Clinical Case Report

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# Laparoscopic splenectomy for littoral cell angioma of the spleen

## A case report

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**Abstract**

**Rationale:** Littoral cell angioma (LCA) is a rare primary vascular neoplasm of the spleen. It can be benign or malignant. Pathology and immunohistochemistry are the gold standards for the diagnosis of LCA. Therefore, splenectomy is recommended for the purpose of diagnosis and treatment, and subsequent follow-up is necessary. There are limited reports about LCA. Here, we present a case of a female patient with LCA undergoing laparoscopic splenectomy in order to provide clinical experience in LCA treatment.

**Patient concerns:** A 32-year-old female attended the outpatient Department of Hepatobiliary Surgery for follow-up of hepatic hemangiomas. The patient presented with intermittent abdominal distension, which was slightly under no obvious inducement.

**Diagnosis:** Physical examination found no signs of abdominal tenderness and rebound tenderness, and liver and spleen were impalpable. The contrast-enhanced computed tomography (CT) showed multiple space-occupying lesions in the spleen, mottled low-density lesions, multiple hypoattenuating nodules with no contrast enhancement on the arterious phase. Delayed contrast-enhanced helical CT scan displayed incomplete filling of hypodense splenic lesions.

**Interventions:** Given that it was uncertain whether it was a benign or a malignant tumor, a laparoscopic total splenectomy was performed.

**Outcomes:** The final pathological diagnosis was LCA. Her postsurgical course was uneventful, and no surgery-related complications were found. No signs of recurrence were observed in the 16 months after the operation.

**Lessons:** LCA was a rare primary vascular neoplasm of the spleen, and laparoscopic splenectomy for LCA was safe and feasible, and postoperative course was uneventful. However, regular follow-up and long-time monitoring after splenectomy for LCA is recommended because of its potential malignant biological behavior.

**Abbreviation:** LCA = littoral cell angioma.

**Keywords:** laparoscopic splenectomy, littoral cell angioma, splenic tumor

## 1. Introduction

Vascular neoplasms are the most common among the non-lymphoid tumors of the spleen with the exception of littoral cell angioma (LCA), which is extremely rare. Falk and his colleague reported 200 vascular neoplasms of the spleen after total splenectomy and identified 17 vascular tumors which were characterized by the cells lining the red pulp splenic sinuses.[1]

LCA can be discovered at any age and its distribution has no gender predilection. The actual incidence rate remains unclear because of its rarity. Up to now, limited cases have been reported. Usually, patients with LCA are asymptomatic, without abdominal pain, persistent fever, chills, weight loss, or other constitutional symptoms, and imaging findings are nonspecific.[2] Therefore, the final diagnosis depends on pathological examination.[3]

## 2. Case report

A 32-year-old Chinese female attended the outpatient department of hepatobiliary surgery for follow-up of hepatic hemangiomas, and computed tomography (CT) scan revealed incidentally multiple splenic foci without hypersplenism. The patient presented with intermittent abdominal distension, which

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*All information about the patient in this manuscript follows the patient's right of informed consent and is fully authorized by the patient. The ethics involved in the manuscript has been reviewed by the Ethics Committee of the Affiliated Hospital of Qingdao University (QDFY WZ 03416).*

