

CASE REPORTS

Rare tumor of the spleen – case report

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ABSTRACT

Sclerosing angiomatoid nodular transformation was first described as a solitary angioma-like disease of the spleen by Martel *et al.* in 2004. It is a relatively new rare benign lesion. The authors present a clinical case of an asymptomatic, 48-year-old female. SANT has been an incidental finding in an abdominal ultrasound. Histological appearance and the immunohistochemical staining after spleen biopsy confirmed the diagnosis. As suggested in previously published articles, a splenectomy was performed for definitive treatment. This case contributes to the differential diagnosis of splenic tumors, its treatment and clinical impact.

Key Words: Spleen tumors, Sclerosing angiomatoid nodular transformation, Splenectomy

1. INTRODUCTION

Sclerosing angiomatoid nodular transformation of the spleen (SANT) is a relatively new rare benign lesion whose pathogenesis has not been fully understood yet. First described^[1] as a solitary angioma-like disease of the spleen in 2004, there are now 129 cases reported in the literature.^[2] The majority of the cases described are solitary nodules. However, there are 6 case reports of multiple SANT of the spleen.^[2] Most patients are asymptomatic and SANT is usually an incidental finding in a routine imaging study. Classically, it is considered a disease of female preponderance. Patients are usually between 30 and 60 years old.^[3] Notwithstanding the fact that SANT has specific imaging findings, differential diagnosis with other tumors or malignant lesions is very difficult.^[3,4] Other than hematopoietic neoplasms like lymphoma, the most common primary nonhematopoietic tumors of the spleen contemplate hemangioma, littoral cell angioma, lymphangioma, hemangioendothelioma, hamartoma and SANT.^[5] The histological appearance and the immunohistochemical staining makes the diagnosis.^[6]

2. CASE PRESENTATION

The authors report the case of a 48-year-old woman with a history of anemia, in regular follow-up. She was asymptomatic and had a normal physical examination. Laboratory test results showed anemia without any other abnormalities. The hemoglobin value was 10.7 g/dl, the MCV was 80.7 fl, the white blood count was 7,600/ μ l, the platelets were 311,000/ μ l and all other data were within normal limits. A routine abdominal ultrasound found indications of an isoechoic nodular image in the superior pole (58.8 mm \times 50.7 mm) of a normal dimensioned spleen. The ultrasound was followed by an abdominal computed tomography (CT) scan that revealed a hypodense tumor of the spleen with 5.5 cm (see Figure 1).

Within a month the patient underwent an ultrasound guided spleen biopsy. The histopathological examination showed nodular splenic tissue with nodular capillary growths separated by fibrous septa, with histiocytic periphery and pseudo-granulomatous appearance (see Figure 2). The result was compatible with SANT.

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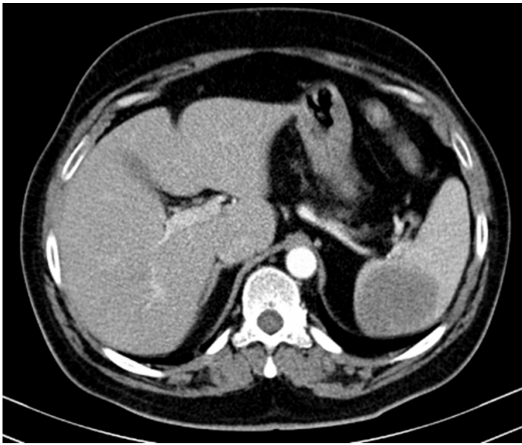


Figure 1. First abdominal CT (axial section) showing the spleen of our patient with a hypodense mass with 55 mm

Control CT scan showed that the tumor was not enlarged (6.0 cm) five months later (see Figure 3). The patient remained asymptomatic with a normal physical examination. According to most of the papers published regarding this topic, we decided for splenectomy as the safest treatment for our patient.

Roughly the spleen weighed 194 g, its external surface was congested, the upper pole showed nodularity (see Figures 4 and 5). Cut surface showed a well-circumscribed mass with 5.3 cm × 5 cm × 4.5 cm, heterogeneous, with fibrous septa traversing throughout, which divided it into discrete brownish nodules with a yellow center.

