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Case Report

Littoral Cell Angioma of the Spleen: A Case Report and Literature Review

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Abstract:

Littoral cell angioma (LCA) is a rare pathology of the spleen often found incidentally or with vague symptoms of abdominal pain, fatigue, or anemia. We present a case of a 45-year-old male with an incidental finding of littoral cell angioma during an emergent exploratory laparotomy for concern of perforated viscus. LCA is a generally benign lesion with minimal risk of malignancy; therefore management of these lesions are dependent on lesion size, potential for rupture, and patient comorbidities. If suspicious imaging features are present, splenectomy should be performed to rule out transformation to angiosarcoma. In this paper, we emphasize the importance of a multidisciplinary approach in managing complex surgical cases and highlight the need for further studies to better understand the natural history and optimal management strategies for LCA.

Key words: littoral cell angioma; splenectomy; malignant transformation

Introduction

Littoral cell angioma (LCA) is a rare, benign vascular tumor of the spleen first described in 1991 [1]. Originating from the littoral cells, a type of endothelial cell located along the inner walls of the splenic sinuses, these tumors exhibit both endothelial and histiocytic features [1]. LCA often presents with non-specific symptoms such as abdominal pain, fatigue, or anemia. It is frequently discovered incidentally during imaging or surgery for other conditions [1-2]. The rarity and non-specific presentation of LCA pose significant diagnostic challenges, with definitive diagnosis reliant on histopathological and immunohistochemical analyses [2]. While splenectomy is often curative for benign LCA, there is potential for malignant transformation, necessitating long-term follow-up. Our case presents a 45-year-old male with incidentally identified LCA of the spleen, highlights the diagnostic and therapeutic challenges, and reviews relevant literature to enhance understanding of this condition.

Case Presentation

Our patient is a 45-year-old male with an extensive medical history, including diabetes mellitus, hypertension, bipolar disorder, alcohol use disorder, and recurrent alcoholic pancreatitis. He also has a history of chronic gastritis and perforated peptic ulcer status post patch repair. The patient presented to the emergency department with sharp, diffuse abdominal pain with hematemesis that started four days after the patient was assaulted. Upon arrival, he was hemodynamically stable but diaphoretic and in acute distress with peritonitis. CT scan showed pneumoperitoneum with concern for a duodenal perforation. Preoperative labs showed a hemoglobin of 14.3, white cell count of 25.6, and total bilirubin of 3.4.

The patient was taken emergently to the operating room for exploratory laparotomy. Bilious abdominal fluid was found upon entry into the peritoneum. Exploration of the left upper quadrant revealed old blood clots and a splenic laceration. The laceration continued to bleed despite conservative attempts with suture repair and packing, therefore a splenectomy was performed.

Attention was then turned to the right upper quadrant to address the perforation. The stomach and duodenum were severely fibrotic from chronic gastritis and prior omental patch repair. This area was examined thoroughly for perforations; however no overt perforation could be identified. Due to the bilious fluid in the abdomen and the patient's prior history of complicated peptic ulcer disease, a distal gastrectomy with Roux-en-Y reconstruction with vagotomy was performed. The patient was then closed and taken to the intensive care unit for further management. Postoperatively, the patient was ultimately extubated and advanced to a regular diet.

Surgical pathology of the distal stomach showed gastritis and focal intestinal metaplasia. Pathology of the spleen revealed littoral cell angioma, confirmed by immunohistochemical staining positive for CD31, CD68, and vimentin [3] (Figure 1-4).

