

## An unusual case of sclerosing angiomatoid nodular transformation: radiological and histopathological analyses

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Received: 02.05.2013 • Accepted: 03.09.2013 • Published Online: 31.03.2014 • Printed: 30.04.2014

**Abstract:** Sclerosing angiomatoid nodular transformation (SANT) is a rare benign primary vascular lesion of the spleen. Its etiology is still debated. Radiological characteristics are less known, although there are some reports regarding histopathological features of SANT. A 21-year-old male patient was admitted to our hospital with fatigue, weight loss, and abdominal pain for 4 months. Physical examination, complete blood count, and biochemical parameters were unremarkable. Dynamic contrast-enhanced computed tomography (CT) of the abdomen was performed. A heterogeneous well-defined hyperdense nodular lesion 3 cm in diameter was detected during the arterial phase. The detected lesion was seen as isodense with the spleen parenchyma during the portal venous and late venous phases. Magnetic resonance imaging (MRI) showed an isointense-hypointense nodular lesion on T1- and T2-weighted images. Intraabdominal LAM or splenomegaly was not detected. Microscopically, it was composed of angiomatoid nodules separated by central stellate fibrous stroma and fibrous septa. The contrast enhancement pattern was described as centrally hypovascular, radially progressive centripetal vascular contrast enhancement, called a spoke-wheel pattern in previously reported cases. We present CT and MRI findings and their correlation with histopathological findings of a case of unusual symptomatic SANT.

**Key words:** Computed tomography, magnetic resonance imaging, sclerosing angiomatoid nodular transformation, spleen

### 1. Introduction

Splenic pathologies are generally seen as secondary involvement by disorders of other organs/systems or caused by infectious spread (1–3). Primary lesions occur rarely (4,5). The overwhelming majority of primary lesions are vascular. Hemangiomas are the most common. Less encountered lesions are lymphangioma, littoral cell angioma, hemangioendothelioma, and hamartoma (6). Malignant vascular lesions might also be seen, as well as benign lesions. Angiosarcoma is a malignant vascular lesion, usually presenting with distant metastasis.

Sclerosing angiomatoid nodular transformation (SANT) is a rare benign primary vascular lesion of the spleen. It is composed of red pulp nonhematolymphoid cells (7). The exact etiology is still unknown (7). Patients are generally asymptomatic. SANT lesions are generally detected incidentally by abdominal imaging, as with most of the other splenic lesions, such as by calcification (7,8). In the literature, reports defining the radiological characteristics of SANT are very rare. The majority of these reports have defined the histopathological features.

We present computed tomography (CT) and magnetic resonance imaging (MRI) findings and their correlations

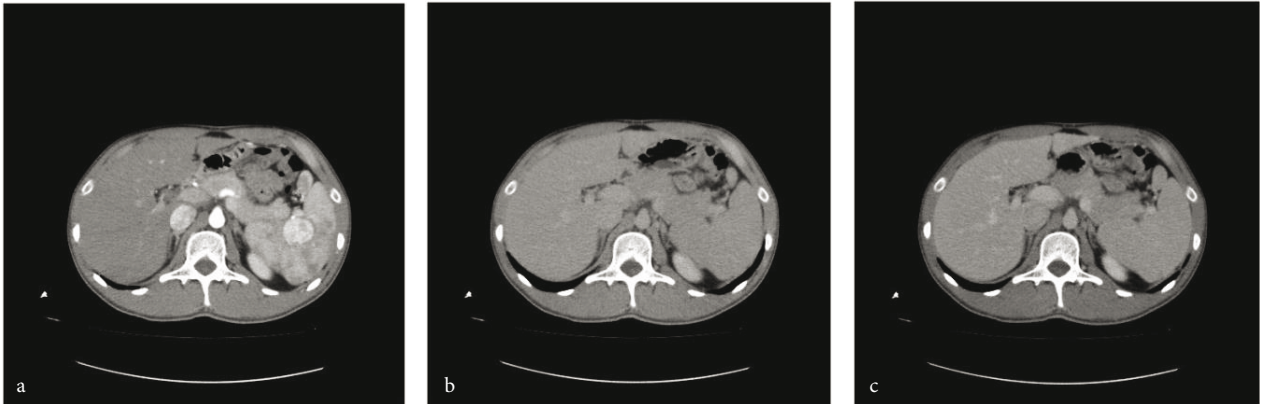
with histopathological findings of a case of unusual symptomatic SANT.

