the impression of lymphoma or metastases, splenectomy was performed. Postoperative pathologic examinations revealed anastomosing vascular channels lined with tall endothelial cells, focal papillary fronds, and normal splenic sinuses at the periphery of the lesion. The final diagnosis was LCA of the spleen.

DISCUSSION

LCA is a rare benign vascular tumor of the spleen that was first described by Falk et al in 1991 [1].
Littoral cells originate from the red cell pulp sinuses and have features intermediate between those of endothelial cells and macrophages. LCA may occur at any age in patients of either sex. Typically, patients with splenic LCA present with benign findings of hypersplenism such as splenomegaly, thrombocytopenia or anemia. In most patients, because of hematologic manifestation and nonspecific imaging findings, splenectomy is performed for definitive diagnosis and treatment.  

To our knowledge, there have been 16 cases with imaging findings of LCA reported in the English-language literature since the description of the disease in 1991 [2-10]. A review of imaging features of LCA with clinical correlation in these cases, including ours, is listed as Table 1.  

Splenomegaly is found in all except three patients at physical examination. All patients have laboratory evidence of hypersplenism except for one of the reported cases and our case.

The sonographic appearance of splenic LCA is variable, including mottled echotexture without

Figure 1. Abdominal ultrasonography showed splenomegaly with heterogeneous echotexture and multiple hypoechoic lesions (arrow heads).

Figure 2. Axial pre-contrast CT scan showed multiple hypodense nodules in the spleen.

Figure 3. Axial a. and coronal b. contrast-enhanced CT images at late portal venous phase showed multiple well-defined, heterogeneously hypoattenuating lesions with partial areas of high density corresponding with the vascular spaces inside the lesions.
### Table 1. Imaging Features of LCA in 17 cases with Clinical Correlation

<table>
<thead>
<tr>
<th>Patient No./ Age (y)/ Sex</th>
<th>Splenomegaly</th>
<th>No. of masses</th>
<th>Ultrasound findings</th>
<th>Postcontrast CT findings</th>
<th>MRI findings</th>
<th>Signs of hypersplenism (anemia and thrombocytopenia)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/39/F</td>
<td>Yes</td>
<td>Numerous</td>
<td>Homogeneously hypoattenuating</td>
<td>*</td>
<td>Yes</td>
<td></td>
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<tr>
<td>2/1.3/F</td>
<td>Yes</td>
<td>Numerous</td>
<td>Isoechoic</td>
<td>*</td>
<td>T1WI: hypointense T2WI: hypointense</td>
<td>Yes</td>
</tr>
<tr>
<td>3/54/M</td>
<td>Yes</td>
<td>Numerous</td>
<td>Homogeneously hypoattenuating and isoattenuating in late portal venous phase</td>
<td>*</td>
<td>Yes</td>
<td></td>
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<tr>
<td>4/59/M</td>
<td>Yes</td>
<td>Numerous</td>
<td>Mottled echotexture of spleen</td>
<td>Homogeneously hypoattenuating and isoattenuating in late portal venous phase</td>
<td>*</td>
<td>Yes</td>
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<tr>
<td>5/66/M</td>
<td>No</td>
<td>Numerous</td>
<td>Homogeneously hypoattenuating</td>
<td>T1WI: hypointense Post-enhanced T1WI: contrast medium pooling on delayed-phase T2WI: inhomogeneously hyperintense</td>
<td>No</td>
<td></td>
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<tr>
<td>6/62/F</td>
<td>No</td>
<td>Numerous</td>
<td>Hyperechoic</td>
<td>*</td>
<td>T1WI: isointense or slightly hypointense Post-enhanced T1WI: capture of contrast medium T2WI: hypointense</td>
<td>Yes</td>
</tr>
<tr>
<td>7/58/M</td>
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<td>Numerous</td>
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<td>*</td>
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<tr>
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<td>9/55/F</td>
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<td>Numerous</td>
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<tr>
<td>10/61/M</td>
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<tr>
<td>11/32/M</td>
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<td>12/41/M</td>
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<td>13/36/F</td>
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<td>14/84/F</td>
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<tr>
<td>15/74/F</td>
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<td>16/39/F</td>
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<td>17/44/F</td>
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<td>Hypoechoic</td>
<td>Heterogeneously hypoattenuating in late portal venous phase</td>
<td>*</td>
<td>No</td>
</tr>
</tbody>
</table>

* The image studies were not performed.