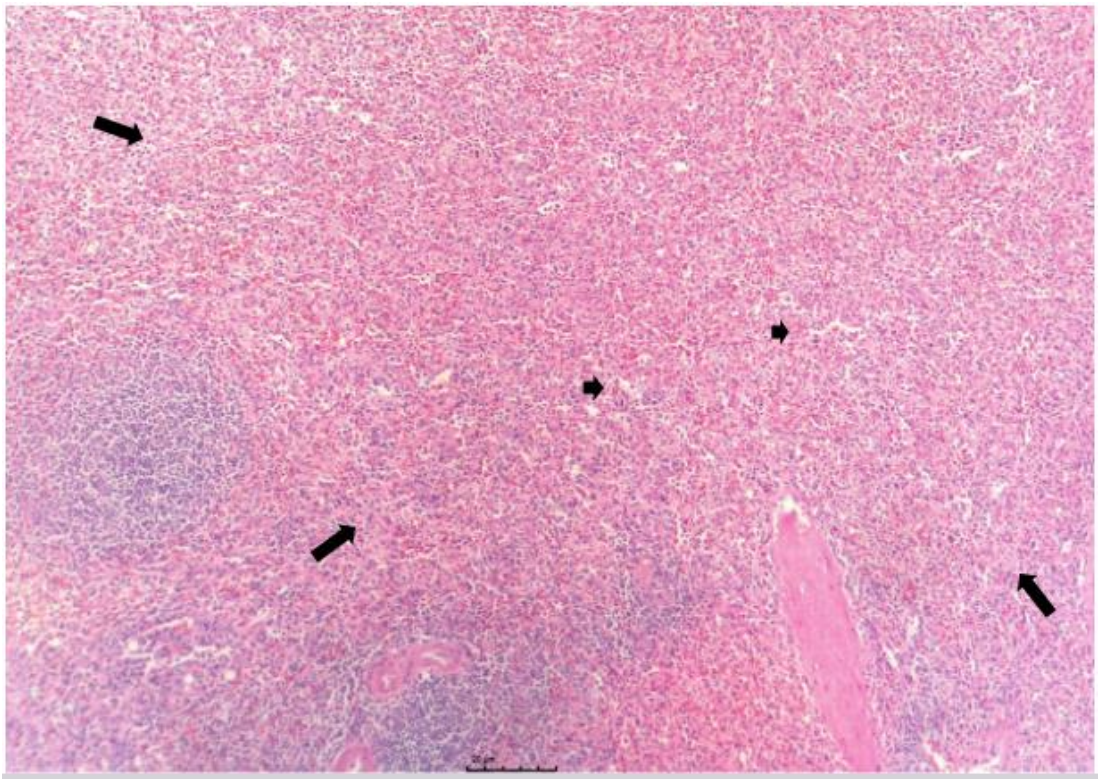

Figure 1: Spleen showing expanded red pulp (arrow), anastomosing and tortuous vascular channels (arrow head) (H&E 40 x) (magnification bar unit 20uM).

