LITTORAL CELL ANGIOMA OF SPLEEN - A RARE CASE REPORT

Pathology

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ABSTRACT

INTRODUCTION: Littoral-cell angioma (LCA) is a rare benign vascular tumour of the spleen. There have been less than 250 cases reported in the literature. Recent reports have described it to be a Benign lesion with congenital and immunologic associations. We report a case of LCA of the spleen.

PRESENTATION OF CASE: A 09-year-old male patient was admitted to hospital with a one-month duration of intermittent upper abdominal pain and nausea. Imaging studies, including Ultrasonography and Colour Doppler Imaging of portal venous systems. Radiology shows moderate gross enlargement-16cm & normal in echotexture with no focal lesion. Splenic vein-10mm in caliber with partial thrombus formation. An open splenectomy was performed and his post-operative recovery was uneventful.

DISCUSSION: Littoral cell angioma of the spleen is a benign vascular tumour that has been infrequently reported in the English literature. While it does have malignant potential, the vast majority are benign. Diagnosis depends on the expression of endothelial markers like CD31 and histiocytic markers like CD68. Malignant potential is enhanced by the presence of splenomegaly as well.

CONCLUSION: This rare condition is made by the presence of rare vascular tumour in the spleens as well.

KEYWORDS

Splenic tumour, Littoral cell angioma, Benign vascular tumours.

INTRODUCTION

Littoral cell angioma of the spleen is a rare vascular tumor that was first described by Falk et al. [1] in 1991. The tumor arises from the littoral cells in the splenic red pulp sinuses. Littoral cell angioma affects both men and women equally with no specific age predilection. It is usually asymptomatic and is discovered incidentally. Some of the patients with this entity may present with symptoms of hypersplenism such as anemia, thrombocytopenia, and splenomegaly, the latter seen in almost all patients with littoral cell angioma. Clinically, a majority of the littoral cell angiomas described in the literature have been found to be benign in nature, but several reports have described malignancy in the littoral cell angioma [2]. Definite evaluation can only be done at pathology. Most of the cases of littoral cell angioma described in the literature have been composed of multiple nodules of varying sizes in the spleen [2,3]; only one case of focal angioma has been described [4]. Our patient was a 09-year-old male who, while being evaluated for abdominal pain, was found to have splenomegaly with dilated splenic vein with partial thrombus. The patient didn't show any other signs or symptoms of hypersplenism. Colour Doppler of portal venous system performed, which revealed 10 mm dilation of splenic vein and thrombus in proximal part. Patient had a history of extra hepatic portal vein thrombosis. Because of the concern for hypersplenism, splenectomy was performed. Pathology revealed the presence of smooth outer surface with multiple brownish area identified on cut surface, the largest being 4.5 cm. This lesion extended up to the capsule but did not penetrate it. An immunohistochemical study confirmed the histologic diagnosis of littoral cell angioma by showing the cells positive for CD31, CD68, and factor VIII.

RADIOLOGIC FEATURES

On ultrasonography, littoral cell angioma appears as an enlargement of spleen (16cm) in size and normal echotexture. Portal vein at porta hepatis is replaced by multiple collaterals suggestive of portal cavernoma formation. Peripancreatic, perigastric, splenic hilar collaterals are noted. CBD normal in caliber. Minimal interbowel fluid is noted, with no focal lesion. On colour Doppler study liver is normal in size with mild diffuse coarse echotexture. Few periportal collaterals are noted. Radiology shows moderate gross enlargement (16 cm) & normal in echotexture with no focal lesion. Splenic vein 10mm in caliber with partial thrombus formation. 20x10 mm sized splenic nodule is noted at splenic hilum. Free fluid was found in peritoneal cavity. Radiologic findings can rarely lead to a definitive diagnosis because many other splenic neoplasms have a similar appearance. Other splenic neoplasms that mimic littoral cell angioma

include hemangiomatosis, lymphangiomatosis, hamartoma, hemangiopericytoma, hemangioendothelioma, and angiosarcoma; malignant processes such as metastases, lymphoma, and Kaposi sarcoma; and infectious processes such as Pneumocystis and Mycobacterium [5]. Splenic metastases from malignant melanoma and leiomyosarcoma, although rare, have been reported. Splenic metastasis from melanoma has an incidence of less than 5%, but the rate is much higher on autopsy (36%) [6]. Sonography is rarely helpful because the findings vary widely. The sonographic features that can be seen include heterogeneous echotexture without any definite nodules, or the lesions may appear isoechoic, hypoechoic, or hyperechoic [2]. MRI of the spleen may further help in the diagnosis by showing hypointense lesions on both T1-weighted and T2-weighted scans because of the hemosiderin content of the tumor.

PATHOLOGIC FEATURES

Littoral cell angioma is a vascular tumor unique to the spleen, originally thought to arise from normal splenic lining (littoral) cells. It varies in size from minute foci to large nodules almost completely replacing the splenic tissue [7]. Definite diagnosis of littoral cell angioma can only be made by histopathology after splenectomy, which is usually performed because of the concern for malignancy or idiopathic hypersplenism. The gross cut surface of the spleen was greyish with a variable colour from dark red or brown to black, depending on the chronicity of the blood in the lesion.

Histopathologically, littoral cell angioma is characterized by the presence of anastomosing vascular channels, which are lined by tall endothelial cells and papillary fronds extending into the vascular channels [1-3]. Some exfoliated cells may also be found within the vascular spaces. The lesions do not have a capsule but are well delineated from the surrounding splenic parenchyma. Littoral cell angioma is a benign vascular tumor of the spleen. It shares some morphologic and immunohistochemical features with hemangiomas of other sites such as immunoreactivity for vascular endothelial markers CD31 and factor VIII. However, it is unique in that almost all cases also express histiocytic marker CD68. It has been proposed, based on the expression of CD68 by the tumor cells, that this tumor may have its origin in the splenic sinus lining cells or littoral cells [1]. In conclusion, littoral cell angioma, although a pathologic diagnosis, should be considered in the differential diagnosis in the presence of multiple hypoattenuating nodules in the spleen, particularly in the presence of symptoms of hypersplenism.

Figure 1 :-Littoral cell angioma of splenic tissue at 4x scanner view.

Figure 2:- Littoral cell angioma of splenic tissue at 10x view.

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