

Received:

16 March 2018

Revised:

17 December 2018

Accepted:

19 December 2018

Cite this article as:

Vigorito R, Scaramuzza D, Pellegrinelli A, Marchianò A. Sclerosing angiomatoid nodular transformation (SANT) of the spleen: A case report on CT and MRI. *BJR Case Rep* 2019; **5**: 20180036.

CASE REPORT

Sclerosing angiomatoid nodular transformation (SANT) of the spleen: A case report on CT and MRI

¹RAFFAELLA VIGORITO, MD, ²DAVIDE SCARAMUZZA, ³ALESSANDRO PELLEGRINELLI and ²ALFONSO MARCHIANÒ

¹Post-graduation School in Radiodiagnostics, University of Milan, Milan, Italy

²Department of Radiology, Fondazione IRCCS Istituto Nazionale Tumori Milan, Milan, Italy

³Pathology Unit, Fondazione IRCCS Istituto Nazionale dei Tumori, Milan, Italy

Address correspondence to: Dr Raffaella Vigorito

E-mail: raffaella.vigorito@unimi.it

ABSTRACT

Solid tumors of the spleen are rare, with an incidence of 0.007% in all operating and autopsy specimens. In terms of microscopic structure and function, the spleen consists of two parts: the white pulp, which plays an important role in the immune system and the red pulp, which filters the blood. Primary splenic neoplasms can be classified into lymphoid neoplasms arising from the white pulp, and vascular neoplasms which arise from the red pulp. Primary tumors arising from vascular elements include benign lesions such as hemangioma, lymphangioma and hamartoma, intermediate lesions such as hemangioendothelioma, hemangiopericytoma and littoral cell angioma as well as the frankly malignant hemangiosarcoma. It is usually difficult to distinguish a benign from a malignant lesion with preoperative imaging studies and cytological exam by fine-needle aspiration (FNA), that is not easily obtained because of the risk of bleeding. Therefore a splenectomy should be necessary for a definitive diagnosis of splenic tumors. Martel and all for the first time described the sclerosing angiomatoid nodular transformation (SANT), like a vascular lesion of the spleen, with benign clinical course consisting by altered red pulp tissue that has been entrapped by a non-neoplastic stromal proliferative process. We describe a rare case of benign splenic mass documented with FDG/PET-CT (referred as equivocal), CT and MRI.

CASE PRESENTATION

A 56-year-old female arrived to IRCCS Istituto Nazionale Tumori Milan for a further diagnostic workup and possible treatment of incidentally hypoechoic 5 cm splenic nodule, found with abdominal ultrasonography during a follow-up, for a previous stomach GIST. The contrast-enhanced ultrasonography (CEUS) report, performed in another Institution, described a lesion with centripetal early enhancement, an anechoic central component, a progressive wash-out 44 s after injection. The nodule was evaluated as “suspicious for neoplastic lesion”.

INVESTIGATION

The patient underwent a contrast-enhanced CT, that demonstrated a solitary, round mass, with lobulated margins. On unenhanced phase the lesion had the same spleen's density. On arterial phase it was heterogeneous, with a lively rim enhancement associated to slight hyperdensity of radiating lines. On venous and delayed phase there was progressive filling. The central scar persisted hypoenhanced in all phases.¹

Axial diameters were 5 × 4.5 cm and craniocaudal distance was 5.2 cm. There was no splenomegaly or lymphadenopathy (Figure 1).

Then she underwent fludeoxyglucose (FDG-PET/CT and MRI. FDG accumulation in the tumor was low and heterogeneous with a maximum standardized uptake value (SUV_{max}) of 4.3, referred as equivocal. No abnormal FDG accumulation has been found elsewhere in the body. Considering the CT findings, a splenic benign vascular tumor was suspected; thus, hematologist and surgeon colleagues decided to perform an MRI (Figure 2).

The MRI showed a 5.4 × 5.0 cm mass, with a craniocaudal diameter of 5.4 cm. Signal's lesion was homogeneous and isointense on T_1 weighted (T_1W), without signal intensity drop on out-of-phase T_1 sequence and inhomogeneous and predominantly decreased on T_2 weighted (T_2W). Morphologically the lesion was rounded, with irregular and smooth margins; there were multiple radiating septa inside, caused by the presence of a central hypointense scar. The diffusion-weighted image (DWI), with a b-value of 800 s mm⁻²,

Figure 3. MRI pre-contrast- (a, b) the lesion isn't clearly recognized on T_1W in phase, without signal intensity drop on out of phase; (c) rounded lesion, with irregular and smooth margins, multiple radiating septa inside, with a typical "spoke wheel pattern" on T_1W , T_1 weighted; (d, e) absence of signal restriction (b -value of 800 s mm^{-2}).

