Sclerosing angiomatoid nodular transformation (SANT): A rare entity of spleen

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Abstract

Sclerosing angiomatoid nodular transformation (SANT) is benign neoplasm originating from vascular cells of the spleen. These tumors are rare between splenic lesions. In this paper, we present a SANT that arose in the upper pole of the spleen of a 61-years-old female. During the examination for anemia and diabetes mellitus, an splenic mass was found incidentally at the CT imaging. Open laparotomy with splenectomy was performed. Histopathological features of this tumor confirmed a rich vascular network in the nodule surrounded by hyalin cortex. In this report, we discuss the differential diagnosis of SANT.

Introduction

Sclerosing angiomatoid nodular transformation (SANT) is an uncommon disorder characterized by benign proliferation of vascular tissue in the splenic red pulpa and was first described by Martel, et al. in 2004 [1]. In the report, they were described 25 cases with angiomatoid nodules surrounded by dense fibrous tissue. About since than 132 cases of SANT have been reported in the English literature [2]. SANT usually presents as a splenic tumoral mass, but it rarely manifests clinically symptoms.

A retrospective review of the surgical database was performed to identified SANT at our institution from January 2010 to January 2016. We found only one SANT of spleen between 117 splenectomy specimens. In this paper, a patient who attended hospital with anemia and diabetes mellitus and by incidentally were found to have SANT on splenectomy specimen is presented.

Case report

A 61-year-old female with a history of chronic pulmonary symptoms and anemia was admitted to Department of Internal Medicine in our hospital for further investigation. A physical examination revealed the patient to be afebrile and no abnormalities other than the vaguely tenderness in the left upper quadrant. She had been suffering from diabetic retinopathy for 3 years. Laboratory examinations revealed hemoglobin 10.6 g/dl, hematocrite 35%, BCR-ABL transcript(-) and JAK-2 (-), while the chest X-ray findings and liver function were normal. Ultrasound of the abdomen was inconclusive. CT imaging showed a 3 cm tumoral mass at the upper pole of spleen (Figure 1). The patient was admitted to our Surgery Department. The tumor was suspected to be a splenic lymphoma or another type of malignant lymphoma. A laparotomy was performed, which revealed a tumoral mass on the spleen. The patient underwent a splenectomy. The postoperative course was uneventful.

The splenic tumor was visible and yellowish-white noduler, irregular tumoral mass in the upper pole (Figure 2). The resected spleen weighed 204 gm and the tumor size was 2.5x2.8x2.5cm. Histologically the mass showed hyalin capsule formation around the noduler lesion with increased vascularity. Immunohistologic studies showed, CD 34(+) cells in capillaries, CD31(+) cells in capillaries, sinusoids and small venules, Factor 8(+), vimentin(+), smooth muscle actin(+), S100(+), Desmin(-) cells in capillaries and sinusoids, and usual staining cells with CD3, CD15, CD20, CD30, CD38, PAX5 and Ki67. The final pathological diagnosis was SANT of the spleen.

Discussion

Primary splenic vascular neoplasms arise from the red pulp of the spleen. Previously, these lesions were called as hamartomas, hemangioendoteliomas, sclerosed hamangiomas capiller hemangioma,
multinoduler hemangioma, and inflammatory pseudotumors. Currently, they classified as sclerosing angiomatoid nodular transformation (SANT) of spleen. Although clinical significance of SANT is unclear, they are important lesions because they may be confused with malignant tumors [3]. Furthermore, some papers reported that SANT showed a relationship with systemic inflammatory reaction, Epstein-Barr virus infection, trauma and splenic hematomas. Our case showed no evidence of primary etiologic reason. The majority of SANT is asymptomatic and discovered incidentally during imaging studies as in our case, or they can present with symptoms related to tumor compression of adjacent organs. Patients presenting with symptoms generally complain of abdominal pain, abdominal distension or a palpable mass [4]. Generally, age of onset is in the fifth decade, but it might observed from second to seventh decade.

On sonography SANT appears as a hypoechoic mass with linear echoes that is suspicious for malignancy. CT is the best imaging technique for evaluating tumor extent. It seems that the presence of the radiating septations in the nodule, spoke-wheel sign, might be diagnostic for SANT. Despite the various diagnostic modalities, SANT remains difficult to distinguish preoperatively from a tumor. Some authors reported that fine needle biopsy of the lesion is a good method for diagnosis but this method carries the risk of dissemination of a malign neoplasm.

Histologically SANT of the spleen are composed by angiomatoid nodules with infiltrative stromal process. Immunohistochemistry is very important for the diagnostic confirmation. SANT have been categorized three types. Type 1 contains well formed capillaries with CD34(+), CD8(-), CD31(+), Type 2 includes vessels similar splenic sinusoids with pattern CD34(-), CD8(+), CD31(+). Type 3 covers mesh-like patterns of small veins with CD34(-), CD8(-), CD31(+). [3]

In our case, histologically, the appearances of angimatoid nodules and vascular staining with CD34 and C31 benefited differentiation from hemangiomas, Littoral cell angioma, hamartoma and angiosarcomas. SANT is still poorly understood, and its diagnosis represents for the lack of specific diagnostic methods as well as biochemical and biological markers. Making a correct diagnosis was very difficult before the operation. Because SANT may be confused with malignant lesions, recommended treatment is splenectomy.

References
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