Research Paper

CT and MRI findings and pathologic correlation on Sclerosing angiomatoid nodular transformation of the spleen

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ABSTRACT

The purpose of this study was to evaluate the CT and MRI characteristics of SANT with pathologic correlation. Six patients who confirmed SANT were included and their clinical history, histopathology, immunohistochemistry and imaging findings were analyzed. Pathologic features of SANT revealed well-circumscribed mass with multiple dark brown nodules and a central fibrous scar in the nodular. Immunohistochemical reports showed a unique characterized by CD34, CD31 and CD8. They showed low density and hypo intensity on unenhanced CT and MRI and progressive enhancement with a radiating line pattern after administration. SANT showed a unique characteristic imaging features on CT and MRI, which had correlation with pathology.

Key words: Sclerosing angiomatoid nodular transformation, CT, MRI, pathology.

INTRODUCTION

The spleen may be affected during several disease processes, and lymphomas particularly. Vascular neoplasms are the most common primary non-lymphomatoid tumors of the spleen. They are comprised of hemangiommas, littoral cell angiommas, splenic hamartomas, angiosarcomas and Sclerosing Angiomatoid Nodular Transformation (SANT)(Elsayes et al., 2005; Zeeb et al., 2009). Before it was defined definitely, SANT of the spleen was referred to by numerous pathologic terms, including cord capillary hemangioma, multinodular hemangioma, sclerosed hemangiommas and inflammatory pseudotumors (Diebold et al., 2008). SANT of the spleen was first defined by Martel et al (2004). It is a rare benign lesion of the spleen and is altered red pulp entrapped by a non-neoplastic stromal proliferation.

SANT of the spleen is populated by endothelial cells, phenotypically recapitulating normal splenic vasculature and small veins. Therefore, SANT can be distinguished on the basis of their special immunohistochemical profiles, because of three distinct types of blood vessels. These include well-formed cord capillaries (CD34+CD8-CD31+), splenic sinusoids (CD34-CD8+CD31+) and small veins (CD34-CD8-CD31+)(Onder et al, 2012; Subhawong et al, 2010; Gutzeit et al, 2009).

To the best of our knowledge, there have been few reports to describe SANT of the spleen, especially on images (Chikkappa et al, 2009; Raman et al, 2013; Falk et al, 2012). The purpose of our study was to find some characteristics on computed tomography (CT) and magnetic resonance imaging (MRI) features on SANT and to help radiologist to diagnose the disease.

MATERIALS AND METHODS

Patients

This retrospective study was approved by our institutional
review board. Between January 2007 and April 2014, nine patients were included with a diagnosis of SANT by pathology. Three of them were excluded from the study because of having no imaging data. Therefore, the final study group consisted of six patients: 4 women and 2 men, with an average of 52.5 years (range, 41–68 years). Five of the patients were found by physical examination, only one of them felt a little abdominal pain. All patients underwent splenectomy.

Imaging methods and analysis

Two patients underwent abdominal CT scans, which were performed using a 64-slice multi detector computed tomography (MDCT) scanner (LightSpeed VCT, GE Healthcare, Waukesha, Wis). The parameters for the MDCT imaging were 120 kV, 200–300 mA, 5 mm thickness and 1.25 mm collimation, a reconstruction thickness of 5 mm. Contrast-enhanced abdominal CT scans were obtained after administration of 90 ml of the iodinated contrast agent iopromide (Omnipaque 300; Schering, Berlin, Germany) at a rate of 3.0 ml/s. The two patients had dual phases, the delay time was 30, 80 s.

Four patients underwent MRI examination, which were performed with a 1.5-T superconducting magnet (Avanto; Siemens Medical solutions, Erlangen, Germany) 6-equipped with phased-array coils. The scans sequences includes: precontrast T1-weighted sequence (TR: 112ms; TE: 4.8ms); T2-weighted sequence (TR: 3500ms; TE: 84ms); dynamic fat-saturated gadolinium-enhanced T1-weighted sequences. Two patients underwent in-/out-of-the-phase sequences before injection.

Two radiologists with more than 10 years of experience in abdominal MRI retrospectively analyzed the images. The radiologists evaluated the CT and MRI images on the presence and number of lesions within the spleen. The shape and margins of each lesion were documented. The attenuation or signal intensity of the lesion was compared with the surrounding normal spleen on unenhanced CT and MRI images. Besides that, enhanced pattern of each lesion was compared with the surrounding normal spleen in each phase, such as the enhancement was homogenous or heterogeneous. The following patterns of enhancement were also documented: enhancement of peripheral radiating lines, rim enhancement, the present and enhancement of a central scar.

