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Sclerosing angiomatoid nodular transformation of the spleen: an unusual cause of abdominal pain

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ABSTRACT

Vascular neoplasms are the most common primary nonhematopoietic tumors of the spleen. They include hemangiomas, littoral cell angiomas, splenic hamartomas, lymphangiomas, hemangioendotheliomas, angiosarcomas, and Sclerosing Angiomatoid Nodular Transformation (SANT)1. SANT is a rare and benign lesion arising from the red pulp of the spleen 2.Martel et al first descried the disease entity in 2004 in middle aged- adults with slight female preponderance , the etiology and pathogenesis are still not clear3, SANT poses diagnostic clinical and radiological challenges4, we present this case report with review of the literature.

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Introduction:

Vascular neoplasms are the most common primary nonhematopoietic tumors of the spleen. They include hemangiomas, littoral cell angiomas, splenic hamartomas, lymphangiomas, hemangioendotheliomas, angiosarcomas, and Sclerosing Angiomatoid Nodular Transformation (SANT)[1]. SANT is a rare and benign lesion arising from the red pulp of the spleen [2].Martel et al first descried the disease entity in 2004 in middle aged- adults with slight female preponderance , the etiology and pathogenesis are still not clear[3], SANT poses diagnostic clinical and radiological challenges[4], we present this case report with review of the literature.

Case report:

A 25 year-old female presented with mild epigastric, left upper quadrant pain for one year. On physical exam the spleen was not palpable and peripheral lymph node examination was normal. Complete blood count and biochemistry parameters were all within the normal range. Her medical and family history was unremarkable.

Abdominal CT showed well defined splenic lesion noted in the lower pole of the spleen measuring about 3.6 x 3.5 cm in maximum anteroposterior diameter. On MRI, the lesion was isointense to parenchyma on T1-weighted sequences and mildly hyperintense with heterogeneous progression on T2-weighted images. A provisional diagnosis of SANT was made and this was discussed with the patient who elected for splenectomy. Laparoscopic splenectomy was performed which revealed a large spleen with multiple superficial nodules. The patient had uneventful recovery and was discharged on the 4th day of surgery. Histopathological examination of the splenic lesion demonstrated multinodular growth pattern on low-power (Figure 1). The nodules composed of slit like, round, or irregularly shaped vascular spaces surrounded by dense collagen fibrosis (Figures 2&3). The vascular channels of varying caliber lined with plump endothelium highlighted by CD34 immunohistochemical stain (Figure 4). There are numerous lymphocytes and plasma cells in the fibrotic stroma with no evidence of malignancy. According to the histopathological findings; the lesion was diagnosed as SANT. The patient was checked at regular intervals after surgery, and the patient remained asymptomatic.

