Case Report

Rare and Unusual Occurrence of Splenic Hamartoma on Ultrasonography

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Abstract

Vascular tumors constitute the most common primary tumors of the spleen. Splenic hamartoma and littoral cell angioma have been reported only in the spleen. Splenic hamartomas are rare benign vascular tumors which are incidentally detected during imaging and are seldom symptomatic. This case report describes a rare sonological appearance of splenic hamartoma in a 31-year-old female with occasional pain in the left hypochondrium.

Keywords: Histopathology, immunohistochemistry, splenic hamartoma, ultrasonography

Introduction

Splenic hamartomas are rare benign vascular tumors that are often incidentally detected. Splenic hamartomas are frequently diagnosed on ultrasonography imaging which has higher sensitivity at detection as compared to computed tomography (CT) where the lesions appear isodense to the adjacent splenic parenchyma.[1] Few instances of splenic hamartoma are associated with anemia, thrombocytopenia, and pancytopenia related to hypersplenism.[2] Syndromic association of splenic hamartoma with tuberous sclerosis and Wiskott–Aldrich-like syndrome has been reported.[3]

Case Report

A 31-year-old female presented to the department of general surgery with complaints of intermittent dull pain in the left hypochondrium for 2 months duration. There was no significant past history related to renal colic and physical examination revealed no left renal angle tenderness. There was mild splenomegaly. Blood investigations revealed thrombocytopenia consistent with features of hypersplenism. The patient was referred for ultrasonography which revealed evidence of a well-defined iso- and hypoechoic mass lesion measuring 4 cm × 4 cm involving the superior pole of the spleen [Figure 1a]. The lesion demonstrated central vascularity on color Doppler [Figure 1b]. Differential diagnoses included atypical hemangioma, splenic hamartoma, and lymphoma. Fine-needle aspiration cytology revealed bizarre-shaped large stromal cells that were scattered throughout the mass lesion which showed irregular morphology of nuclei, such as reniform, oval, convoluted, or multilobulated with scarcely visible cytoplasm. Mitotic activity, necrosis, or infiltrative growth pattern were not identified [Figure 2]. The patient subsequently underwent splenectomy in view of the increasing intensity of pain in the left hypochondrium. The resected spleen specimen weighed 300 g with the evidence of a well-circumscribed solid mass lesion at the superior pole measuring 4 cm × 4 cm. Histopathology of the resected specimen revealed a mixture of sinusoid-like vascular channels lined by plump endothelial cells which stained positive for CD8 on immunohistochemistry. There were no areas of mitotic figures, necrosis, or nuclear atypia. A final diagnosis of splenic hamartoma is made based on the findings of histopathology and immunohistochemistry.

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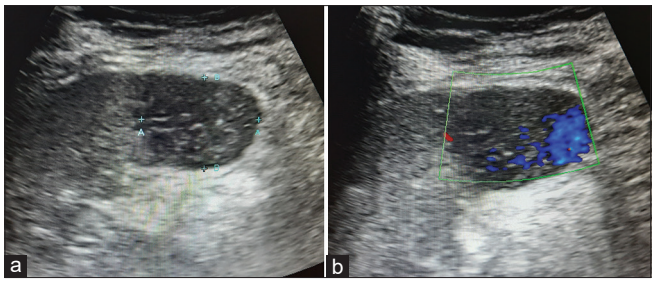

Figure 1: (a) Longitudinal ultrasonography image demonstrating a well-defined iso- and hypoechoic mass lesion involving the superior pole of the spleen. (b) Longitudinal ultrasonography image demonstrating intense color uptake on Doppler study consistent with rich vascularity of the splenic lesion

Discussion

Splenic hamartomas are benign and often asymptomatic lesions which may occur in any age group, with equal sex preponderance.[4] The larger size of splenic hamartomas in females is attributed to hormonal influence with splenomegaly and complications like spontaneous rupture of the spleen more often encountered than in males.[5] Features of hypersplenism such as anemia, thrombocytopenia, and pancytopenia have been reported more frequently in children with splenic hamartoma than in adults.[6] Rare syndromic association of splenic hamartoma with tuberous sclerosis and Wiskott–Aldrich-like syndrome has also been reported.[3]

