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Case Report
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A Rare Cause of Splenic Vascular Neoplasm Requiring Splenectomy: Littoral Cell Angioma

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Abstract

Littoral cell angioma is an uncommon primary vascular neoplasm of the spleen. It is made up of littoral cells that line the red pulp's splenic sinuses. Netra Rana et al. reported littoral cell angioma to be a benign incidental lesion. Only after histopathology and immunohistochemistry testing can a final diagnosis be made. Immunohistochemistry is a useful tool in diagnosing littoral cell angioma because it can demonstrate the distinct hybrid endothelial-histiocytic phenotype of littoral cells, which aids in making an accurate diagnosis. We present a rare etiology of splenic neoplasm treated with splenectomy. To our knowledge, there are only a few cases reported in the medical literature.

Keywords: Littoral cell angioma; Splenomegaly; Splenectomy

Introduction

Littoral Cell Angioma (LCA) is an uncommon primary vascular neoplasm of the spleen seen in the red sinus shore cells of the Reticuloendothelial System (RES) and was first discovered by Falk et al. in 1991 [1-3]. The major clinical manifestation is isolated splenomegaly of unknown etiology and the diagnosis practically demands splenectomy. Splenectomy because the diagnosis of Littoral cell angioma is confirmed *via* a histological study. In this article, we would like to report a rare case encounter of splenic vascular neoplasm.

Case Presentation

A 60-year-old female with a past medical history of hypertension, sarcoidosis, lower leg edema and prior history of thyroid disease presents to the office for possible splenectomy consideration. In 2021, the patient underwent a Computer Tomography (CT) scan of the abdomen and pelvis to investigate cholelithiasis. During this scan, multiple splenic lesions were incidentally discovered, along with isolated splenomegaly of 18 cm in greatest dimension, without associated hepatomegaly (Figure 1). The patient has not experienced any symptoms related to these findings, such as weight loss, fatigue, nausea, vomiting, diarrhea, constipation, pain, fevers, chills, or vision changes. She underwent extensive testing to try to determine the cause of these findings.

The tests included Complete Blood Count (CBC), Comprehensive Metabolic Panel (CMP), Lactate Dehydrogenase (LDH), uric acid, Angiotensin-Converting Enzyme (ACE), HIV/HEP panel, Computer Tomography of the thorax (CT thorax), and flow cytometry, all of which were negative. Due to her inconclusive laboratory results she underwent a splenic biopsy. The report demonstrated the presence of numerous polymorphous lymphocytes in a background of scattered spindle cells with mild atypia. Laparoscopic splenectomy was recommended to her.

Discussion

Primary tumors of the spleen are divided into two categories, lymphoid tumors and nonlymphoid tumors [4,5]. The most common non-lymphoid tumors are vascular tumors, which can be either benign or malignant [5]. Benign vascular tumors include hemangioma, lymphangioma, hemangioendothelioma, and Littoral Cell Angioma (LCA). Since its first cited case in 1991 [3], only 29 cases of LCA have been described in the literature, owing to its exceeding rarity.

LCA tumors share a unique, soft tissue capsule morphology. While derived from the red pulp sinuses lined by endothelial cells, these capsules express both endothelial and histiocytic antigens [6]. Due to this, LCA tumors have both immunophenotypic and morphologic features that differentiate them from other benign splenic tumors, including their expression of CD68 and Lysozyme macrophage markers and FVIII and CD31 endothelial markers, respectively [6].

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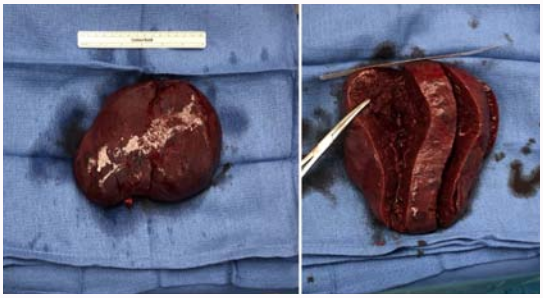

Figure 1: Image showing splenectomy.

Littoral cell angiomas can occur in all age groups but typically present in middle age with nonspecific complaints of abdominal pain and splenomegaly on sonographic and CT imaging. The appearance of splenic LCA varies widely, ranging from multiple blood-filled nodules grossly to solitary lesions that engulf the entire parenchyma of the spleen [6]. Microscopically, canonical findings of LCA include aggregates of eosinophilic globules, likely derived from phagocytosed red blood cells, lymphocytes and plasma cells [7]. Treatment of LCA is open or hand-assisted laparoscopic total splenectomy with tissue preservation for histologic analysis.

Of the 29 documented cases of LCA, 9 have been associated with other cancers, including visceral epithelial malignancies (colorectal adenocarcinoma, renal and pancreatic adenocarcinoma) as well as other liquid and solid cancers including non-Hodgkin's lymphoma, unspecified tumors of the liver and brain, epithelial ovarian cancer and a non-small cell lung cancer [6,8]. Given its diverse presentation, a thorough workup for a second visceral neoplasm may be necessary in these patients, although no predictive genetic or molecular associations have been found.

Conclusion

The incidence of tumors of the spleen are rare when compared to that of tumors of other visceral organs. Littoral cell angioma represents a rare, benign vascular tumor of the spleen that usually presents as non-specific abdominal pain and is typically diagnosed by histology. Although these tumors are benign, their association with other visceral neoplasms is still under investigation.

References

1. Rana N, Ming Z, Hui M, Bin Y. Case report: Littoral cell angioma of spleen. Indian J Radiol Imaging. 2009;19(03):210-12.
2. Peckova K, Michal M, Hadravsky L, Suster S, Damjanov I, Miesbauerova M, et al. Littoral cell angioma of the spleen: A study of 25 cases with confirmation of frequent association with visceral malignancies. Histopathology. 2016;69(5):762-74.
3. Falk S, Stutte HJ, Frizzera G. Littoral cell angioma. A novel splenic vascular lesion demonstrating histiocytic differentiation. Am J Surg Pathol. 1991;15(11):1023-33.
4. Bailey A, Vos J, Cardinal J. Littoral cell angioma: A case report. World J Clin Cases. 2015;3(10):894-9.
5. Fotiadis C, Georgopoulos I, Stoidis C, Patapis P. Primary tumors of the spleen. Int J Biomed Sci. 2009;5(2):85-91.
6. Du J, Shen Q, Yin H, Zhou X, Wu B. Littoral cell angioma of the spleen: Report of three cases and literature review. Int J Clin Exp Pathol. 2015;8(7):8516-20.
7. Michal M, Skalova A, Fakan F, Koza V, Svojgrová M. Littoral cell angioma of the spleen. A case report with ultrastructural and immunohistochemical observations. Zentralbl. Pathol. 1993;139:361-5.
8. Bisceglia M, Sickel JZ, Giangaspero F, Gomes V, Amini M, Michal M. Littoral cell angioma of the spleen: An additional report of four cases with emphasis on the association with visceral organ cancers. Tumori. 1998;84:595-9.