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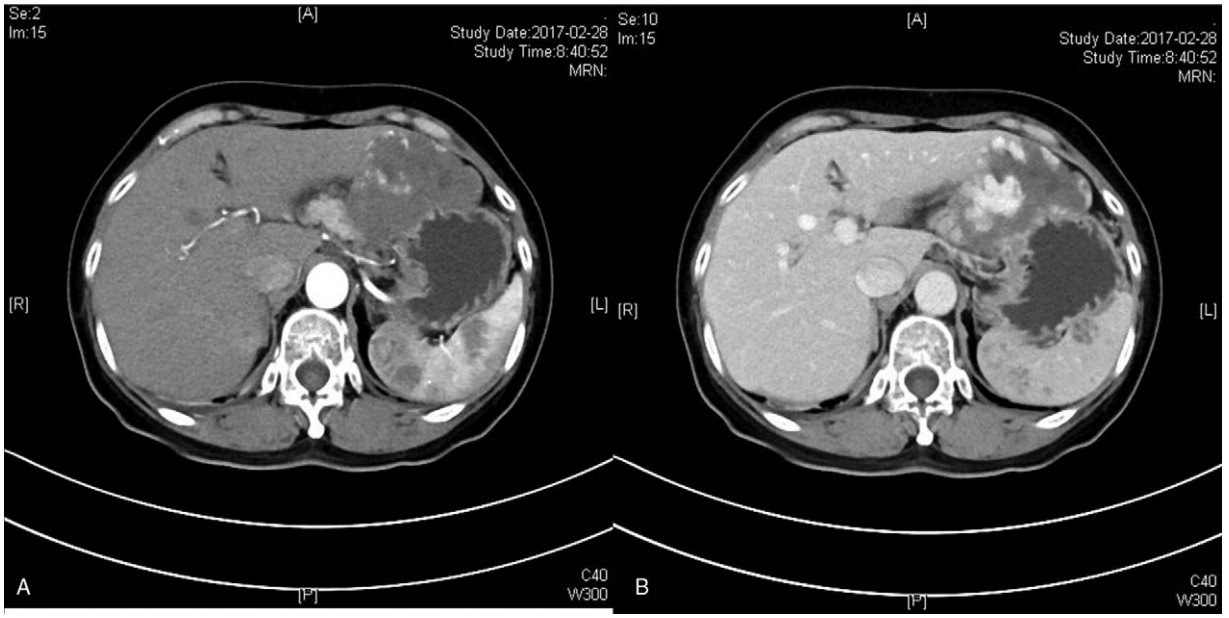


**Figure 1.** Arterious phase (A) shows multiple space-occupying low-density nodules with no contrast enhancement in the spleen. Delayed phase (B) shows incomplete filling of hypodense splenic lesions.

was slightly under no obvious inducement, with no fever, emesis, nausea, or vomiting. Physical examination found no signs of abdominal tenderness and rebound tenderness, and liver and spleen were impalpable.

Laboratory examination results were as follows: blood routine examination (hemoglobin 132 g/L, red blood cell count $4.61 \times 10^{12}$/L, white blood cell count $10.04 \times 10^{9}$/L, blood platelet count $180 \times 10^{12}$/L), liver enzymes (ALT 10.0 U/L, AST 17.0 U/L, GGT 16.0 U/L, ALP 86.0 U/L, TBIL 12.7 μmol/L) and renal function (BUN 5.65 mmol/L, SCr 42.0 μmol/L). Serum tumor markers AFP, CA19-9, CEA, and CA125 were all within normal limits.

Her past medical history included hemangioma for 20 years, and asthma for 30 years, and she underwent hysteromyomectomy 8 years ago. She did not smoke or drink. The splenic ultrasound revealed multiple hypoechogenic foci measuring 5 to 38 mm in diameter. The contrast-enhanced CT showed multiple space-occupying lesions in the spleen, mottled low-density lesions, multiple hypoattenuating nodules with no contrast enhancement on the arterious phase. Delayed contrast-enhanced helical CT scan displayed incomplete filling of hypodense splenic lesions. Imaging findings raised the suspicion of a vascular neoplasm of the spleen (Fig. 1A and B).

