

Littoral Cell Angioma of Spleen: A Rare Case Report

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Abstract: The most common primary tumours of spleen are vascular in origin and are mostly benign. Littoral cell angioma is a rare primary splenic vascular tumour that arises from littoral cells lining the splenic red pulp sinuses. It is usually asymptomatic and is discovered incidentally on imaging for other pathologies. Here we present a case of littoral cell angioma that was discovered incidentally in a 17year old female being investigated for hepatosplenomegaly and anaemia and subsequently underwent splenectomy.

Keywords: Littoral cell angioma, Vascular neoplasm, Immunohistochemistry.

INTRODUCTION

The most common tumours of spleen are vascular in origin and are mostly benign. Littoral cell angioma is a rare primary splenic vascular tumour. It was first described by Falk *et al.* in 1991 [1]. This neoplasm takes origin from the littoral cells lining the red pulp sinuses with intermediate features between those of endothelial and histiocytic cells [2, 3]. In the literature only few cases have been reported.

CASE REPORT

A 17 year old female presented with intermittent epigastric pain, non-radiating type, not related to food intake and relieved spontaneously or with medication. There is no history of fever, weight loss, bone pains. She is born out of consanguineous marriage. Her childhood and teenage was nil remarkable. On general examination she is thin built girl with severe pallor, no abnormal facies, no lymphadenopathy. Physical examination revealed marked splenomegaly with mild hepatomegaly.

Laboratory tests revealed normocytic normochromic anaemia with hemoglobin of 5gm/dl and thrombocytopenia. Bone marrow aspiration and biopsy were done from right posterior iliac crest revealed erythroid hyperplasia with absent iron stores. No other abnormal cells noticed. Serological examination for Hepatitis A, B, C was negative. Liver function tests were within normal limits. Hemoglobin electrophoresis was normal. Abdominal ultrasonography revealed splenic span of 20cms. CT abdomen revealed moderate splenomegaly with many minute hypointense lesions. Based on clinical, radiological and laboratory findings preoperative diagnosis of tropical splenomegaly was

made and ruled out storage disorders and hemolytic anaemia. Subsequently patient underwent splenectomy. This patient had an uneventful post-operative recovery and has remained well at four months of post-operative follow-up.

Post-operative pathological examination revealed a markedly enlarged spleen weighing 2.2kgs and measuring 28x19x10cms. Cut surface revealed hemorrhagic appearance and fine mottling.



Fig. 1: Marked diffuse splenomegaly

Histopathological examination showed splenic capsule and trabeculae with few residual lymphoid follicles. Rest of the parenchyma showed dilated vascular sinusoids lined by plump endothelial cells with abundant cytoplasm and vesicular nuclei, many cells showed hemophagocytosis. Focal areas showed fibrosis, areas of hemorrhage and hemosiderin laden

macrophages. No atypical cells or atypical mitosis was noted.

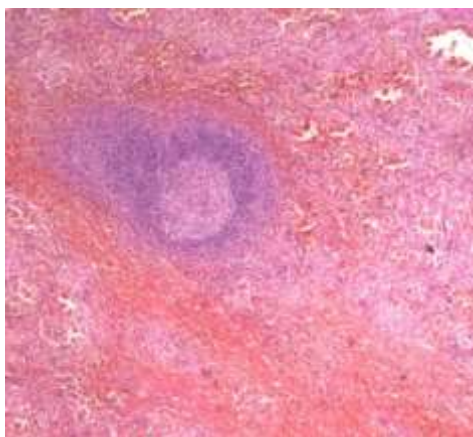


Fig. 2: (40x) H &E. Spleen with remnant lymphoid follicle and dilated vascular channels

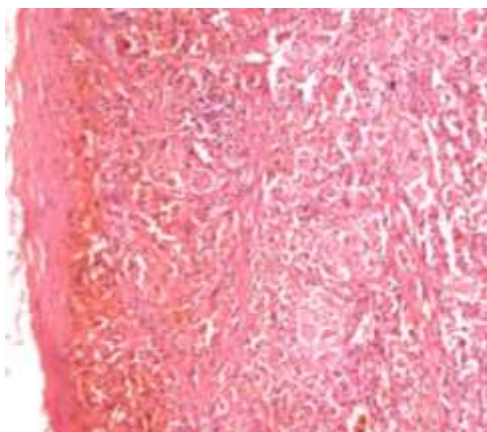


Fig. 3: (10x) H & E. Splenic capsule with dilated vascular channels and littoral cells