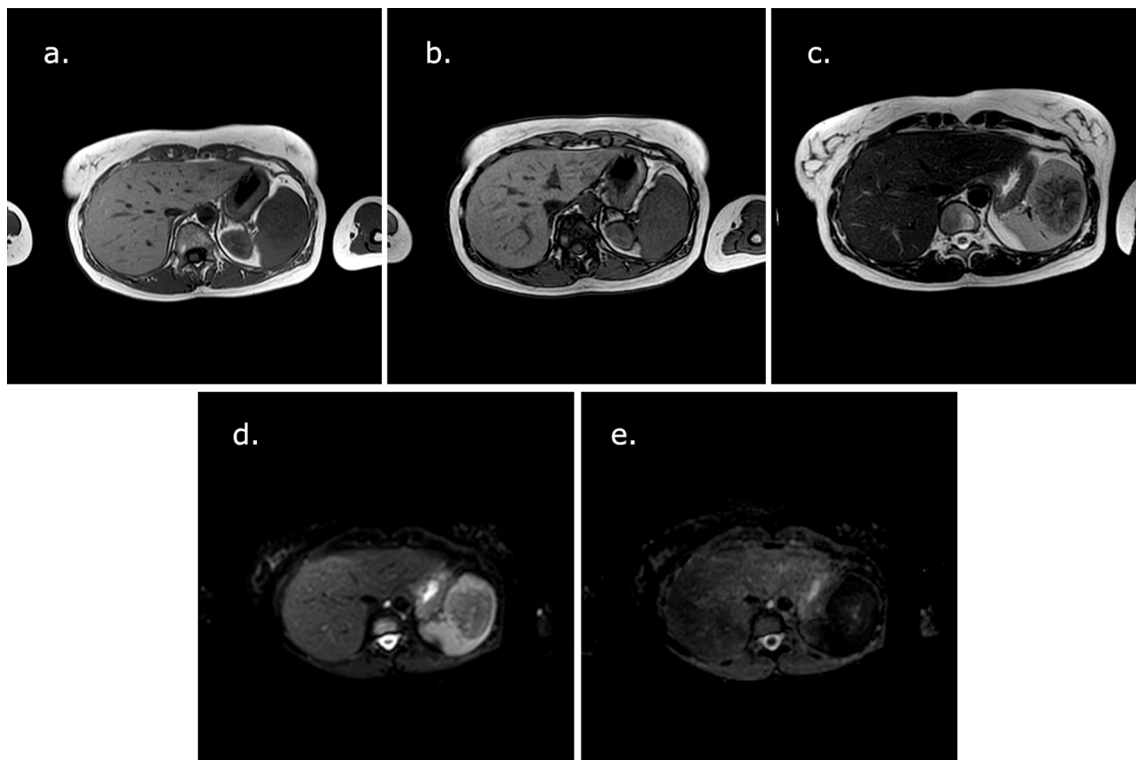
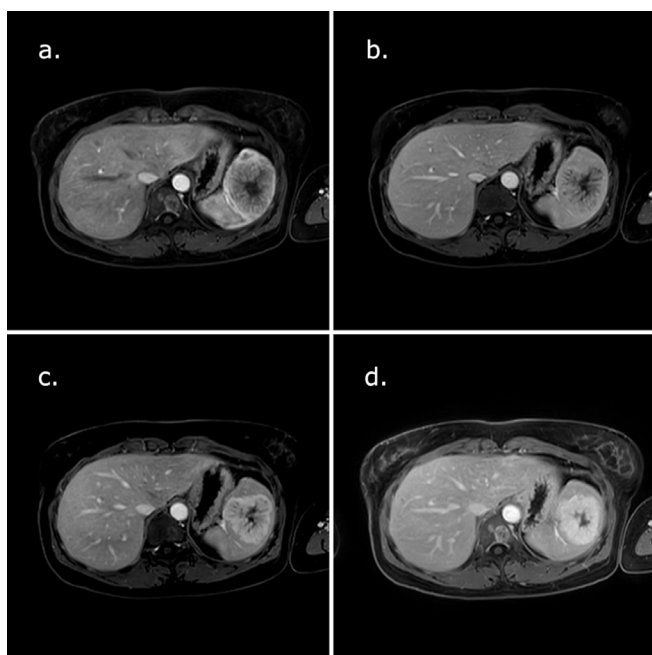


Figure 4. MRI after gadolinium administration: (a) on arterial phase peripheral and septa enhancement is recognizable; (b-d) on venous and delayed phase progressive and centripetal filling, with non-enhancing central hypointense scar is recognizable.

