

CASE REPORT

Sclerosing angiomatoid nodular transformation of the spleen

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Case

A 56-year-old gentleman presented under the surgical team with left upper quadrant pain and mild anemia (Hb 11.2 gm/dL). He reported feeling well aside from intermittent left upper quadrant pain and denied any weight loss. He underwent a CT scan which showed a 10 × 10 × 9 cm rounded lesion within the spleen with small areas of central calcification (Fig. 1). Spleen size was normal. There was no evidence of lymphadenopathy or any other neoplastic process from the CT scan. Subsequent blood tests revealed normal LDH and serum protein electrophoresis. Bone marrow biopsy showed active trilineage hemopoiesis without any evidence of lymphoma or other infiltrative neoplastic process.

He initially underwent splenic core needle biopsy. This showed paucicellular fibrosis with medium-caliber vessels embedded in fibrous tissue showing signs of obliteration and/or recanalization. He subsequently underwent splenectomy to characterize these splenic lesions as splenic biopsy itself was not completely diagnostic.

Resected spleen (175 × 110 × 95 mm) weighed 578 g. It contained a central bulky stellate mass of white compact tissue (50 × 40 × 45 mm), surrounded by multiple poorly defined hard dark-brown nodules (Fig. 2). Microscopically, the center of the lesion consisted of almost

Key Clinical Message

Sclerosing angiomatoid nodular transformation (SANT) of spleen is a very rare benign entity with unknown etiology. Here, we report this unusual case in a fit middle-aged gentleman and discuss various diagnostic modalities along with the management of this condition.

Keywords

Incidental splenic mass, splenectomy, splenic biopsy, spokes wheel pattern.

acellular hyaline, whereas the peripheries comprized macro-nodular aggregates of round angiomatoid structures, each composed of a chaotic meshwork of capillary-like vessels in the middle (CD34+, CD31+, CD8–), rimmed by a band of collagen-rich connective tissue, and scant smooth muscle cells. There was no nuclear atypia in this case (Fig. 3,4,5,6).

The appearances were diagnostic for sclerosing angiomatoid nodular transformation of the spleen (SANT), a very rare entity first recognized in 2004, distinct from much more frequent hamartomas and angiomas. In retrospect, features of this lesion were also seen in the core needle biopsy, which, however, was hardly diagnostic on its own.

This patient has remained well and is now 2 years post splenectomy.

Discussion

Sclerosing angiomatoid nodular transformation was originally described in 2004 by Martel et al [1]. Most people are asymptomatic at presentation. Common presentations are incidental splenic mass, abdominal discomfort, or splenomegaly. There is slight female preponderance and predominantly affects middle aged adults [1, 2].

Radiologically, they present as solitary, round, lobulated mass which is centrally hypodense with peripheral-

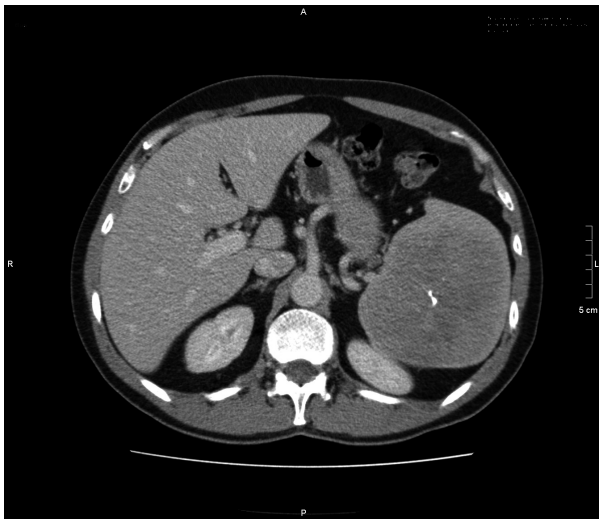


Figure 1. Computed tomography (CT).



Figure 2. Spleen Macro.

enhancing portions. Contrast CT/MRI scan shows heterogeneously hypo-enhancing lesion during arterial and venous phase with an early peripheral-enhancing radiating lines and delayed enhancement of the central area due to fibrous tissue. MRI appearances on T1-weighted images have been described as “spokes wheel pattern” [3, 4].