Given that it was uncertain whether it was a benign or a malignant tumor of the spleen, a laparoscopic total splenectomy was performed for the purpose of diagnosis and treatment and informed written consent was obtained from the patient for publication of this case report and accompanying images. Grossly, the spleen measured $11\,cm \times 6.5\,cm \times 2.5\,cm$. The largest lesion measured $2\,cm \times 2.3\,cm \times 3.6\,cm$. Histopathologically, the tumor was characterized by multiple cystic structures with anastomosing vascular channels (H & E staining ×50) (Fig. 2). Photomicrograph of the histologic specimen (H & E staining ×400) showed a high power view of the cystic structures demonstrating tall columnar endothelial cells without cytologic or nuclear atypia that lined the cyst-like spaces (Fig. 3). Immunohistochemistry showed that the cells compromising the tumor were positive for CD21, CD31, CD68, and CD163 (Fig. 4).

According to the histology and immunohistochemistry findings, the diagnosis of LCA of the spleen was thus established. The patient was discharged 6 days after the splenectomy. Her postsurgical course was uneventful, intermittent abdominal distention disappeared, and no surgery-related complications were found. After surgery, Aspirin (100 mg qd) was used to prevent thrombosis. One month after discharge, the patient's blood routine and clotting routine were normal, and Aspirin was stopped to use. The follow-up imaging findings at 16 months after surgery showed no signs of recurrence. From the patient perspective, the result of surgical treatment was satisfactory.

## 3. Discussion

Most of LCA cases are benign, and malignant LCA was reported only by 3 studies.[4–6] Fernandez et al[7] reported that the liver metastasis occurred 4 years after splenectomy for LCA.

The etiology of LCA remains unknown. Several studies have shown an association of LCA with immune system dysfunction, including Crohn disease, and Gaucher disease, but almost no association was reported between LCA and visceral tumors including renal cell cancer and seminoma.[8,9]

Usually, patients with LCA present no obvious symptoms. Splenomegaly, abdominal pain, thrombocytopenia, and anemia are common clinical presentations. In most cases, LCA, as a neoplasm of the spleen, is found in patients who undergo splenectomy for other reasons or medical examination for anemia or thrombocytopenia.

LCA is multiple, and few solitary neoplasms were reported. To date, imaging examination cannot determine whether the lesion is benign or malignant because some other neoplasms of the spleen such as Kaposi sarcoma,[10] angiosarcoma hemangiomatosis,[11] hamartoma,[12] lymphoma,[13] hemangioendothelioma, and angiosarcoma,[14,15] share similar appearance to LCA. In

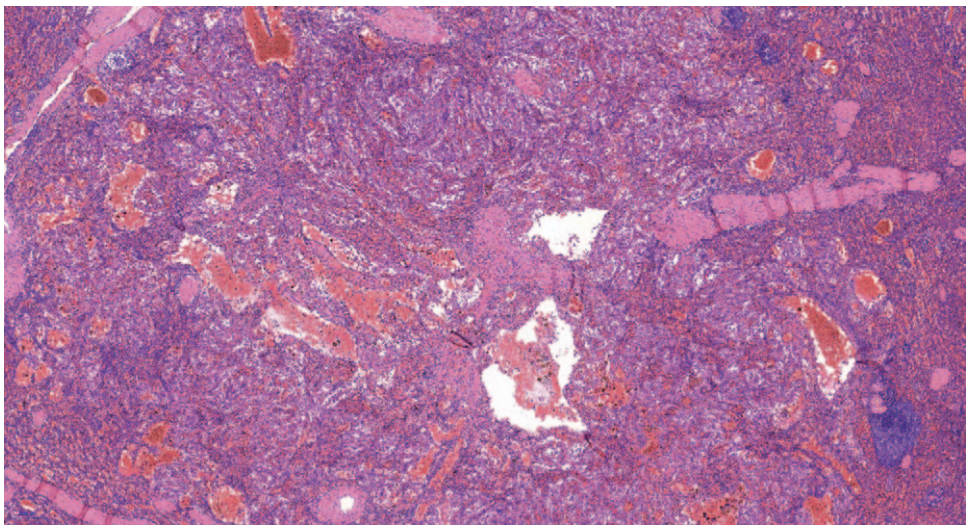

**Figure 2.** Photomicrograph of the histologic specimen (H & E staining,×50) shows that the tumor has multiple cystic structures with anastomosing vascular channels and cyst-like hemorrhagic spaces (H & E staining,×50).

