Littoral Cell Angioma of the Spleen Treated by Laparoscopic Splenectomy

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ABSTRACT

Massive splenomegaly due to multifocal littoral cell angio-
oma was discovered incidentally in a 55-year-old man
during a workup for an unrelated condition. The tumor
was removed successfully by laparoscopic splenectomy.
We report the second case of littoral cell angio-
ma of the spleen treated laparoscopically.

Key Words: Littoral cell angioma, Laparoscopic splenec-
tomy, Splenomegaly, Thrombocytopenia.

INTRODUCTION

Littoral cell angioma (LCA) of the spleen is a new subtype
of benign vascular tumor that involves the cells that line
the splenic sinus channels. This tumor expresses both
vascular and histiocytic antigens. Splenectomy is both
diagnostic and therapeutic. We present a case of LCA of
the spleen treated successfully with laparoscopic splenec-
tomy. Our review of the literature revealed only 1 other
report of splenic LCA treated laparoscopically.1

CASE HISTORY

A 55-year-old man with a history of right-sided flank pain
presented to the emergency department. The pain was
caused by a right renal stone that passed without treat-
ment. Incidentally on the computed tomographic (CT)
scan that was part of his workup, an abnormally large
spleen with numerous poorly defined hypodense areas
was noted (Figure 1). His past medical history was sig-
nificant for hypertension, type 2 diabetes mellitus, Crohn’s
disease, and obesity, but negative for any family history of
cancer. His previous surgical history included laparo-
scopic cholecystectomy. The patient had no known drug
allergies, and his current medications included calan SR,
enalapril, doxazosin, glyburide, metformin, and asacol.
He had a distant history of tobacco use and used alcohol
occasionally. He denied any constitutional symptoms,
such as fever, night sweats, and chills.

On physical examination, he was a well appearing, obese
man who was 71 inches tall and weighed 315 lbs. His
blood pressure was 176/70 and his heart rate was 92.
Abdominal examination revealed an obese abdomen
without tenderness, distension, masses, hernias, or orga-
nomegaly. The rest of his physical examination was un-
remarkable.

Laboratory results revealed normal white blood cell count
and platelet count. He was slightly anemic with a hemoglo-
bin of 13.7 g/mL (normal range, 14.0 to 18.0). The
differential count was also within normal limits. The basic
metabolic panel was within normal limits except for glu-
cose of 225 mg/dL (normal range, 70 to 130).

Magnetic resonance imaging confirmed multiple abnor-
mal areas of very low signal intensity on T1-weighted images, and mixed signal intensity was present on the T2-weighted images. Postcontrast films showed abnormal enhancement in all of the abnormal areas except 1 medial area, which resembled a simple cyst. The differential diagnosis at this point was lymphoma, metastatic disease, or an infectious process with microab-
scess formation.

During laparoscopic splenectomy, the spleen was noted to be very large with multiple irregularities on its surface. It was removed through one 5-mm viewing port and two 5-mm and one 12-mm working ports. The 12-mm port was enlarged to a 4-cm incision to remove the spleen. On gross pathology, the spleen weighed 788 grams and measured 19x10x6 cm. The patient had several spongy hemorrhagic circumscribed nodules that measured up to 6 cm in diameter. On microscopic pathology, the sectioned specimen revealed splenic parenchyma with nodular collections of varying sized vascular channels forming discrete but not circumscribed angiomas, some of the spaces of which showed organizing thromboses separated by compressed red pulp. The cells lining these varied from flat to polygonal to cuboidal and had benign uniform nuclei, sometimes containing hemosiderin pigment, sometimes were vacuolated, and rarely showed erythrophagocytosis. Histiocytic markers factor 8 immunoperoxidase stain and CD68 stained brightly to light up these cells. No evidence was present of a malignant component.

The patient did well postoperatively and was discharged to home on postoperative day 2 tolerating a regular diet. One year postoperatively, he is doing well.