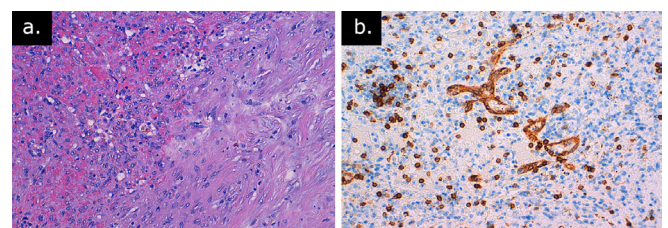


US is capable to detect vascular splenic masses, but the ultrasound findings of SANT are inconclusive,⁵ because SANT has variable appearance, variable echogenicity, and sometimes internal septa; there is usually internal vascularity on the Color Doppler.

We have found a few reports concerning contrast-enhancement behavior on CEUS; Cao et al. showed a lesion with a diffuse enhancement pattern from 11 s after injection, homogeneously hyperechoic in comparison with the splenic parenchyma in 21 s. It turned out to be isoechoic in 4 s and hypoechoic in 30 s after the injection, with a persistent enhancement up to about 7 min.⁶

A few authors have described CT and MRI findings of SANT; on CT it is solitary and round, with lobulated or smooth margins, iso- or hypoattenuating on an unenhanced scan. After medium contrast injection, the lesion shows peripheral enhancement in

Figure 5. Pathology (a) angiomatoid nodule upper left, fibrous stroma lower right (EE 20 ×); (b) cd8 positive splenic sinusoid in the angiomatoid nodule (CD8 20×).



the arterial phase, with progressive centripetal filling in a radiating pattern, especially in the delayed phase, while its center continues to be hypodense.⁷

This “spoke wheel” pattern is recognizable also on an MRI; the lesion is usually heterogeneous, iso- to hypointense on T_1 , hypointense on T_2 . Following a gadolinium administration, it shows a peripheral and septal enhancing pattern with a hypointense central scar in all sequences; the enhancement persists on venous and delayed phase.⁸

For Yoshimura et al.⁹ DWI (b -value 800 s mm⁻²) findings correlate well with the pathological findings of SANT, the multinodular high-intensity area as the angiomatoid nodules and the peripheral low-intensity area as fibrous tissues, in both cases described.

However, in the differential diagnoses also other hypervascular focal lesions of spleen, benign or malign, must be considered.

Hamartoma on a CT appears iso- or hypodense on unenhanced phase and following contrast administration demonstrates heterogeneous enhancement. On an MRI it is isointense on T_1 , and heterogeneously hyperintense on T_2 , with heterogeneous enhancement on immediate post-contrast images, uniform and intense enhancement on delayed phase, with or without central hypovascular areas.⁸

Littoral cell angiomias of the spleen are always multiple hypointense lesions with sharp margins on non-contrast CT, with an early arterial enhancement, delayed filling and eventually isointense referred to the surrounding splenic parenchyma. They are in 50% of cases hypointense on both T_1 - and T_2 weighted images, due to hemosiderin deposition.¹⁰ After contrast administration, it shows mild heterogeneous enhancement on the immediate post-contrast images, with homogeneous enhancement on delayed imaging.¹¹

Angiosarcoma has often bigger dimension and more aggressive behavior, with distant metastasis. On CT it can be either solitary or multiple, nodular, poorly marginated masses with heterogeneous attenuation. However, hyperattenuation can be seen in the setting of acute hemorrhage. Some lesions can show peripheral

enhancement. On an MRI, these lesions are hypointense on both T_1 - and T_2 weighted images relative to the spleen. In larger lesions hemorrhage and tumor necrosis areas can appear.⁸ These lesions demonstrate early contrast-enhancement with variable non-enhancing areas, related to necrosis or hemorrhage.

Actually differential diagnosis with only pre-operative imaging is difficult, because SANT is a rare entity and there are a few images in the literature. Moreover there is a realistic risk of rupture in cases of large spleen masses, so the current direction is splenectomy and histological diagnosis.