addition, no correct diagnosis of LCA can be established before surgery. The final diagnosis depends on the histological and immunohistochemical results. Tumor markers including Ets-related gene, friend leukemia integration 1 transcription factor, vascular endothelial growth factor receptor 2, Claudin-5, lymphatic vessel endothelial hyaluronan receptor 1, and Wilms tumor-1, especially used alone, cannot establish a final diagnosis of LCA, as the expression of these markers can also be detected in other tumors.[16] LCA unique to the spleen is an uncommon primary vascular neoplasm of the spleen that originates from the littoral cells lining the splenic red pulp sinuses. But the tumor cells show a slight immunophenotypic difference from this normal cellular component of the spleen. In addition to CD34, CD21, CD31, and focal CD68 have been demonstrated in normal spleen-lined cells. Currently, the histiocyte marker CD163 is present in the littoral cells of LCA.[17] Histopathologically, the feature of LCA is CD31(+), CD163(+), CD68(+), and CD21 (+).[18] However, fine-needle aspiration is not recommended due to the possibility of bleeding and malignant cells dissemination. From a clinical point of view, splenectomy is performed for both diagnostic and therapeutic purposes.

Compared with open surgery, laparoscopic splenectomy has its potential advantages such as less bleeding, shorter post-operation stay, and less postoperative pain. Marzetti et al[19] reported a 79-year-old female with cardiopathy and obesity who underwent laparoscopic splenectomy, and no postoperative complications or morbidity was observed. Cai et al[20] also supported laparoscopic splenectomy and found laparoscopic splenectomy for LCA was safe, feasible, with favorable postoperative clinical outcomes. However, due to its rarity,

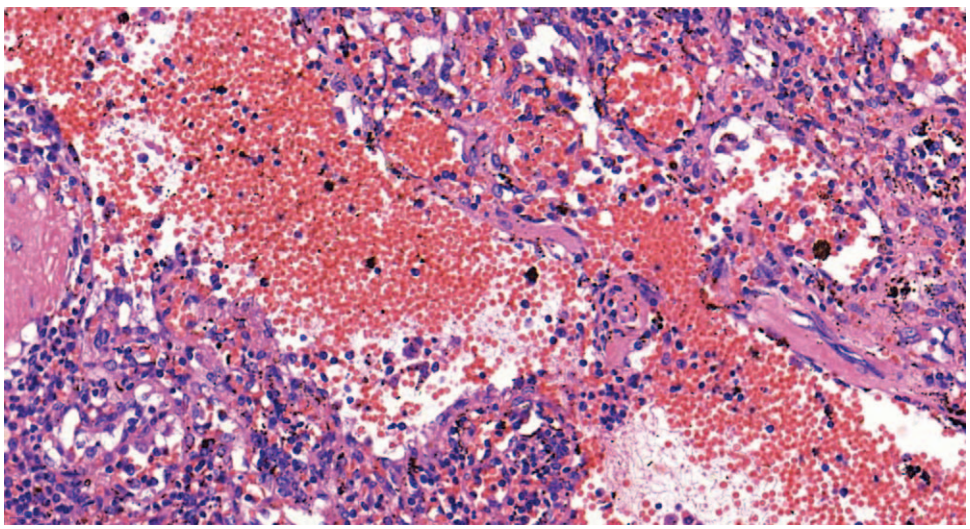

**Figure 3.** Photomicrograph of the histologic specimen (H & E staining,×400) demonstrates tall columnar endothelial cells without cytologic or nuclear atypia that line the cyst-like spaces.