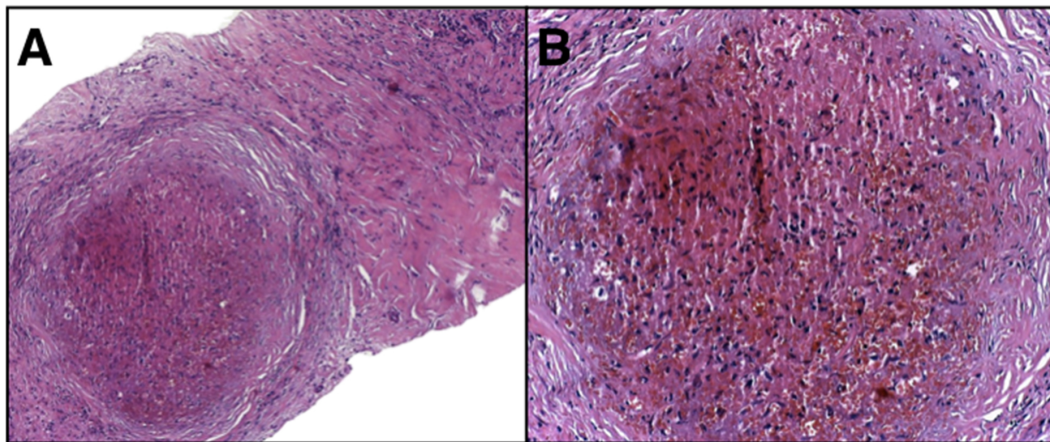


Figure 2. Microscopic examination of the spleen. A: Individual nodule of sclerosing angiomatoid nodular transformation shows bands of sclerosis surrounding an arrangement of vasculature; B: Splenic tissue with capillary growths separated by fibrous septa and histiocytic peripheral cells with pseudo-granulomatous appearance

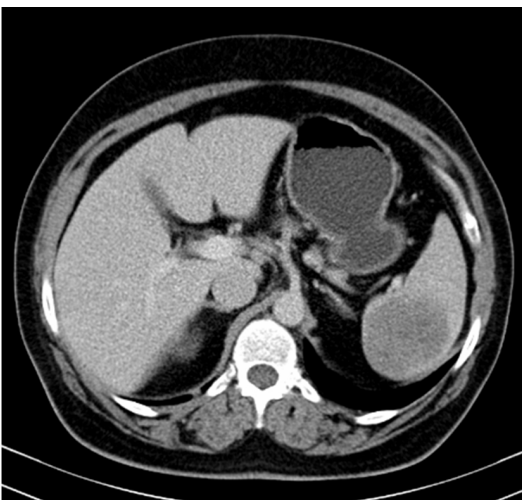


Figure 3. Abdominal CT (axial section), five months later, revealing a hypodense mass of the spleen with 60 mm

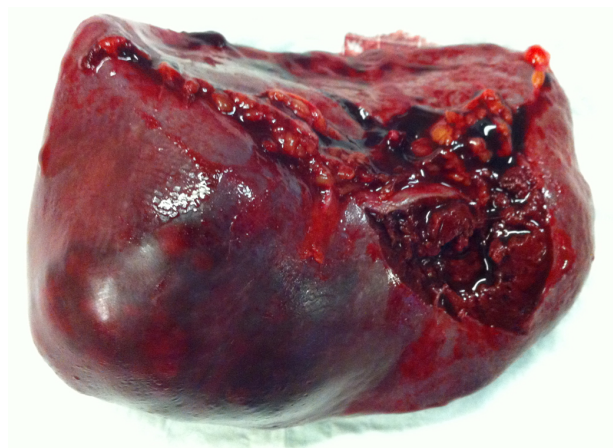


Figure 4. Sclerosing angiomatoid nodular spleen. Spleen weighing 194 g, after surgical removal, showing a nodular tumor in the upper pole



Figure 5. Macroscopic aspect of the spleen. The cut surface of the spleen reveals a beefy-red coloured congested parenchyma. The upper pole contains a well-circumscribed 5.3 cm nodule with a central area of pallor and fibrosis

Microscopy confirmed the previous biopsy. It showed nodular splenic tissue, with peripheral sclerosis and vascular spaces. The fibrous septa formed by fibroblasts, hemosiderophages and lymphomononuclear cells, separated

the nodules. Immunohistochemistry revealed a variable admixture of CD8, CD31 and CD34-positive vessels which indicates derivation from vein, sinusoidal and capillary-like elements and confirmed the diagnosis of SANT of the spleen (see Figure 6). One year later the patient remains asymptomatic.

3. DISCUSSION

The term SANT first appeared in scientific literature in a 2004 paper which examined a series of 25 cases. This uncommon splenic lesion had however been recognized earlier by other authors under different names such as splenic hamartoma, cord capillary hemangioma and multinodular hemangioma.^[6] SANT is a benign splenic condition that is more common in women rather than in men. However, as the number of reported cases grows, it was recently described that gender difference in prevalence is getting smaller.^[6] It has variable clinical presentation. Most patients are asymptomatic as published on a series by Diebold *et al.* Nevertheless, some patients presented abdominal pain and discomfort or splenomegaly.^[5] When symptomatic, the most common symptom is abdominal pain. Cytopenias, pelvic pain, flank pain, palpable left upper quadrant mass, and long standing fever are alternative presentations.^[6] The splenic lesions frequently represent incidental findings on imaging studies performed for different motives, as the present case reports.