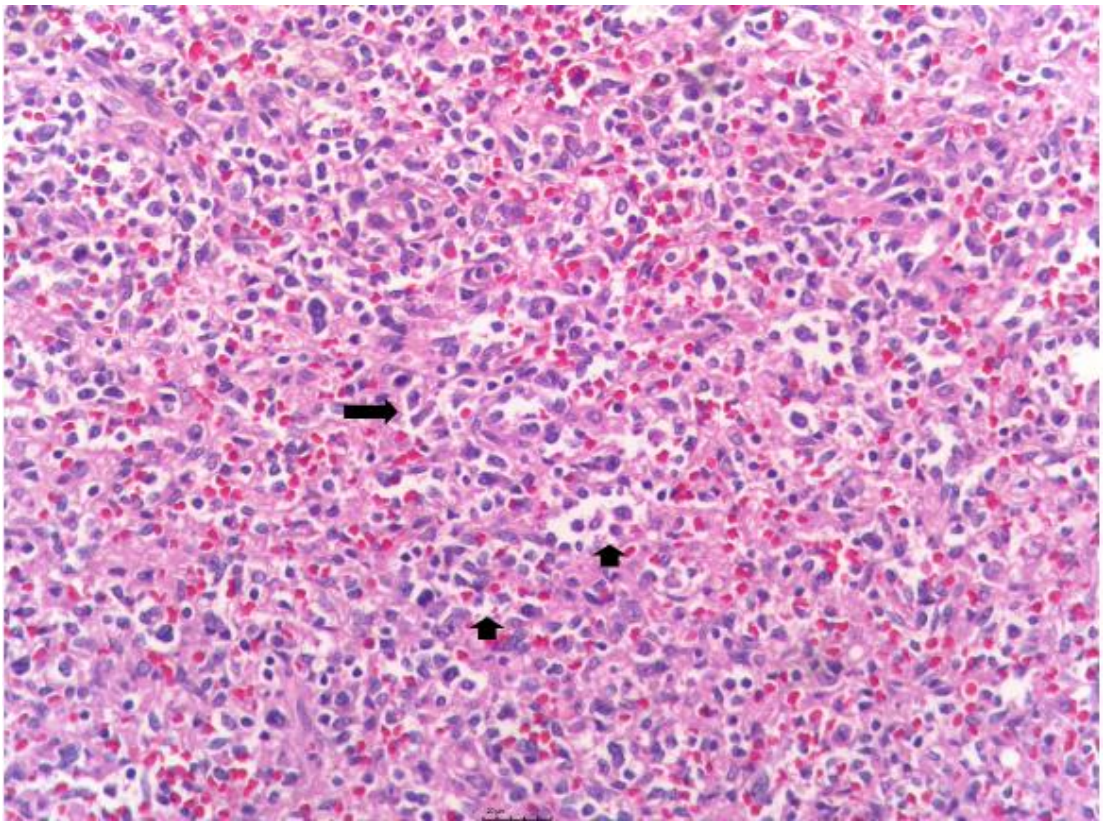

Figure 2: Anastomosing and tortuous vascular channels (arrow), with sloughing of tumor cells into vascular spaces (arrow head) (H&E 200X) (magnification bar unit 20uM).