It is noted that these lesions are FDG-avid. In few reported cases where PET CT is employed, FDG-avid multiple splenic nodules with a “prunes on bread” appearance in the maximum-intensity-projection image (MIP image) are seen. In sectional PET/CT images, a

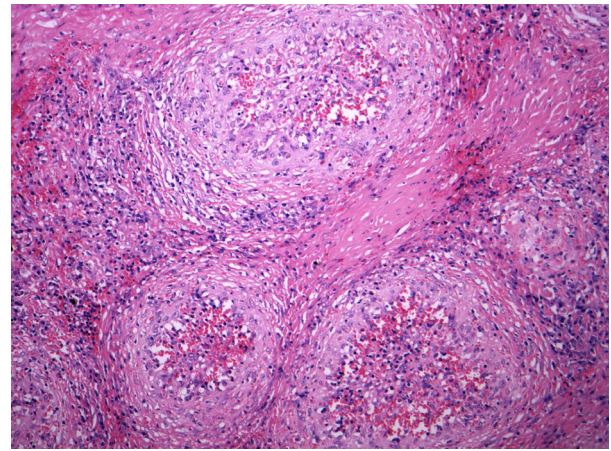


Figure 3. Spleen H+E.

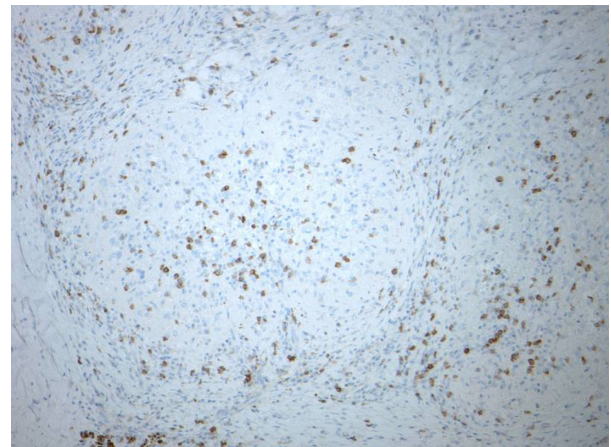


Figure 4. CD8 immunostain.

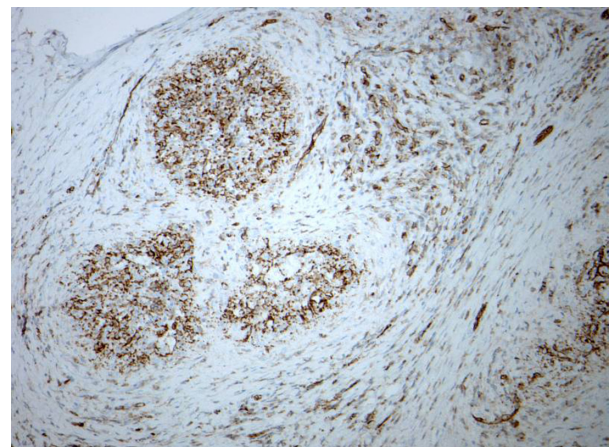


Figure 5. CD31 Immunostain.

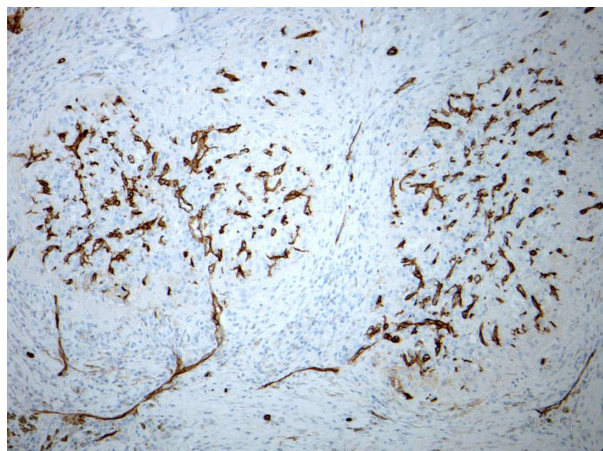


Figure 6. Cd34 immunostain.

central cold area with peripheral increased FDG uptake in the splenic nodule is noted [5].

