Title: Littoral Cell Angioma: A Rare Diagnosis

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Introduction

Littoral cell angioma (LCA) is a rare primary vascular tumor of the spleen. Originating from littoral cells, these specialized endothelial cells line the red pulp sinuses of the spleen and are involved in removing damaged red blood cells from circulation.

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.

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.

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.

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.