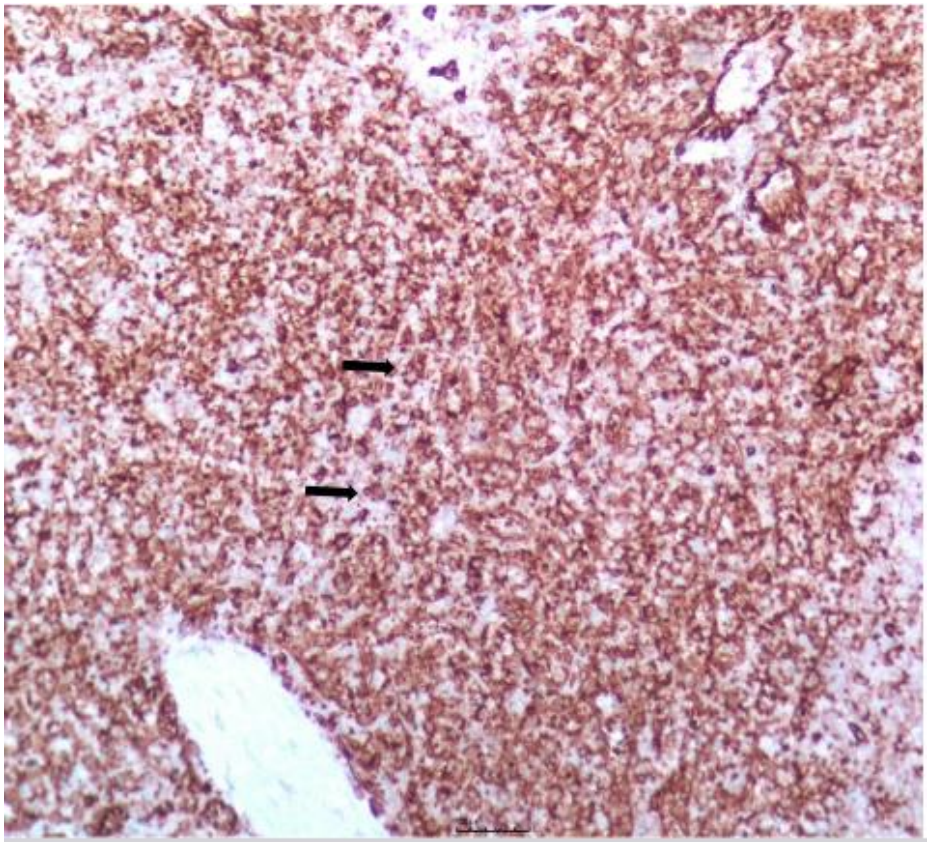

Figure 3: Tumor cells are positive for CD31 (arrow) by immunostain showing endothelial differentiation (H&E 100X) (magnification bar unit 20uM).

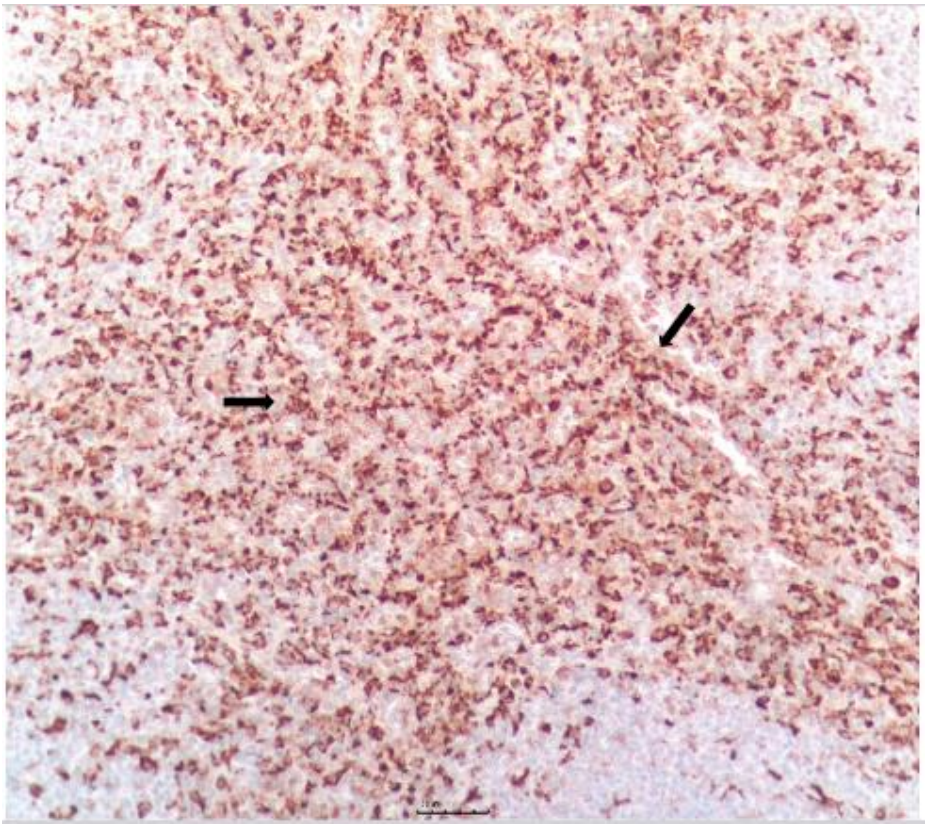

Figure 4: Tumor cells are positive for CD68 (arrow head) by immunostain showing histiocytic differentiation (H&E 100X) (magnification bar unit 20uM).

Discussion

Littoral cell angioma (LCA) is a rare benign vascular tumor of the spleen, with fewer than 500 cases reported in the literature [2]. LCAs arise from the littoral cells lining the red pulp sinuses of the spleen, which exhibit both endothelial and histiocytic features. This dual nature characteristic facilitates its diagnosis, as these cells uniquely express markers seen in both vascular and macrophage lineages. The pathophysiology of LCA involves the proliferation of littoral cells into anastomosing vascular channels, leading to the formation of the tumor [1].