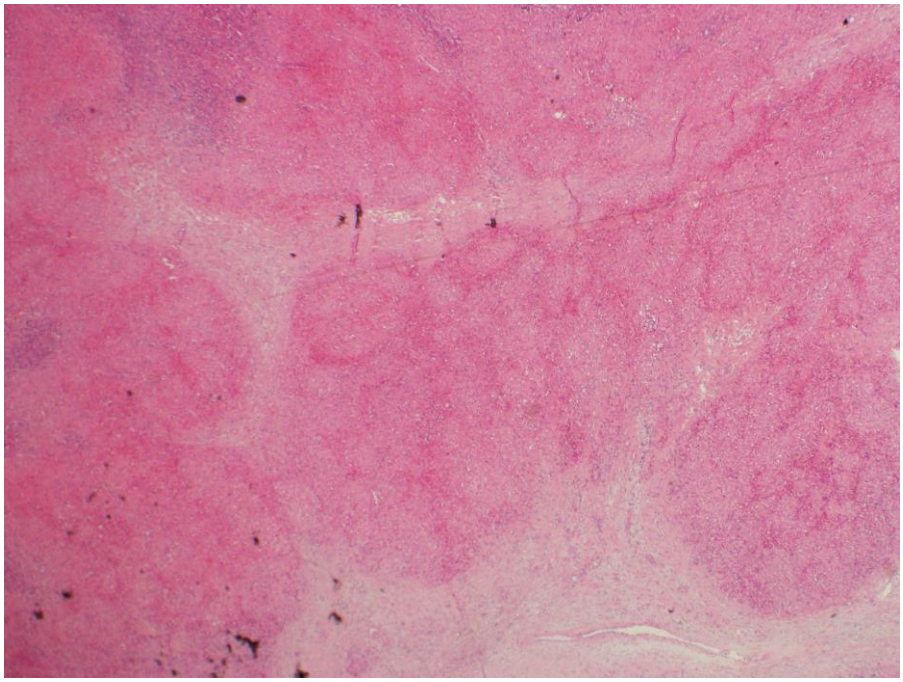
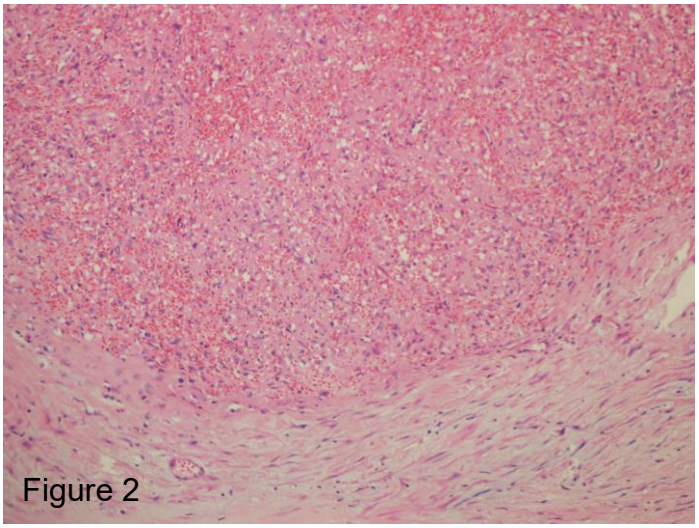
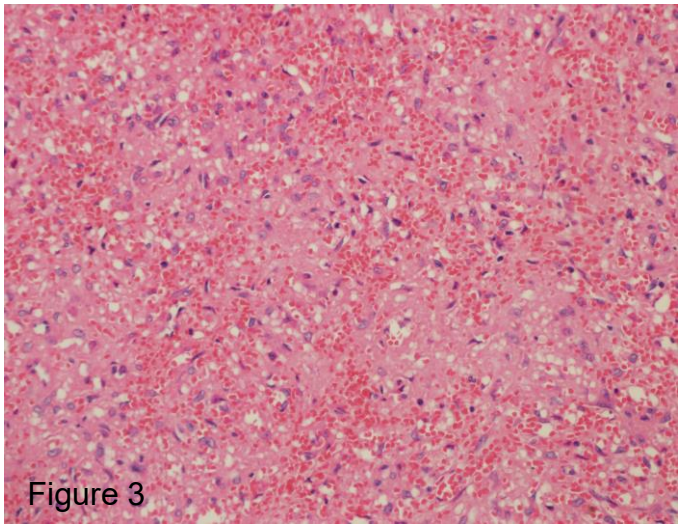


Figure 1 Multinodular growth pattern on low-power





Figures 2&3 The nodules composed of slit like, round, or irregularly shaped vascular spaces surrounded by dense collagen fibrosis.