### 2. Case report

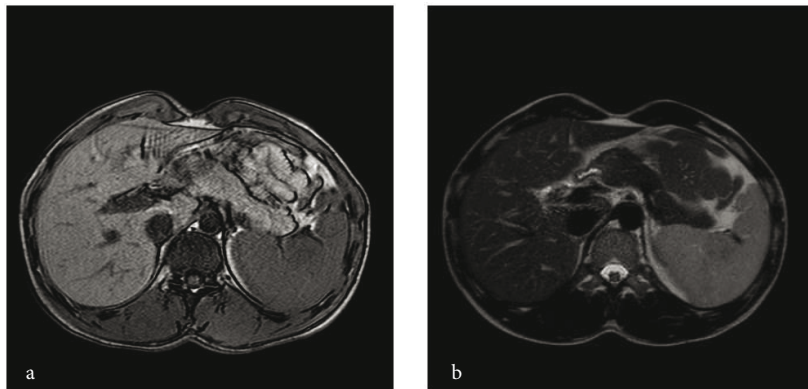
A 21-year-old male patient was admitted to our hospital with fatigue, weight loss, and abdominal pain for 4 months. Physical examination, past medical history, complete blood count, and biochemical parameters were unremarkable.

Dynamic contrast-enhanced CT of the abdomen was performed. A heterogeneous well-defined hyperdense nodular lesion 3 cm in diameter was detected during the arterial phase. The detected lesion was seen as isodense with the spleen parenchyma during the portal venous and late venous phases (Figures 1a–1c). MRI showed an isointense-hypointense nodular lesion on T1- and T2-weighted images (Figures 2a and 2b). After intravenous gadolinium administration, the lesion appeared hyperattenuated in the arterial, portal venous, and late venous phases (Figures 3a–3c). Intraabdominal lymphangioleiomyomatosis or splenomegaly was not detected. Percutaneous needle biopsy taken from the biggest lesion (3 cm in diameter) was inadequate for the diagnosis. Therefore, surgery was planned.

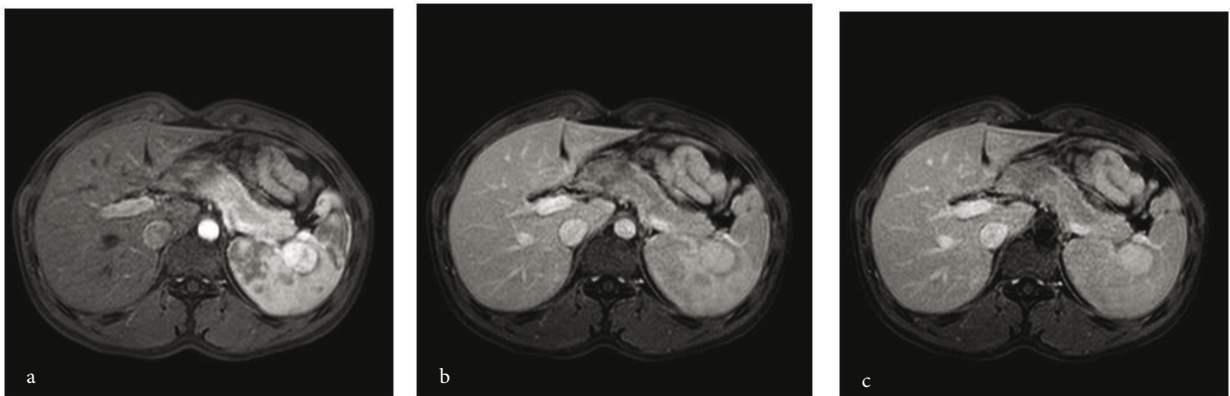
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**Figure 1.** A well-defined nodular lesion seen as hyperdense in the arterial (a) and as isodense in the portal venous (b) and the late venous (c) phases at the hilum of the spleen.



**Figure 2.** Splenic lesion seen as iso-hypointense on T1-weighted (a) and T2-weighted (b) images.



**Figure 3.** Hyperattenuating splenic lesion seen as hyperdense in the arterial (a) and as isodense in the portal venous (b) and the late venous (c) phases at the hilum of the spleen.

A splenectomy was performed, and gross examination demonstrated a mildly enlarged spleen with parameters including dimensions of  $11 \times 8 \times 7$  cm, weight of 200 g, and a well-demarcated nodular lesion  $3 \times 2.5 \times 2.3$  cm in size at the hilum, differentiated from the surrounding parenchyma with indistinct color and peripherally located

millimetric creamy white nodules. Microscopically, multiple microangiomaoid nodules in red pulp were detected. Vascular spaces were lined with endothelial cells surrounded by epithelioid proliferation cells with ovoid nucleus and insignificant cytoplasmic border. A sclerotic fibrous component was not prominent.