Although the etiology of LCA remains unclear, it is often associated with certain immunologic disorders [4] such as hemophagocytic syndrome, Castleman disease, and lymphoproliferative disorders, which may contribute to its development. Chronic inflammation or immune dysregulation are proposed as potential triggers for the proliferation of littoral cells. Moreover, despite its benign nature, there have been rare reports of malignant transformation of LCA, including the development of angiosarcoma, a highly aggressive tumor of endothelial cells. This underscores the importance of long-term monitoring, as the transformation into angiosarcoma can lead to poor prognosis if not detected early [5].

LCAs can present with a wide range of clinical manifestations, from asymptomatic cases detected incidentally to symptomatic cases with significant splenomegaly, abdominal pain, or symptoms related to hypersplenism such as anemia, thrombocytopenia, or leukopenia [2, 6]. Patients may present with vague abdominal discomfort or fullness due to the enlarged spleen impinging on surrounding structures. Occasionally, LCA may present with acute events like splenic rupture, leading to intra-abdominal hemorrhage and requiring urgent surgical intervention [7]. In the context of our patient, this angioma may have ruptured in the days before admission, leading to the unexpected finding of bleeding in the left upper quadrant.

Due to the nonspecific and sometimes absent symptoms associated with littoral cell angioma, this condition is very difficult to diagnose preoperatively [2, 6]. Furthermore, the clinical presentation may also overlap with other clinical conditions affecting the spleen [4]. A high index of suspicion and detailed diagnostic evaluation to arrive at an accurate diagnosis. Hematologic abnormalities in peripheral blood counts due to hypersplenism are common in LCA and can complicate the clinical picture, requiring thorough investigation to rule out other hematologic disorders [4].

Multiple imaging modalities are commonly used for the initial detection and evaluation of littoral cell angioma. However, they rarely provide a definitive diagnosis due to the similar appearance of LCA to both benign splenic neoplasms and malignant tumors [8]. Ultrasound is often the first imaging technique employed, but it offers low specificity and is rarely helpful in diagnosing LCA due to its widely varying findings. Ultrasound features of LCA range from a heterogeneous echotexture without specific nodules to lesions that appear hyperechogenic, hypoechogenic, or echogenic [9-13]. Contrast-enhanced computed tomography (CT) scans and magnetic resonance imaging (MRI) are more specific modalities. On CT, LCA usually appears as multiple hypoattenuating nodules that may enhance heterogeneously after contrast administration [8]. In our patient, the CT scan revealed a normal-appearing spleen.

However, a small LCA located deep in the splenic parenchyma may not be readily visible on a CT scan. MRI of the spleen can aid in the diagnosis by revealing iso- or hypointense lesions on T1-weighted images and hyperintense lesions on T2-weighted images, with variable contrast enhancement patterns reflecting the vascular channels within the tumor [8] (Figure 5). Moreover, diffusion-weighted imaging (DWI) sequences, apparent diffusion coefficient (ADC) maps, and dynamic contrast graphic images may be helpful as diagnostic tools in the differential diagnosis of malignant and benign splenic processes [8].

In cases suspicious of malignant neoplasia, characterized by intense vascularization on MRI, some case reports recommend further evaluation with positron emission tomography (PET) scans [8, 14]. PET imaging can assess metabolic activity, aiding in the differentiation of benign from malignant lesions. Typically, PET scans show no increased fluorodeoxyglucose uptake in the splenic abnormalities identified on prior CT examinations [8]. Advancements in imaging modality and tailored imaging protocols help significantly narrow down the differential diagnosis, though pathological data is still needed for a definitive diagnosis.

