Littoral-cell Angioma of the Spleen

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Abstract
Littoral-cell angioma (LCA) is a rare vascular tumor of the spleen. It was thought to be a benign, incidental lesion. However, many recent reports have described it to be a malignant lesion with congenital and immunologic associations. We report a case of LCA of the spleen, which has been infrequently communicated in the literature. A 41-year-old female patient admitted to our hospital with a three-week history of weakness, weight loss, anorexia, and intermittent upper abdominal pain which improved slightly with antacid medication. Imaging studies, including computed tomography (CT) and magnetic resonance imaging (MRI), showed multiple lesions in the spleen. Laparoscopic splenectomy was performed.

Keywords: Littoral-cell angioma, red pulp, spleen


Introduction
Littoral-cell angioma (LCA) is a rare primary tumor of the spleen that was first described by Falk et al. in 1991. Two forms of LCA have been described; the more commonly encountered diffuse multiple nodular form and the rare solitary form. It originates from the specialized endothelial cells lining the sinus channels of the splenic red pulp, called “littoral cells”. Clinically, a majority of the LCA described in the literature have been found to be benign in nature, but several reports have described malignancy in the LCA. Definite evaluation can only be done at pathology. We report a case of LCA in a patient who had symptoms of weakness and intermittent upper abdominal pain which improved slightly with antacid medication. The combination of CT and MRI showed multiple lesions in the spleen.

Case Report
A 41-year-old female patient was admitted to our hospital with a three-week history of weakness, weight loss, anorexia, and intermittent upper abdominal pain which improved slightly with antacid medication. Physical examination was normal. A normal splenomegaly was found. Results of routine laboratory tests were normal. An ulcerous lesion in the duodenum was detected gastroscopicaly. An enhanced CT scan of the abdomen showed multiple round, hypodense lesions in the spleen (Figure 1). Abdominal MRI revealed multiple splenic hypointense lesions (Figure 2). Our presumptive preoperative diagnosis was lymphoma or hemangioma. After appropriate preoperative vaccination for Streptococcus pneumoniae, Haemophilus influenzae B, and Neisseria meningitidis, laparoscopic splenectomy was performed. Gross examination of the spleen showed a 305 grams yellowish-brown nodular organ measuring 17 cm × 8 cm × 6 cm. Two dark-brown, well-circumscribed nodules were identified; one measuring 6.5 × 0.4 cm near to the pancreatic tail and other measuring 2 × 1 × 0.3 cm just underneath the capsule. Histopathologic examination showed multiple, anastomosing vascular lesions that vaguely resembled splenic sinusoids lined by tall endothelial cells. Immunohistochemical staining (Figure 3) was positive for both endothelial (CD 31, factor VIII) and histiocytic (CD 68) markers (Figure 4). No cytologic atypia and mitotic figures were found. The postoperative course was uneventful and the patient was discharged on day five. Six months after the surgery, the patient was asymptomatic.

Discussion
Littoral-cell angioma (LCA) is a rare primary tumor of the spleen that was first described by Falk et al. in 1991. Two forms of LCA have been described; the more commonly encountered diffuse multiple nodular form as in our case and the rare solitary form. It originates from the specialized endothelial cells lining the sinus channels of the splenic red pulp, called “littoral cells”. LCA of the spleen may occur at any age (one to 77 years; median age, 50 years), with no sex-based predilection (female: male ratio, 5:3). Some of the patients have splenomegaly, abdominal pain, pyrexia of unknown origin, or hypersplenism. The majority of LCA are asymptomatic. Clinically, a majority of the LCA described in the literature have been found to be benign in nature, but several reports have described malignancy in the LCA. Definite evaluation can only be done at pathology. On ultrasound, the findings vary widely from heterogeneous echotexture without specific nodules to hyperechogenic, hypoechochogenic, or isoechogenic appearing lesions. CT scan typically shows multiple hypoattenuating nodules. MRI of the spleen shows hypointense lesions on T1 and T2 weighted scan due to the hemosiderin content of the tumor.
Morphologic diagnosis is based on the presence of anastomosing vascular channels lined with tall endothelial cells, focal papillary fronds, and normal splenic sinuses at the periphery of the lesion. It shares some morphologic and immunohistochemical features with hemangiomas of other sites such as immunoreactivity for vascular endothelial markers CD31 and factor VIII. All cases also express histiocytic marker CD68.

The pathogenesis of LCA remains unclear but, given its association with autoimmune disorders such as Crohn’s disease and Gaucher’s disease, immune system dysfunction has been postulated as a possible pathogenic mechanism.8,9

LCA is a benign vascular tumor of the spleen. Some of the patients who had LCA are associated with malignancies of visceral organs including adenocarcinoma of colorectum, malignant lymphoma, lung, pancreas, kidney, and hepatocellular carcinoma.10,11,12 It has also been associated with aplastic anemia13 and myelodysplasia.14

LCA may appear as single or multiple lesions in the spleen. An extensive list of possibilities such as multiple hemangiomas, lymphoma, metastatic disease, and disseminated infections caused by fungi, mycobacteria, Pneumocystis carinii, and sarcoidosis, should be considered in the differential diagnosis of multinodular splenomegaly.7

In conclusion, LCA are primary vascular neoplasms of the spleen and are usually an incidental finding. Clinically, a majority of the LCA described in the literature have been found to be benign in nature, but several reports have described malignancy in the LCA.6 We recommend a close clinical follow-up of patients with LCA of the spleen.
References