Immunohistochemical analysis of angiomatoid nodules showed CD34+, CD31+, CD8-, and HHV8-cells. Residual lymphoid cells were CD3- and CD20-. Ki-67 proliferation rate was approximately 4%-5%. Histopathological and immunohistochemical analyses were compatible with SANT.

### 3. Discussion

The vast majority of the primary splenic lesions are vascular (4,5). Hemangiomas are the most common of this group. Less commonly encountered lesions are lymphangioma, littoral cell angioma, hemangioendothelioma, and hamartoma (6). Hemangioma presents as a well-defined isodense-hypodense lesion. Cavernous types enhance heterogeneously, whereas the capillary types enhance homogeneously. Unlike the hemangiomas showing nodular enhancement in the liver, splenic hemangiomas show circular enhancement. Lymphangiomas are multiloculated cystic lesions and demonstrate MRI signals consistent with the proteinaceous content (9).

Hamartomas demonstrate homogeneous solid echogenic masses on ultrasonography and increased blood flow on color-Doppler images. CT and MRI are the other methods used for diagnosis. It is difficult to differentiate from the malignant types using imaging methods. Therefore, the exact diagnosis can only be established by histopathological findings (10).

Angiosarcoma, a malignant primary splenic lesion, presents as a heterogeneous cystic-solid lesion on ultrasonography images. Solid components show increased blood flow on color-Doppler images. They present as ill-defined lesions with punctuated or massive calcifications in a radial pattern. They can be accompanied by a hemorrhage, demonstrating hyperdensity on CT images. They show heterogeneous enhancement on postcontrast images. MRI is a useful imaging method for demonstrating the hemorrhage and necrotic areas in these lesions (11).

Histopathological properties of SANT were first defined by Martel et al. in a case series of 25 patients (7). They defined SANT as a benign, mostly asymptomatic, primary vascular lesion of the spleen. Some patients may have abdominal pain. They are generally detected by abdominal imaging incidentally. The mean age of the patient population was 53.7 years (age range: 22-74 years). The female-to-male ratio was 2:1 (7). In the literature, there are some reports about SANT; most of them have defined the histopathological features. Radiological description of SANT is limited to a few cases. CT findings were first defined by Li et al. They described SANT as a hypodense lesion in the portal phase and isodense

with central calcification in the late venous phase (12). Kim et al. retrospectively evaluated 7 patients with histopathologically correlated SANT using CT, MRI, ultrasonography, and positron emission tomography (PET) CT. They concluded that SANT was a single, well-defined lesion showing heterogeneous enhancement on CT, centripetal progressive enhancement on MRI, and increased fludeoxyglucose 18F (FDG) activity on PET. Follow-up CT scans were applied to 5 patients and tumor growth was found (13). SANT was defined by Lee et al. and Zeeb et al. as a peripherally contrast-enhanced hypodense lesion in the portal venous phase, shown as an isodense lesion similar to the surrounding parenchyma except for the central stellate area in the late venous phase (14,15). The enhancement pattern of SANT is described as centrally hypovascular, radially progressive centripetal vascular contrast enhancement, called the spoke-wheel pattern. The spoke-wheel pattern occurs secondary to angiomatoid nodules separated by central stellate fibrous stroma and fibrous septa (4). Karaosmanoglu et al. described both CT and MRI findings of SANT. The lesion showed the spoke-wheel pattern in both CT and MRI. SANT appeared centrally hyperintense on T1-weighted and hypointense on T2-weighted images because of the hemorrhagic component. Hyperintense radiations from the periphery to the central area on T2-weighted images were shown as radially progressive centripetal vascular contrast enhancement in the late venous phase in postcontrast T1-weighted images (5).

Histopathological and immunohistochemical analyses of our case were compatible with SANT without the prominent sclerotic fibrous component. As a result, the lesion did not show the characteristic spoke-wheel pattern. Therefore, this case is an unusual example of SANT, appearing hyperattenuated in the arterial phase and hypoattenuated in the portal venous and the late venous phases.

Thacker et al. detailed CT and MRI findings similar to the other reports mentioned above using FDG-PET and <sup>99m</sup>Tc-sulfur colloid SPECT CT. The SANT showed a hypermetabolic activity by FDG-PET. The <sup>99m</sup>Tc-sulfur colloid SPECT CT showed the lack of uptake due to the absence of reticuloendothelial cells (4).

Diagnosis can be made either by percutaneous biopsy or by splenectomy. Percutaneous biopsy, however, has potential risks for complications causing splenic rupture, especially for the vascular lesions such as SANT. Diagnosis and curative therapy are generally best established by surgery (4,5). Our patient underwent splenectomy for exact diagnosis and treatment. No complication was observed after surgery. The patient was discharged uneventfully.

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