Performing fine needle aspiration (FNA) cytology or image-guided biopsy could theoretically assist in diagnosing LCA [2]. However, due to the vascular nature of LCA, such procedures carry a significant complication rate (10.3%) [15]. Our patient with LCA, for example, required splenectomy for hemostasis. Thus, rather than risk uncontrolled bleeding from disrupting the splenic capsule during biopsy, splenectomy may be a more favorable surgical option for both diagnosis and therapy [16].

The characteristic histologic features of LCA include anastomosing vascular channels lined by tall endothelial cells that exhibit histiocytic features, such as phagocytosis and hemophagocytosis. These cells are positive for endothelial markers (CD31), histiocytic markers (CD68), and vimentin, but negative for CD34 and factor VIII-related antigen, which differentiates LCA from other vascular tumors of the spleen [3]. Immunohistochemical staining is crucial for diagnosing LCA and involves positive staining for CD31, CD68, and variably for CD21 and CD163, with negativity for CD34, which distinguishes LCA from other splenic vascular tumors [2, 17]. This immunophenotypic profile is key to its diagnosis and helps exclude other neoplastic processes. Confirmation of these findings through microscopic evaluation and immunohistochemistry ensure accurate differentiation from other splenic lesions.

In symptomatic patients, splenectomy addresses clinical manifestations related to splenomegaly and hypersplenism, such as abdominal discomfort and hematologic abnormalities [18]. For asymptomatic patients in whom LCA is incidentally discovered, the management strategy can vary. Observation without surgical intervention may be appropriate, given that LCA is a benign lesion with minimal risk of malignancy. The decision to biopsy should proceed with caution given the high risk of complications. The decision to proceed with surgery depends on factors such as the size of the lesion and spleen, the potential for rupture, and patient comorbidities. However, splenectomy is often pursued to definitively rule out malignancy, particularly in cases where imaging or clinical presentation raises concerns for littoral cell angiosarcoma (LCS), a rare but aggressive malignancy that can mimic LCA radiologically and clinically [16].

Postoperatively, patients generally have a favorable prognosis, with alleviation of symptoms and minimal risk of recurrence. Regular follow-up is recommended, particularly to manage postoperative complications, such as infections due to asplenia, and to monitor for rare instances of malignant transformation on an accessory spleen or metastasis after splenectomy [2].

Due to the rarity of this generally benign lesion, there is currently no guideline for postoperative monitoring for malignant transformation. For patients with malignant LCA or those with high-risk features, adjuvant therapies including chemotherapy or radiotherapy may be considered, although data on their efficacy are limited. A regimen of etoposide, paclitaxel, and vincristine appeared to have an anti-tumor effect in one case of malignant LCA [5]. A cohort of 25 cases reported that LCA expresses vascular endothelial growth factor receptor (VEGFR)-2 and -3, which suggests another avenue of therapies to target littoral cells [19]. The decision to avoid splenectomy should incorporate factors such as lesion size, imaging features, and patient comorbidities, alongside a

multidisciplinary team. Regular follow-up is recommended to monitor for rare complications, including malignancy or spontaneous rupture, as well as to manage potential postoperative infections in patients who undergo splenectomy.

Conclusion

This review of littoral cell angioma underscores the rarity and diagnostic challenges of this vascular neoplasm. There are currently no concrete guidelines for management of littoral cell angioma due to the rarity of the diagnosis. As littoral cell angioma is generally a benign lesion with minimal risk of malignancy, we recommend conservative management with serial imaging for small incidentally found lesions without suspicious features. Lesions that are large, symptomatic, or have suspicious features should undergo splenectomy due to the high bleeding risk in splenic biopsy. Further studies regarding the natural history of littoral cell angioma and the rate of malignant transformation to angiosarcoma are required to guide clinicians for long-term management of littoral cell angioma.

Conflicts of Interest

There are no conflicts of interest to disclose.

Informed consent was obtained from all individual participants included in the study.

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