Littoral Cell Angioma: A Rare Diagnosis

Jacob Elnaggar, PhD[1], Donnell White, III, PhD[1], Colin Cunningham, MD[2], and Brian Boulmay, MD[2]

[1]School of Medicine, Louisiana State University Health Sciences Center, New Orleans, LA, 70112

[2]Section of Hematology and Oncology, Louisiana State University Health Sciences Center, New Orleans, LA, 70112

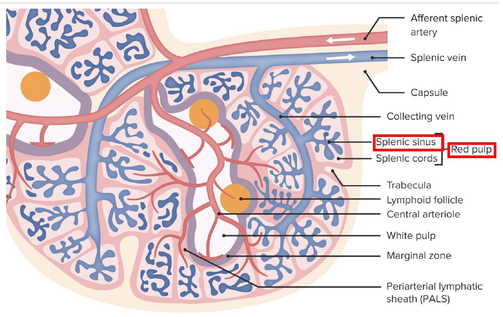
Introduction

Littoral cell tumors are rare primary splenic neoplasms which include the more common, typically benign littoral cell angioma (LCA); the less common, potentially malignant littoral cell hemangioendothelioma (LCHE); and the aggressive littoral cell angiosarcoma (LCAS)

LCA arise from the littoral cells lining the sinusoids of splenic red pulp that was first described in 1991 by Falk et al in Frankfurt, Germany

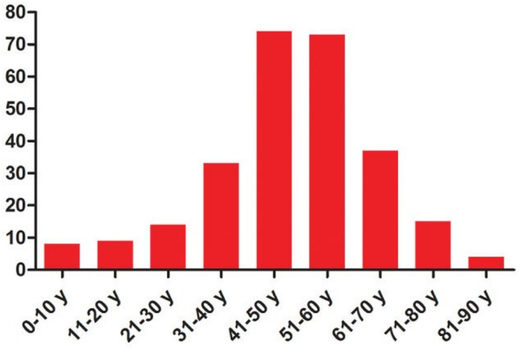
Due to its extremely low incidence and limited awareness, fewer than 500 cases have been documented since it was first described 34 years ago.

Given this lack of data, there is little known about the epidemiology, natural history, pathogenesis, clinical manifestations, and prognosis. There is also conflicting opinion as to the management of patients



Basic demographics

Total number of LCA patients	435	171(English)/264(Chinese)
Male/Female	0.90	192/213
Age (mean, SD)	48.2(16)	267
Age range	26 d–86 y	267



Clinical manifestations

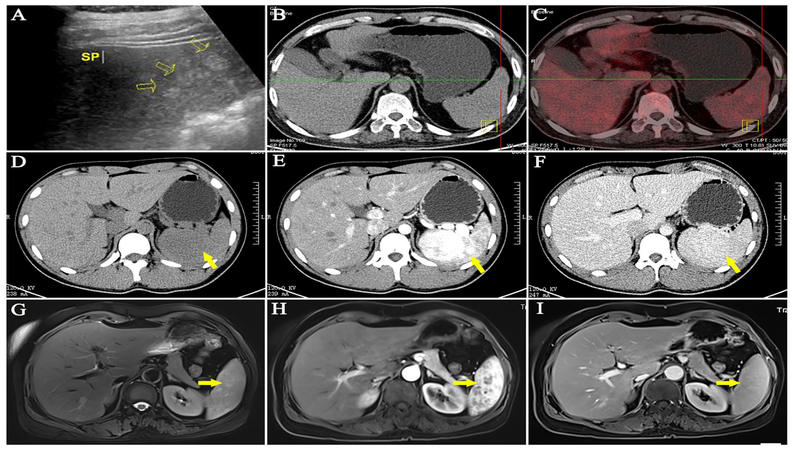
Asymptomatic or incidental finding	49.7%	156/314*
Upper abdominal discomfort[$]	39.2%	123/314
Fever	5.4%	17/314
Fatigue	8.3%	26/314
Dizziness	2.9%	9/314
Purpura	2.2%	7/314
Loin pain	1.9%	6/314
Splenomegaly	69.7%	255/366[#]
Thrombocytopenia	48.8%	127/260[&]
Anemia	31.4%	69/220[^]

Case Description

A 56-year-old man presented to the emergency department with a chief complaint of abdominal pain and constipation. CT scan revealed splenomegaly (16 cm) with multiple hypoenhancing nodules in the splenic parenchyma. Differential diagnosis included lymphoma, solid tumor metastasis, or venous lakes. He was subsequently discharged and referred to oncology.