In conclusion, SANT is a rare hypervascular benign lesion, recently classified as an independent entity. Knowledge of the typical “spoke wheel” aspect, and of contrast-enhanced pattern, recognizable on CT or MRI, can be helpful in a preoperative diagnosis, avoiding histological examination. A good diagnostic accuracy of DWI MRI imaging is also reported.

LEARNING POINTS

1. SANT of the spleen is a distinct benign pathological entity, microscopically characterized by multiple angiomatoid nodules separated by collagenous bundles.
2. At diagnostic examinations SANT morphology can mimic other benign or malignant splenic lesions.
3. Typical morphology and contrast-enhancement behavior on CT and MRI can be helpful for radiologic diagnosis. SANT is usually a solitary, rounded, with lobulated or smooth margins, multiple radiating septa inside, caused by the presence of hypodense/hypointense a central scar. Following intravenous administration of a contrast medium on arterial phase a peripheral and septa enhancement has been observed, with progressive and centripetal filling, particularly during the delayed phase, with a persistent non-enhancing central scar.

CONSENT

Written informed consent for the case to be published (including images, case history and data) was obtained from the patient(s) for publication of this case report, including accompanying images

REFERENCES

1. Moriyama S, Inayoshi A, Kurano R. Inflammatory pseudotumor of the spleen: report of a case. *Surg Today* 2000; **30**: 942–6. doi: <https://doi.org/10.1007/s005950070051>
2. Martel M, Cheuk W, Lombardi L, Lifschitz-Mercer B, Chan JK, Rosai J. Sclerosing angiomatoid nodular transformation (SANT): report of 25 cases of a distinctive benign splenic lesion. *Am J Surg Pathol* 2004; **28**: 1268–79.
3. Falk GA, Nooli NP, Morris-Stiff G, Plesec TP, Rosenblatt S. Sclerosing Angiomatoid nodular transformation (SANT) of the spleen: case report and review of the literature. *Int J Surg Case Rep* 2012; **3**: 492–500. doi: <https://doi.org/10.1016/j.ijscr.2012.06.003>
4. Atas H, Bulus H, Akkurt G, Ozkan H. Sclerosing Angiomatoid nodular transformation of the spleen: an uncommon cause of abdominal pain. *Euroasian J Hepatogastroenterol* 2017; **7**: 89–91. doi: <https://doi.org/10.5005/jp-journals-10018-1221>
5. Wang HL, Li KW, Wang J, HI W, KW LI. Sclerosing angiomatoid nodular transformation of the spleen: report of five cases and review of literature. *Chin Med J* 2012; **125**: 2386–9.
6. Cao J-Y, Zhang H, Wang WP. Ultrasonography of sclerosing angiomatoid

- nodular transformation in the spleen. *World J Gastroenterol* 2010; **16**: 3727–30. doi: <https://doi.org/10.3748/wjg.v16.i29.3727>
7. Karaosmanoglu DA, Karcaaltincaba M, Akata D. CT and MRI findings of sclerosing angiomatoid nodular transformation of the spleen: spoke wheel pattern. *Korean J Radiol* 2008; **9 Suppl**: S52–5. doi: <https://doi.org/10.3348/kjr.2008.9.s.s52>
 8. Bowerson M, Menias CO, Lee K, Fowler KJ, Luna A, Yano M, et al. Hot spleen: hypervascular lesions of the spleen. *Abdom Imaging* 2015; **40**: 2796–813. doi: <https://doi.org/10.1007/s00261-015-0523-8>
 9. Yoshimura N, Saito K, Shiota N, Suzuki K, Akata S, Oshiro H, et al. Two cases of sclerosing angiomatoid nodular transformation of the spleen with gradual growth: usefulness of diffusion-weighted imaging. *Clin Imaging* 2015; **39**: 315–7. doi: <https://doi.org/10.1016/j.clinimag.2014.10.015>
 10. Abbott RM, Levy AD, Aguilera NS, Gorospe L, Thompson WM. From the archives of the AFIP: primary vascular neoplasms of the spleen: radiologic-pathologic correlation. *Radiographics* 2004; **24**: 1137–63. doi: <https://doi.org/10.1148/rg.244045006>
 11. Falk S, Stutte HJ, Frizzera G. Littoral cell angioma. A novel splenic vascular lesion demonstrating histiocytic differentiation. *Am J Surg Pathol* 1991; **15**: 1023–33.