Patient No. 17: our case
discrete lesions [5], isoechoic [3], hypoechoic [8], and hyperechoic nodules [7, 9]. There are only 6 cases reports with sonographic study in the medical literature and our case is hypoechoic.

On abdominal CT images without or with contrast enhancement, numerous hypodensiating lesions within the spleen are present in all except for one patient [10]. On delayed contrast-enhanced images, LCA may show homogeneously enhancement and become isodense to the remaining splenic parenchyma in three cases (not present in our case), the findings suggest that contrast material filling in the vascular spaces inside LCA. It may help differentiate LCA from other lesions [4, 5, 10].

On MR images, LCA typically appear marked hypointensity on both T1- and T2-weighted pulse sequences, a finding that reflects the presence of hemosiderin in the lesions due to the hematophagocytic capacity of the neoplastic cells [3, 6, 7, 11]. However, one case shows inhomogeneously hyperintensity on unenhanced T2-weighted images as the same findings in hemangioma of the spleen [6].

The differential diagnoses for LCA of spleen include neoplasms (metastatic disease, lymphoma, hemangiomas, lymphangioma) and nonneoplastic disorders such as infectious diseases and granulomatous diseases [10, 11].

Metastatic disease and lymphoma may affect the spleen in a diffuse manner, leading to difficulty in distinguishing them according to the distribution pattern. Hypoechoic nodules are the characteristic sonographic findings of both metastases and lymphoma [12].

After IV contrast material administration, splenic metastases and lymphoma typically appear as hypodense lesions with variable enhancement on CT images. Typical findings such as nodal disease are not present in patients with LCA. Hemangiomas are the most common benign primary splenic tumor. On sonography, hemangiomas may be homogeneously hyperechoic but may also show mixed echogenicity and cystic components [13]. At CT, they are typically hypoattenuating masses with contrast enhancement pattern similar to that of hepatic hemangiomas or a pattern of mottled contrast enhancement on delayed images [14]. Lymphangiomas are benign neoplasm of the spleen composed of endothelium-lined cystic spaces containing proteinaceous fluid. These cystic spaces appear hypodense on unenhanced and contrast-enhanced CT [11]. On sonography, the cystic component of the lesion is confirmed by the presence of multiple anechoic nodules of various sizes with posterior acoustic enhancement [11].

Infectious processes with microabscess may mimic LCA. Fungus, Pneumocystis carinii, and infections from Mycobacterium species may cause multiple microabscesses of the spleen that are hypodense on contrast-enhanced CT and homogeneously hypoechoic on sonography [15]. In these cases, the patient’s clinical condition and immune status will help distinguish an infection from LCA.

Sarcoidosis may involve the spleen in a diffuse manner. On CT, irregularly distributed hypodense lesions are seen in an enlarged spleen. Associated adenopathic, pulmonary, and mediastinal diseases may help establish the diagnosis of sarcoidosis. The sonographic features of splenic sarcoidosis include increased echogenicity of the splenic parenchyma with focal hypoechoic or mixed echogenic lesions [16].

In conclusion, LCA is a benign neoplasm of the spleen composed of multiple vascular spaces. The most common imaging findings are splenomegaly with innumerable masses. It must be considered in the differential diagnoses of multiple splenic lesions with heterogeneous hypoattenuation on the late portal venous phase of contrast-enhanced CT and multiple hypoechoic nodules found on sonography, particularly if the patient has clinical signs of hypersplenism.

**REFERENCES**

10. Levy AD, Abbott RM, Abbondanzo SL. Littoral cell angioma of the spleen: CT features with clinicopatho-
logic comparison. Radiology 2004; 230: 485-490
脾臟的Littoral cell血管瘤：病例報告

劉育朋¹ 鄭旭萌¹ 黃榮貴¹,²
馬偕紀念醫院 放射線科¹
台北醫學大學 放射線學科²

Littoral cell血管瘤是相當罕見的脾臟血管腫瘤。在此我們報導一個四十四歲的女性病例，臨床表現為無症狀的脾臟腫大。腹部超音波及電腦斷層掃瞄發現脾臟中有多個大小不一的病灶，由於無法排除腫瘤的可能性，患者接受脾臟切除手術並證實此病灶為Littoral cell血管瘤。

關鍵詞：脾臟，腫瘤