A Splenic Surprise: Navigating Littoral Cell Angioma in an Elderly Patient

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Introduction: Littoral Cell Angioma (LCA) is a rare primary vascular tumor of the spleen. Typically, LCAs are incidentally found as a single or multiple benign tumor(s) appearing among the littoral cells which line the venous sinuses of splenic red pulp. While less common, there are also reported cases of malignant subtypes of LCAs. We present a case who was admitted out of concern for intra-abdominal hemorrhage, later finding multiple LCAs identified following splenectomy.

Case report: A 72-year-old female with a past medical history of hypertension and iron deficiency anemia who presented to the Emergency Department following a near-syncopal episode. Before the near fall, she developed abdominal pain, diffuse weakness, and vomiting. She denied prior episodes of syncope or a traumatic history. The patient's pertinent vitals were a blood pressure of 88/58 mmHg and a heart rate of 106 beats per minute. On physical exam, ecchymosis over the thoracoabdominal area was appreciated, and the patient was alert and fully oriented. Laboratory studies were concerning for a hemoglobin of 6.8, and platelets 67,000 prompting multiple transfusions of red blood cells and cryoprecipitate. CT abdomen was obtained and revealed hepatosplenomegaly, perihepatic fluid, peri-splenic fluid, and a splenic hematoma. An active Grade 3 splenic rupture prompted splenectomy and a partial liver lobectomy. Subsequent CT imaging was concerning for portal vein thrombosis and two noncommunicating fluid collections in the left upper quadrant and subscapular hepatic region. Her hospital course was complicated by urosepsis and worsening renal function, prompting intravenous antibiotics and one-time hemodialysis. Pathology from the spleen was significant for multiple hemorrhagic nodules with closely placed sinusoids and cuboidal endothelial cells, compatible with littoral cell angioma. Congo red staining was also concerning for amyloidosis. Oncology was consulted and the patient was started on an outpatient multi-drug chemotherapy regimen. The patient, however, had passed away at her home two months after her admission.

Discussion: Littoral cell angiomas are typically an incidental finding on abdominal imaging, although there have been rare cases of splenic rupture, as seen in this patient. Diagnosis of an LCA involves identifying characteristic morphology (tall columnar endothelial cells lining cyst-like spaces) and utilizing immunohistochemical studies. LCAs are comprised of cells that express histiocytic markers, such as CD68 and lysozyme, and endothelial markers, like CD31 and factor VIII. This contrasts with the surrounding splenic cells, which only have endothelial markers. The current recommended treatment for LCAs is splenectomy with follow-up, given their risk of malignant transformation. In the present case, the patient required a splenectomy secondary to spontaneous splenic rupture, likely due to LCA burden, a rare outcome for this condition. Although LCA has been associated with many inflammatory disorders and malignancies, there has been no documented case associated with amyloidosis. This case emphasizes the intricate interplay of complex splenic pathologies in elderly patients with multiple comorbidities, underscoring the significance of early recognition and the implementation of comprehensive management strategies to enhance outcomes in these challenging scenarios.