Littoral Cell Angioma – A Rare Primary Splenic Lesion with Malignant Potential

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Introduction

Littoral cell angioma (LCA) is a rare primary splenic tumor arising from littoral cells which lined the red pulp sinuses of the spleen. Clinically, these tumors present with vague symptoms including splenomegaly, abdominal pain, thrombocytopenia, and fatigue.

Since its first description in the literature by Bhatt et al. in 1991, LCA was believed to be benign. However, several more recent reports have described LCA with malignant histologic features or malignant behavior. Additionally, LCA has been associated with other visceral tumors, including non-small cell lung cancer, pancreatic neuroendocrine tumors, papillary thyroid cancer, and colorectal adenocarcinoma. For this reason, correct identification of this neoplasm is essential so that patients can undergo long-term surveillance for further malignancies.

On imaging, LCA resembles lymphangioma, hamartoma, hemangioma and lymphoma. As such, pathological analysis of the tumor remains essential for definitive diagnosis.

Methods

A 74 year old female presented to the hospital with recent chest pain, abdominal pain, and 10-pound weight loss. Upon imaging, abdominal CT with contrast showed splenomegaly and innumerable hypodense lesions. Bone marrow biopsy and flow cytometry performed were suggestive of an underlying neoplastic process but were not conclusive. Subsequent CT guided spleen biopsy was consistent with littoral cell angioma with a background proliferation of small clonal B-cells. Surgery was indicated, and a total splenectomy was performed. The specimen was submitted to the pathology laboratory for evaluation.

Results

Macroscopic examination of the 717 grams, 19.5 x 12 x 6.5 cm spleen specimen revealed a spongy nodular surface with numerous small ill defined cysts. Histology showed multiple nodules of cystic vascular lesions with dilated irregular anastomosing lumens (Figure 1) lined by cuboidal cells (Figure 2) and rare eosinophilic granular cells (Figure 3). By immunohistochemistry the lining cells are positive for endothelial markers, factor VIII (Figure 4) and CD31 (Figure 5), as well as histiocytic marker CD68 (Figure 6), ratifying the diagnosis of littoral cell angioma. Additional immunostains for CD3 and CD20 were negative (Figure 7), excluding the possibility of associated lymphoma. There was no atypia, necrosis or high mitotic activity to suggest malignancy.

Figures

TUMOR **SPLEEN**

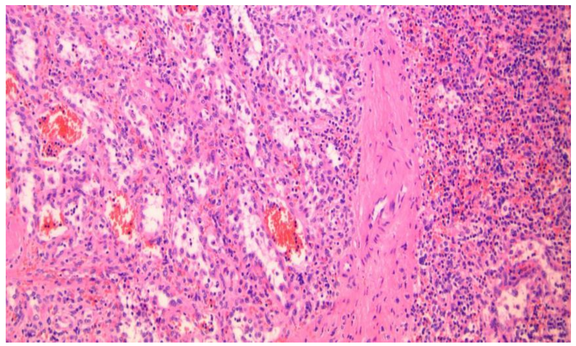

Figure 1 (4X)

Results (Continued)

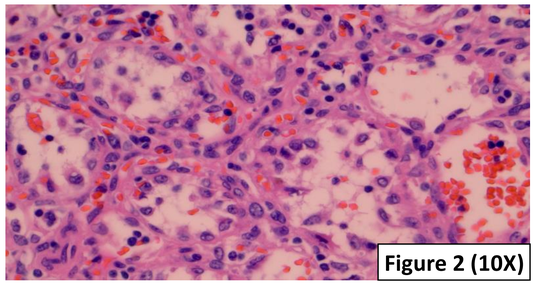

Figure 2 (10X)

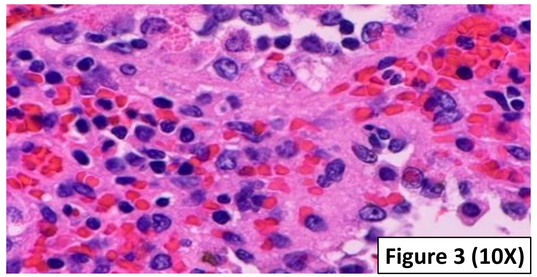

Figure 3 (10X)

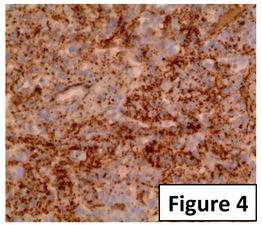

Figure 4

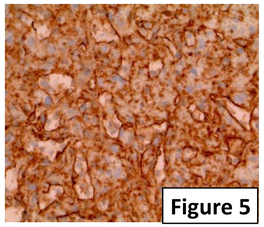

Figure 5

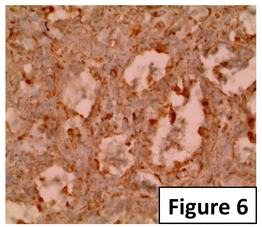

Figure 6

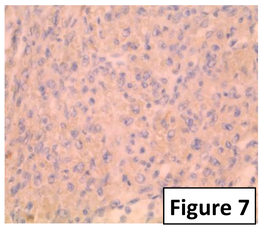

Figure 7

Conclusions

Littoral cell angioma (LCA) is a rare, typically benign, primary vascular lesion of the spleen, arising from littoral cells. LCA most commonly presents with splenomegaly, and has multifocal as opposed to solitary lesions. Histologically, these neoplasms exhibit sinus-like anastomosing vessels lined with tall and plump endothelial cells with focal papillary fronds and normal splenic sinuses at the periphery of the lesion. It is characterized immunohistologically by dual endothelial and histiocytic features, negative for CD 34 and CD 8, while positive for CD 68 and CD 21. LCA is distinguished from littoral cells by its poor expression of formin homology domain protein 1 (FHOD1).

While typically benign, cases of malignant transformation have been noted in the literature, particularly in cases of massive preoperative splenomegaly defined by one literature review as a splenic weight greater than or equal to 1500 grams and/or splenic length greater than 20 cm preoperatively. Additionally, LCA has been rarely associated with visceral malignancies such as colorectal carcinoma, pancreatic neuroendocrine tumors, non-small cell lung cancer, and papillary thyroid cancer. Therefore, familiarity with this rare lesion is essential to render a correct diagnosis and properly monitor these patients postoperatively for further complications such as recurrence or dissemination.

References & Acknowledgements

To view the complete list of references and submitted abstract, please point your phone's camera at the QR code to the left and click on the link that appears.