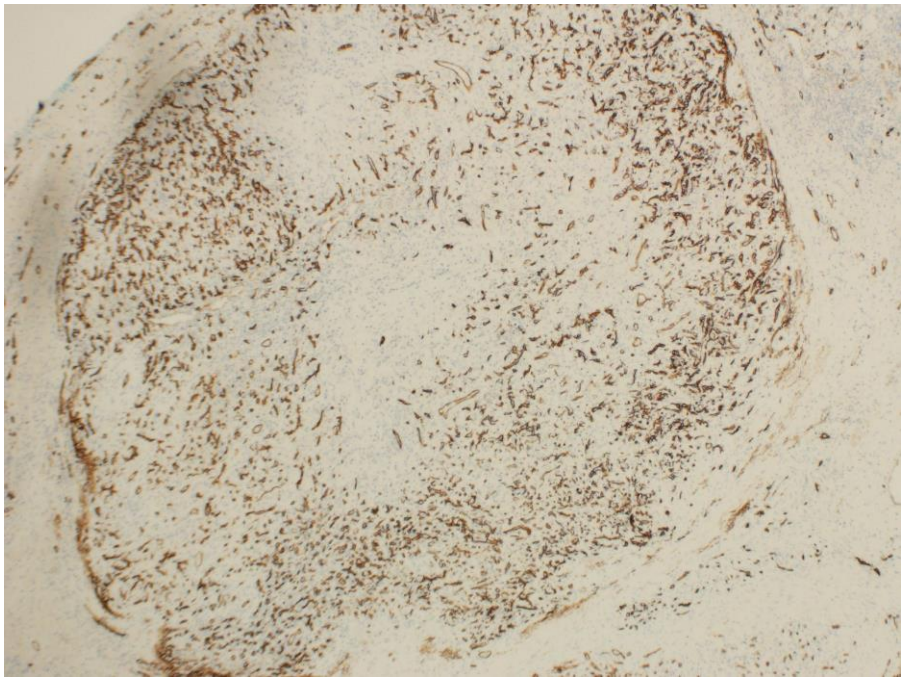


Figure 4The vascular channels of varying caliber lined with plump endothelium highlighted by CD34 immunohistochemical stain

Discussion:

Lymphoid tumors, such as lymphoma are the most common neoplasms of the spleen, whereas vascular tumors were rarely reported. Among those, hemangiomas, hemangio-endotheliomas, and hamartomas are the most common variants. On the contrary, SANT is an extremely rare and benign vascular lesion. SANT is usually described as described as multiple angiomatoid nodules embedded in a fibrosclerotic stroma, and vascular spaces surrounded by endothelial cells within each individual nodule. Etiology and pathogenesis were not clearly described in the literature, however multiple theories are found in literature. Martel et al reported on the largest series with 25 patients and suggested that angiomatoid nodules developed as a consequence of vascular obstruction [5].

Weintrib et all suggested that infection withEpstein-Barr virus may be associated with SANT in some cases[6], however Chang et al

reported on 20 cases of SANT using in situ hybridization did not find evidence of EBV RNA in any of the specimens [7]. More recently, Kuo et al showed that patients with SANT had elevated levels of immunoglobulin (Ig)G4 and IgG antibodies, considering IgG4-related sclerosing process in the pathogenesis of SANT[8], similar finding were reported by Kahiwagi et who observed 4 to 13 fold increase in IgG14 in SANT spleens as compared to normal spleen[9]. In our case, no virologic and immunological associations were found.

Patient with SANT usually have vague and non-specific abdominal pain[10-12]. As a result; the majority of SANT are found incidentally on radiographic images or intra-operatively during surgeries of unrelated conditions[13-15]. Other unusual presentations include, Splenic abscess[16;17], it can also mimic metastatic tumors and therefore causes major diagnostic dilemma[18-24] .This was the case in our patient who suffered from intermittent non-specific abdominal pain for one year and was diagnosed with SANT incidentally with a CT scan. More than 100 case reports showed patient age rang from 3 years[25] to 74 [5] , however most cases present in early adulthood.

Radiological methods including US, CT, and MRI are useful in the diagnosis of SANT. Li et aldescribed the CT appearance as an isodense appearance relative to splenic parenchyma at the late portal phase with a calcified focus[26]. Spoke wheel appearance can also identify SANT on MRI and CT [27], while Cao et al used Contrast Enhanced Ultrasound for diagnosis[28], however it has to be said that there is no single radiological method that has been consistently used for diagnosis.

In our case, a preliminary diagnosis of SANT was considered according to the CT findings of the lesion. SANT was not easily diagnosed pre-operatively due to the rarity of the entity. Differential diagnosis of SANT includes both benign and malignant lesions, such as hemangioma, littoral cell angioma, inflammatory myofibroblastic tumor, and hamartoma. Percutaneous biopsy was avoided due to the serious potential complications such as hemorrhage and seeding of tumor cells in case of it being a malignant lesion.

If SANT is suspected, Splenectomy is performed in order to exclude malignancy and prevent potential risk of abdominal bleeding. Open or laparoscopic splenectomy are the mainstay surgical approach in patients with SANT, and allows complete cure. In our case laparoscopic spleenectomy was performed for the tumor. SANT has a good prognosis without a risk of recurrence, and no malignant transformation was reported to date.

In conclusion, SANT is a rare and unusual cause of abdominal pain, and represents a diagnostic dilemma. High level of alertness from radiologists and clinicians should exist with the presence of solitary splenic mass with nonspecific clinical presentations. Large series are necessary for better understanding of this new pathological entity.

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