Hemangioma is the most common benign primary vascular tumor of the spleen followed by littoral cell angioma and splenic hamartoma.[7] Angiosarcoma and Kaposi's sarcoma constitute the most common primary malignant tumors arising from the spleen.[8]

On ultrasonography, splenic hamartomas have an appearance of an iso- and hyperechoic mass showing increased vascularity on color Doppler. On CT, splenic hamartomas appear isodense causing contour deformity showing heterogeneous contrast enhancement. Splenic hamartomas appear isointense to the adjacent splenic parenchyma on T1-weighted sequences of magnetic resonance imaging and heterogeneously hyperintense on T2-weighted sequences.

Vascular tumors of the spleen such as hemangioma, hamartoma, littoral cell angioma, angiosarcoma, lymphangioma, hemangioendothelioma, lymphoma, sclerosing angiomatoid nodular transformation of the spleen, metastases from melanoma, bronchogenic and ovarian carcinoma, and granulomatous lesions like sarcoidosis must be included in the differential diagnosis of solid mass lesions of the spleen.[9] Solid lesions of the spleen usually are asymptomatic and detected incidentally on imaging. However, larger lesions carry a risk of spontaneous rupture of the spleen.

Splenic hamartoma occurs as solitary or multiple, well-circumscribed lesions ranging between <1 cm and 20 cm in size with no surrounding capsule and causing compression of the normal splenic parenchyma. On histopathology, splenic hamartoma comprises sinusoid-like vascular channels with absent intervening white pulp. On immunohistochemistry, the key feature of splenic hamartoma is CD8 positivity of the plump endothelial cells lining the vascular channels.[10]

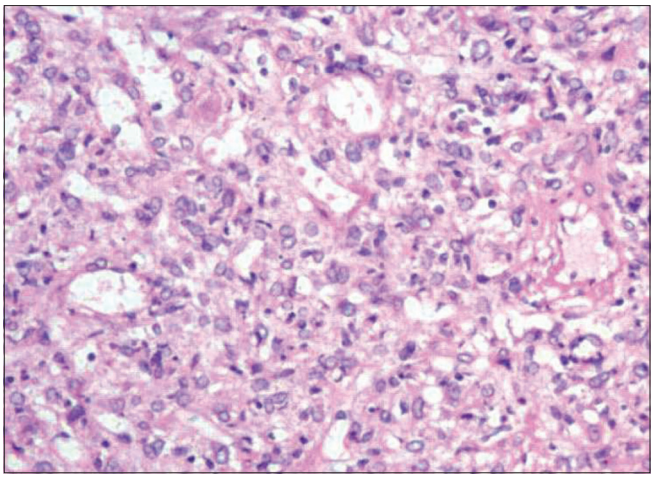

Figure 2: Histopathology image of the resected spleen specimen demonstrating disorganized blood vessels intermingled with splenic red pulp element consistent with features of splenic hamartoma (H and E, ×400)

Despite the fact that the final diagnosis of splenic hamartomas is established by a histopathological evaluation, this article highlights the importance of preoperative imaging evaluation on ultrasonography. Splenic hamartomas are solid, well-defined lesions with homogeneous echogenicity compared to the normal splenic parenchyma on ultrasonography, but may appear heterogeneous and may demonstrate cystic changes. Furthermore, splenic hamartomas should be differentiated from the more common and ominous lesions of the spleen which include hemangioma, angiosarcoma, and metastases.[11] Nevertheless, there is considerable overlap in the imaging patterns of vascular lesions of the spleen, and histopathological evaluation remains the only definite means of diagnosing splenic hamartomas.

Conclusion

Splenic hamartoma is a rare benign vascular tumor of the spleen most often detected incidentally. Interpretation of imaging findings with histopathological and immunohistochemistry correlation can help differentiate this benign entity from malignant lesions of the spleen. Splenic hamartoma should be included in the imaging differential diagnosis of focal splenic masses.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other

clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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