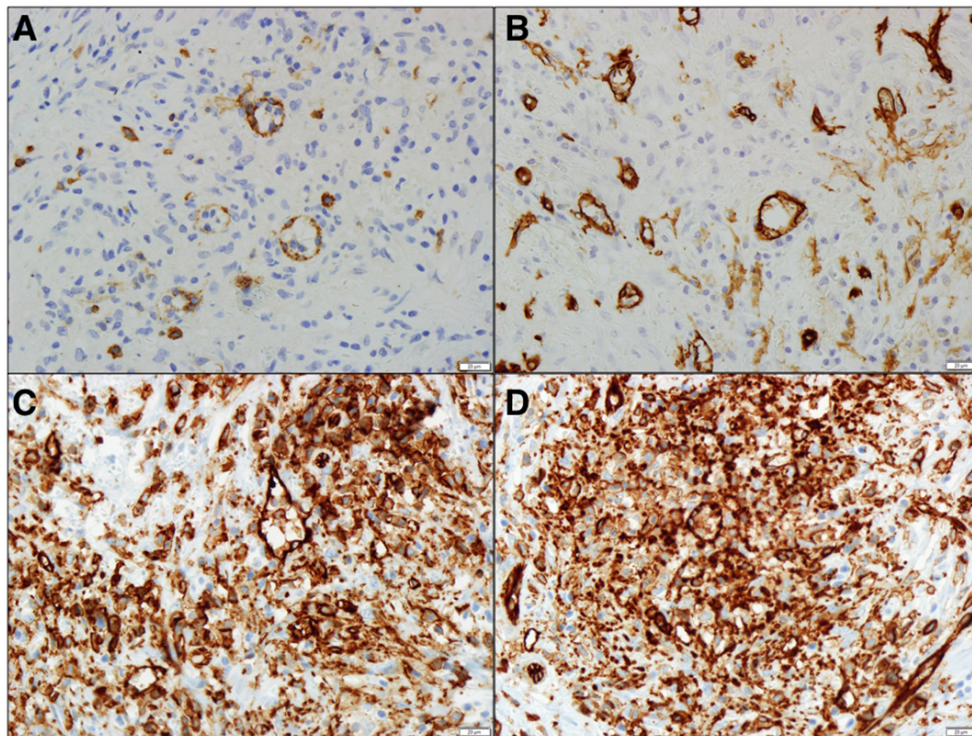


Figure 6. Immunohistochemistry examination of the spleen. A: CD8 immunostain highlights sinusoid-like spaces; B: CD34 immunostain highlights the capillaries; C and D show immunoreactivity for CD31

SANT presents itself on CT scans and magnetic resonance imaging has a hypodense mass. The differential diagnosis includes neoplasms like lymphoma, metastatic lesions and benign lesions such as hamartoma,^[5] which emphasizes the importance of the diagnostic confirmation. Spleen biopsy can be performed and makes the diagnosis.^[2] Ultrasound guided percutaneous splenic biopsy is an accurate and relatively safe procedure. Major complication risk (1.3%) does not exceed other solid abdominal organ biopsies, and has less mortality and morbidity than splenectomy.^[7] However, the authors couldn't find any published cases where splenectomy wasn't the first choice for diagnosis. The benign course, distinctive morphologic appearance, immunophenotype suggest that SANT is a dissimilar non-neoplastic vascular lesion. Individual nodules are composed of slit-like, globular or irregular-shaped vascular spaces interspersed by a population of thin or ovoid cells and lined up by pudgy endothelial cells conferring them an angiomatoid appearance.^[5] Usually they have clear margins with normal parenchyma. SANT is reliably distinguished from other tumors by immunohistochemical staining. Angiomatoid nodules display three distinct types of vessels: CD34⁺/CD8⁻/CD31⁺ small veins, CD34⁺/CD8⁺/CD31⁺ sinu-

soids and CD34⁺/CD8⁻/CD31⁺ capillaries.^[5] Other splenic vascular lesions don't meet these criteria. In this case, the authors decided to perform an ultrasound-guided spleen biopsy as it meant a shorter length of stay in the hospital and an earlier return to normal activity. The result was compatible with SANT. Nevertheless, and according to the literature, we decided to perform a splenectomy.

SANT should be considered in any patient with a splenic lesion containing an inflammatory or angiomatoid component. The majority of diagnosis are incidental, given that symptoms are absent in most cases, or are very unspecific when present. The final diagnosis is only possible with a histopathology which is feasible by ultrasound-guided spleen biopsy.

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CONFLICTS OF INTEREST DISCLOSURE

The authors have declared no conflicts of interest.

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