Histology of the spleen consistently shows multinodular appearance at lower power examination. Individual nodules have angiomatoid appearance. Nuclear atypia is rare and necrosis is almost never seen. Immunostaining of the vessels simulate the composition of normal splenic red pulp and features are different from angioma, hemangioma of the spleen. Myofibroblasts are highlighted within the nodules and endothelial lining cells are D2-40 negative supporting vascular over lymphatic origin. Vessels in the angiomatoid nodular are the combination of three different types: capillaries (CD34+/CD8-/CD31+), small veins (CD34-/CD8-/CD31+), and sinusoids (CD34-/CD8+/CD31+). Some cells in SANT are CD68+ and SMA+ [1, 6].

Differential diagnoses of splenic lesions include both benign and malignant disease. It is difficult to rule out other benign pathological conditions of the spleen such as inflammatory pseudotumor or hamartoma using imaging modalities. However, it may be possible to differentiate benign and malignant splenic lesions with a combination of clinical history and radiological modalities. Asymptomatic patient with hyper echoic lesions point toward benign, whereas patients with systemic symptoms and hypo echoic lesions on the scan should point toward possible malignant etiology. However, nodular carcinomatous metastasis to spleen cannot be sufficiently be diagnosed by imaging modalities alone.

Diagnosis of SANT requires high clinical suspicion. Although histological examination of the spleen is always employed to give the diagnostic yield, there is a debate if one should be subjecting an asymptomatic patient through a surgical procedure to obtain diagnosis. There were diagnostic clues from splenic biopsy in our case, but the diagnosis of most of the published cases has been

based on a splenectomy specimen. Splenic biopsy has been employed and Weinreb et al. suggests that good core biopsy can be used to distinguish SANT from other lesions. However, there is a worry about risk of intra peritoneal seeding if the lesion biopsied proves to be angiosarcoma and other complications such as splenic rupture and bleeding are noted with splenic biopsy.

Pathogenesis of SANT is unclear. Martel et al. postulated that SANT was a response to stromal proliferation and that the internodular zones were similar to inflammatory pseudotumor [1, 7]. SANT may represent a hamartomatous transformation of splenic red pulp in response to an exaggerated nonneoplastic stromal proliferation. In majority of cases, the etiology is unknown. A link to Epstein–Barr virus was postulated by Weinreb et al. but this has not been consistently found in other case series. SANT-like changes may be occasionally seen in vicinity of splenic metastases, but there was no evidence of any malignant/metastatic process in this case.

Splenectomy serves as both diagnostic and therapeutic option. SANT is a benign lesion which may be nevertheless symptomatic in some patients. Splenectomy is curative.

Conflict of Interest

None declared.

References

- Martel, M., W. Cheuk, L. Lombardi, B. Lifschitz-Mercer, J. K. Chan, and J. Rosai. 2004. Sclerosing angiomatoid nodular transformation (SANT): report of 25 cases of distinctive benign splenic lesion. *Am. J. Surg. Pathology* 28:1268–1279.
- Falk, G. A., N. P. Nooli, G. Morris-Stiff, T. P. Plesec, and S. Rosenblatt. 2012. SANT of the spleen: case report and review of literature. *Int. J. Surg. Case Rep.* 3:492–500.
- Lewis, R. B., et al. 2013. Sclerosing angiomatoid nodular transformation of the spleen: CT and MRI features with pathological correlation. *Gastroint. Imaging*, 200, 4.
- Devrim, A., et al. 2008. CT and MRI findings of sclerosing angiomatoid nodular transformation of the spleen: spoke wheel pattern. *Korean J. Radiol.* 99(Suppl.):S52–S55.
- Feng Y. M., Y. C. Huang, C. W. TU, W. S. Kao, and D. G. Tu. 2013. Distinctive PET/CT features of splenic SANT. *Clin. Nucl. Med.* 38:e465–e466.
- Wang, H. L., K. W. Li, and J. Wang. 2012. Sclerosing angiomatoid nodular transformation of the spleen: report of five cases and review of literature. *Chin. Med. J. (Engl.)* 125:2386–2389.
- Diebold, J., et al. 2008. Is sclerosing angiomatoid nodular transformation of the splenic red pulp identical to inflammatory pseudotumour? Report of 16 cases. *Histopathology* 53:299–310.