DISCUSSION

Vascular tumors, the most common primary tumors of the spleen, have been divided into 3 types as differentiated by level of aggressiveness: benign (hemangiomas, lymphangiomas, and hamartomas), intermediate (hemangioendotheliomas), and malignant tumors (angiosarco-
mas).

LCA of the spleen was first described in 1991 as a new subtype of vascular tumor that arises from the normal littoral cells that line the sinus channels of the splenic red pulp and expresses both vascular and histiocytic anti-
gens. A malignant subtype, littoral cell angiosarcoma has also been described and possesses cytologically atypical
cells and malignant morphologic features, such as invasion of surrounding organs.

Clinically, patients with LCA present with splenomegaly, thrombocytopenia, anemia, or with constitutional symp-
toms, such a fever of unknown origin. Patients can also present completely asymptatically. The male-to-female
distribution is nearly equal.

CT findings consistent with LCA are multiple small ovoid areas of hypoattenuation on contrast-enhanced CT performed during the portal venous phase. These findings can also be seen in other primary splenic neoplasms that lead to diffuse involvement of the spleen. Consequently, metastatic disease, lymphoma, sarcoidosis, Kaposi’s sar-
coma, or infectious processes that lead to microabscess formation may mimic LCA.

LCA has characteristic morphologic and immunohisto-
chemical characteristics that differentiate it from other vascular splenic tumors. Gross pathology often reveals a large spleen with multiple nodular lesions. Microscopi-
cally, the lesions vary from narrow sinus-like channels to large spaces lined with cuboidal to tall pseudopapillated cells, sometimes containing hemosiderin and red cells. These spaces are surrounded by compressed red pulp, all different findings from the more common cavernous hem-
angioma. Immunohistochemical staining for factor 8 and CD68 are characteristic of LCA, showing both vascular and histiocytic phenotypes. Our patient’s specimen revealed gross, microscopic, and immunohistochemical changes diagnostic of splenic LCA.

The treatment for LCA is splenectomy, which is both diagnostic and therapeutic. Interestingly, one third of the previously reported cases were associated with either ma-
lignant lymphomas or visceral organ cancer, a correlation that was not seen in our patient.
Although LCAs of the spleen are considered a benign clinical entity, scattered reports have been made of tumors with more malignant behavior presenting with metastatic disease. These patients were found to have microscopic changes consistent with LCA with malignant areas and areas with cellular atypia. These cases suggest that due to the subtle differences between benign and malignant tumors, patients should be followed for signs of recurrence and metastasis.

Our search of the literature revealed only 1 other report in English of laparoscopic splenectomy for LCA. Studies have shown laparoscopic splenectomy to be superior to open splenectomy for certain diseases, such as idiopathic thrombocytopenic purpura, with its attendant benefits of decreased pain medication needed, earlier oral intake, and decreased length of stay. Kercher et al found that a laparoscopic approach was beneficial for massive splenomegaly (defined as craniocaudal dimension $\geq$17 cm and weight $\geq$600 grams), which is also supported by our experience with this patient. Brodsky et al also support laparoscopic splenectomy for an assortment of diseases, even in the setting of splenomegaly. Rosen et al found laparoscopic splenectomy to be safe for both benign and malignant hematologic conditions, including Idiopathic Thrombocytopenic Purpura and a case of LCA, with a conversion rate in their series of 5%. The ability of our patient to tolerate a regular diet and be discharged by postoperative day 2, as well as the avoidance of a large painful subcostal incision that would have hindered pulmonary toilet in this obese patient, underscore the benefit of laparoscopic splenectomy.

**CONCLUSIONS**

Primary tumors of the spleen are quite rare when lymphoid and hematologic tumors are excluded. LCA should be considered in the differential diagnosis of multinodular splenectomy, especially in the presence of anemia, thrombocytopenia, or splenic lesions in the absence of associated adenopathy. LCA with splenomegaly is amenable to laparoscopic splenectomy diagnostically and therapeutically.

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