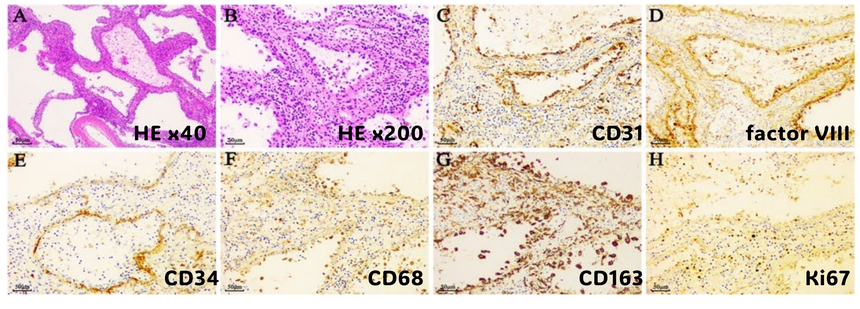
A follow-up ultrasound confirmed splenomegaly (16.5 cm) with heterogeneous echotexture but no discernible masses. The patient reported feeling well overall, with no fever, chills, abdominal pain, night sweats, or lymphadenopathy. Initial lab work was unremarkable, including tumor markers (CA 19-9, PSA, AFP) and routine blood tests.

Abdominal CT with Splenomegaly and LCA



Six months later, repeat CT imaging showed stable splenomegaly with similar lesions. The differential was expanded to include lymphoma, atypical hemangiomas, LCA, and sarcoidosis, while metastasis was considered less likely due to lesion stability. A fine-needle aspiration of the spleen was performed. The biopsy showed a vascular lesion consistent with a littoral cell origin, confirmed by immunoreactivity for CD68 and CD31, but not CD34. While littoral cell angioma was suspected, the diagnosis was uncertain due to the rarity of the condition and limited sample material.

Histopathology and IHC Characteristics



Given the uncommon nature of LCA, there was conflicting guidance regarding management in the medical literature. The case was presented at multi-disciplinary tumor board, and no consensus was reached between observation and splenectomy. After discussing options, the patient chose monitoring due to his asymptomatic course. A year later, a repeat CT showed stable lesions, and the patient continues to be observed.

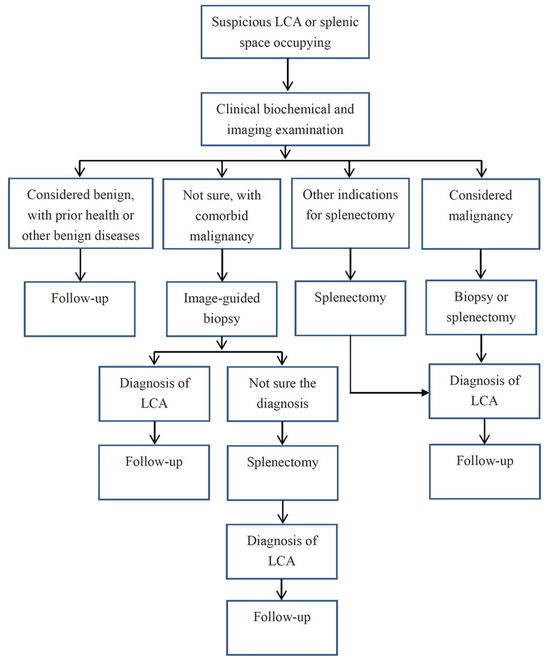
Treatments and Prognosis

Open splenectomy	81.0%	325/401
Laparoscopic splenectomy	19.0%	76/401
Diagnosis by percutaneous splenic biopsy	83.3%	10/12
No recurrence or metastasis	91.4%	159/174
Recurrence or metastasis	0.57%	1/174
Death	9.2%	16/174

Discussion

LCA, though rare and often benign, presents significant diagnostic and management challenges due to the lack of specific guidelines and limited literature. Splenectomy is curative but carries risks. This case exemplifies the need for a careful, multidisciplinary approach, considering the patient's preferences, particularly due to the limitations in current evidence for managing LCA.

Proposed New Mode of Diagnosis and Treatment for LCA



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