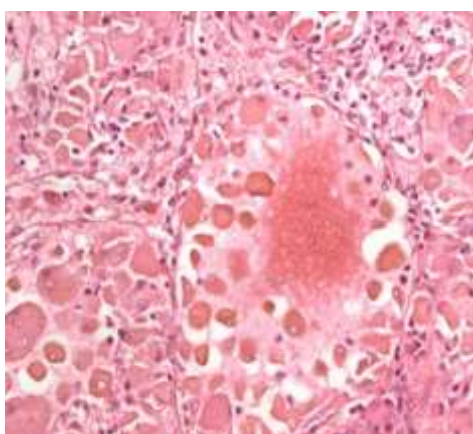


Fig. 4: (40x) H & E. Dilated vascular channels lined by plump and prominent endothelial cells

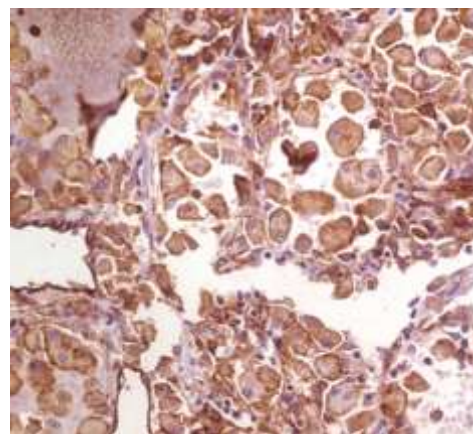


Fig. 5: (40x) Immunohistochemistry. CD 68 diffuse positivity in lining endothelial cells

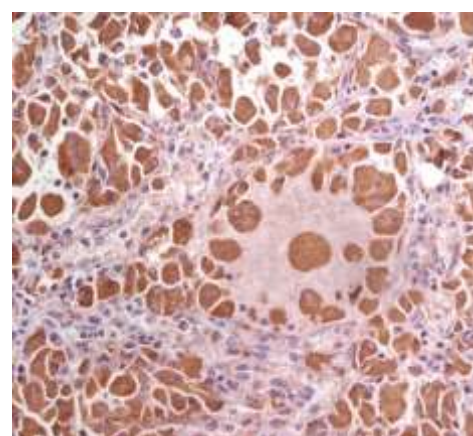


Fig. 6: (40x) CD 31 diffuse positivity in lining endothelial cells

Immunohistochemistry showed positivity for both CD 31 and CD 68 in the lining endothelial cells (clones 9G11 and KP1 respectively).

DISCUSSION

Littoral cells normally are seen lining the sinusoids in the red pulp. Littoral cell angioma of spleen arising from these littoral cells may present as an incidental finding [3]. It is usually diagnosed in patients undergoing splenectomy for splenomegaly of unknown origin or during radiographic evaluation of other abdominal processes [4]. LCA presents in all age groups and has no sex predilection. Patients present with either mild or moderate splenomegaly, pain abdomen, fever or hypersplenism [3].

Grossly they can appear as single or more often multiple nodular lesions [5]]. Less commonly, these lesions can be solitary or completely replace the splenic parenchyma [6]. Imaging studies like CT, MRI, USG and Tc 99mlabeled RBC scintigraphy are useful in evaluation [8].

Association with other visceral malignancies of colon, rectum, kidney, lung, pancreas, hepatocellular carcinoma and lymphomas is reported in the literature [7]. So a thorough clinical workup is needed to rule out these possibilities. A possibility of malignant potential of Littoral cell angioma of spleen is not firmly established [2]. Recently it has been reported to have malignant potential [8]. Two case reports have described variants of LCA with histological features of malignancy [9, 10].

Histologically it shows anastomosing vascular channels lined by tall columnar cells with hemophagocytosis. Papillary fronds can be seen. Lining cells are positive for both endothelial and histiocytic markers. CD 31, CD68, CD4 and EBV receptor of B lymphocytes is exclusively positive in Littoral cell angioma and negative for CD8 and CD34 [11]. Combination of morphological and immunohistochemical analyses that show a hybrid endothelial histiocytic phenotype establish the diagnosis of LCA [3].

Differential diagnosis includes other vascular tumours like hemangioma, hemangioendothelioma and infective etiology, hamartoma, angiosarcoma.

LCA Treatment includes surgical removal and can result in complete resolution.

CONCLUSION

Littoral cell angioma is a rare vascular tumour of spleen, the diagnosis of which is difficult to make preoperatively. Although vast majority of cases are benign, it may be associated with other malignancies and may itself also have malignant potential. The clinical presentation varies from pyrexia of unknown origin to splenomegaly, hypersplenism or as acute abdomen. Awareness of this entity littoral cell angioma, should be considered in the differential diagnosis of splenic hypodense lesions as splenectomy alleviates the symptoms. Long-term follow-up is advised for the possibility of development of synchronous tumours or metastatic lesions.

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