Pathology

Two pathologists with more than 15 years of experience in hematopathology reviewed the all patients. The pathology and surgical records were reviewed for the number and sizes of the lesion. Histopathology reports and slides were also observed for all cases. The lesions were used hematoxylin-eosin (HE) stained and the diagnosis of SANT on the basis of criteria originated by Martel et al (2004). Immunohistochemistry reports were available for all patients. These were assessed for immnoreactivity to the vascular markers (CD8, CD31 and CD34). One observer reviewed the histologic pathology findings and correlated them with the imaging features.

RESULTS

Patients’ features

Finally, six patients were included in the study.

Imaging features

On CT scans, the two lesions were solitary, round and lobulated margin, and the margins were clear. On unenhanced CT images, both lesions were homogenous and slightly hypointenuating. After contrast injection, the lesions showed heterogeneous and slightly enhancing on arterial phase. Besides that, rim enhancement could be seen by the two lesions. On the portal venous phase, the two lesions showed progressive enhancement, and radiating lines could be observed in the lesions (Figures 1a, 1b and 1c). Both on the arterial phase and on the portal venous phase, the two lesions showed hypointenuating relative to the surrounding normal spleen.

Four patients underwent MRI examination and all of them were solitary and round. On T1-weighted image, two lesions showed isointensity mainly and the other two lesions showed hypointensity. On T2-weighted images, all of the four lesions showed heterogeneous, predominantly hypointensity and radiating lines, just like scars or spoken wheel pattern, could be observed on all lesions. There was no intensity decrease with chemical shift image. On gadolinium-enhanced images, all lesions were heterogeneous enhancing during the arterial phase. And on the portal venous phase and the delay phase, all lesions showed progressive enhancement. However, the radiating lines, which revealed on T2-weighted image, still showed hypointensity, without enhancement. The mainly lesions showed isoointensity on the delay phase compared with the surrounding spleen (Figures 2a, 2b, 2c and 2d).

Pathological features

All patients underwent splenectomy. A solitary splenic tumor was found in all patients, with an average size of 6.5 cm (range: 4.5–8 cm). On histologic examination, all lesions showed a well-circumscribed but non-encapsulated bosselated mass with multiple dark brown nodules, interspersed with a fibrous stroma.
Figure 1. 46-year-old man with sclerosing angiomatoid nodular transformation. Axial (a) unenhanced CT scan showed a hypotenuating lesion with round margin in the spleen (arrow). Axial (b) arterial phase contrast-enhanced axial CT image showed round lesion with rim enhancement (arrow). Axial (c) portal venous phase contrast-enhanced axial CT image showed progressive enhancement just like radiating lines (arrow).

Figure 2. Forty one (41) year-old woman with Sclerosing angiomatoid nodular transformation. Axial (a) T1-weighted MR image showed hypointensity on the lesion with round margin (arrow). Axial (b) T1-weighted MR image showed slightly rim enhancement in arterial phase (arrow). Axial (c) T1-weighted MR image showed progressive enhancement like ‘spoke wheel’ on portal venous phase (arrow). Axial (d) T2-weighted image showed heterogeneous hypointensity lesion with hypointense central scar (arrow).
Microscopically, SANT composed of slit like, round, or irregularly shaped vascular spaces often surrounded by dense collagen fibrosis or fibroid rims. Centrally, the lesions were populated by vascular channels of varying caliber lined with plump endothelium interspersed with spindle cells (Figure 3). Necrosis, significant nuclear atypia and calcification were not present.

All of the six lesions on immunohistochemistry reports revealed CD34-positive, CD31-positive and CD8-negative cells in the peripheral sector, while the central portion was mainly comprised of CD31-positive, CD34-and CD8-negative cells. CD68 showed partly positive and immunostained histocytes within the splenic cord and scattered sinusoidal macrophages.

DISCUSSION

SANT is a recently described lesion within the spleen consisting of vascular nodules in a fibrous stroma. It was first defined by Martel et al. (2004). Martel et al. (2004) and Falk et al. (2012) also noted that the pathogenesis of this entity was unclear and hypothesized that SANT might be splenic hematoma that had undergone an unusual form of sclerosis, with a particular reactionary transformation of red pulp due to an exaggerated stromal response (Falk et al., 2012; Cao et al., 2015). It is mostly found by accident, and most patients feel asymptomatic (Kim et al., 2011). In our study, lesion was found in five of the six patients by physical examination, and only one patient felt a little abdominal pain. SANT appears to be a benign lesion. The all patients in our study had no recurrence after splenectomy. Also, no reports said that SANT had malignant behavior (Sitaraman et al., 2010; Awamleh and Perez-Ordonez, 2013).