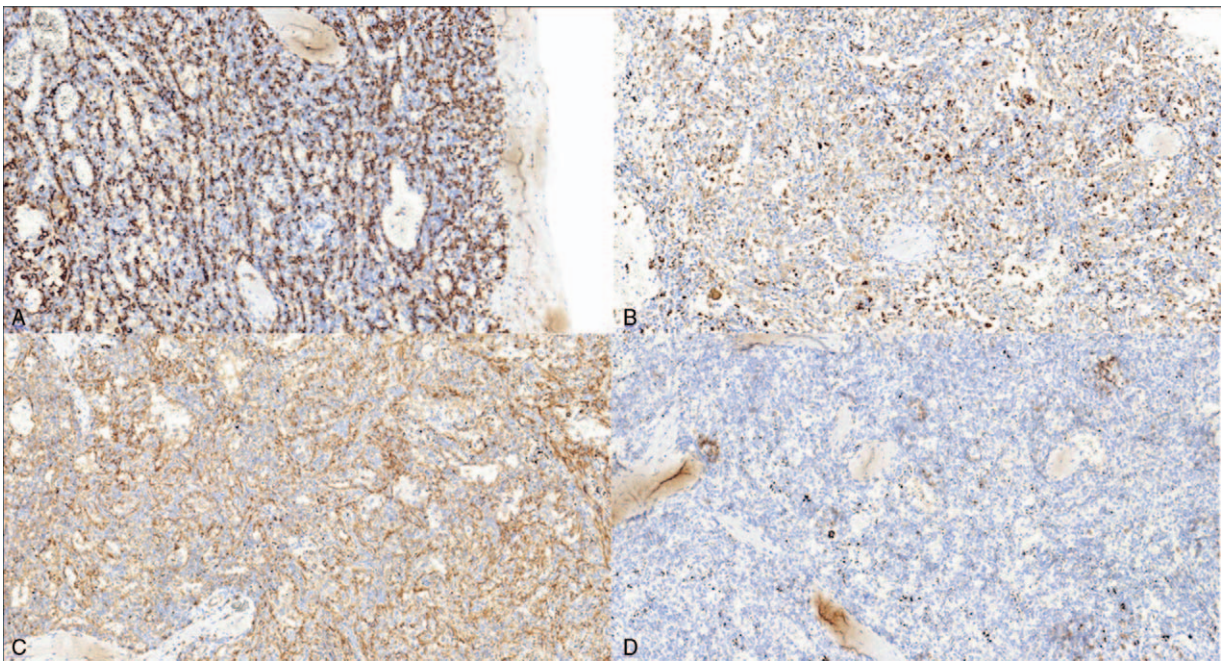


**Figure 4.** (A) The anastomosing channels lined cells expressed antigen CD163. (B) CD68 was positively expressed. (C) The anastomosing channels lined cells expressed antigen CD31. (D) CD21 was positively expressed.

only a few studies have reported laparoscopic splenectomy for LCA, and some concerns should be taken seriously: it is extremely difficult to perform laparoscopic splenectomy for massive splenomegaly, and laparoscopic splenectomy may lead to deterioration of oncological outcome. According to our experience in the present case, laparoscopic splenectomy for LCA was safe, and there were no signs of recurrence in the 20 months after the operation.

In conclusion, splenectomy is the main method for the purpose of diagnosis and treatment of LCA. The main "take-away" lessons of this case report were that LCA was a rare primary vascular neoplasm of the spleen, and laparoscopic splenectomy for LCA was safe and feasible, and postoperative course was uneventful. However, regular follow-up and long-time monitoring after splenectomy for LCA should be carried out because of its potential malignant biological behavior.

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## Author contributions

Man-Jiang Li wrote the case report; Xuan Zhou contributed the pathology analysis and provided the collection of pathological images; Li-Qun Wu and Yun-Jin Zang critically revised the intellectual content and contributed to the design of the paper; Jing-Yu Cao, Cheng-Zhan Zhu, and San-Shun Zhou provided the collection of literature.

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