Previous papers reported that SANT is considered to be a disease of female preponderance (about 1.3:1 female-to-male ratio). And the patients usually present in the 30 to 60 year old group (Abbott et al., 2004; Lee et al., 2007). The size of reported lesions ranges from 3 to 17 cm in diameter. In our study, male occurrence rate was higher than female occurrence rate. The reason might be relatively small samples. However, the year period and the size range were similar with the papers having reported.

Gross examination typically reveals a mass of red-brown nodules embedded in dense fibrous stroma that form a central scar. The histological hallmark of SANT is the angiomatoid appearance of these nodules, which contain multiple types of blood vessels normally found in splenic red pulp; cord capillaries, splenic sinusoids and small veins are present and can be identified by their various staining patterns (Hou et al., 2010; Giovagnoni et al., 2005). Other splenic primary vascular lesions, such as littoral cell angioma, conventional hemangioma and hemangioendothelioma, lack the nodular pattern and staining pattern of SANT. In our study, all the six lesions were found in the central scar by pathology and the special staining pattern (CD34, CD31, CD8) were correspond to the studies that have already been reported.

The imaging features on SANT have only been reported in the radiology literature in a small number of case reports, and a series of them were about sonographic findings (Wang et al., 2012; Koreishi et al., 2009; El Demellawy et al., 2009). To the best of our knowledge, symmetrical imaging findings on SANT were limited. Our study showed several imaging characters with pathologic correlation. On CT images, the lesion demonstrated diffuse peripheral enhancement, just like a rim, on the arterial phase on the both cases. And on the portal venous phase, the lesions showed progressive centripetal filling in a radiating pattern, while the center remained hypodense. After gadolinium-enhanced images, persistently peripheral enhancing radiating lines could be observed on the arterial phase, the portal venous phase and the delay phase. And this appearance is referred to 'spoken wheel', just as
Karaosmanogh et al. (2009) and Gutzeit et al. (2008) proposed. These CT and MRI imaging findings corresponded on histopathology to the concentration of angiomatos nodules around the periphery of the lesion, also in a radiating pattern between the branches of the fibrous scar. Besides that, all the six cases showed progressive pattern of enhancement. This symptom might be attributed to fibrous tissue and the angiomatos tissue in the nodule, which had a character of delayed enhancement.

On unenhanced CT scan, only hypodensity lesion could be seen compared with the surrounding spleen. We could not find out more detailed structure. However, low signal intensity on T2-weighted images was present in all the four lesions that underwent MRI examination. The lesions appeared as hyperintensity seen at the periphery and a centrally hypointense region on T2-weighted images. And the hypointensity area was showed as radiating bands corresponding with areas of fibrous on pathology. Therefore, MRI-enhanced examination might provide more information for us to diagnose SANT of the spleen.

The differential diagnosis of a splenic mass is broad and composes of many benign and malignant entities and sometimes it’s very difficult for us to distinguish them from each other. Hemangiomas are the most common benign lesion in the spleen and they can also show progressive enhancement (Abbott et al., 2004). They may be distinguished from SANT by their high T2 signal intensity. Littoral cell angiom is a rare vascular neoplasm of the spleen, and it may be differentiated due to its typical multiple hypodense nodules and considering the solitary appearance of SANT of the spleen. Lymphoma is the most common malignant tumor of the spleen and can present as a solitary or mass or multiple nodules, but it shows little enhancement and it has the appearance of splenomegaly and intraabdominal lymphadenopathy (Warshauer and Hall, 2006). These features may be helpful clues for making the differential diagnosis. Angiosarcoma is the most prevalent primary malignant vascular entity and it may sometimes be difficult to differentiate from other benign lesions (Awamleh and Perez-Ordonez, 2007). However, it has highly aggressive biological nature and it always has happened distant metastasis when it grows as the same size as the SANT.

There are also several limitations in our study. Firstly, SANT of the spleen is a rare benign lesion of the spleen, so the samples are relatively small. Secondly, the number of the patients underwent CT scanning is a few and the observing result may have deviation.

Conclusion

SANT is a rare benign lesion of the spleen. It is classically considered as a female-predominant disease and is considered the incidence may increase, after its recent description as a separate clinic pathologic entity. Histopathologically, these lesions reveal multiple confluent angiomatos nodules and it has an unique immunohistochemical profile characterized by CD34-CD31+CD8+ sinusoids, CD34+CD31+CD8- capillaries and CD34-CD31+CD8- small veins. On CT and MRI imagings, SANT displays its special features, including peripheral enhancing radiating lines and progressive on arterial, portal venous phase and delay phase. Peripheral enhancing radiating lines, called spoken wheel pattern, corresponding histologically to multiple angiomatos nodules concentrated around the periphery and in a radiating. Besides that, SANT showed hypointensity on T2-weighted images and it is also a very important sign. We believe that remembering these imaging features may help us distinguish SANT from other solid splenic lesions.

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