Sclerosing Angiomatoid Nodular Transformation (Sant) Involving Spleen: A Case Report

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ABSTRACT

Background: Sclerosing angiomatoid nodular transformation (SANT) also known as multinodular haemangioma is a recently recognized, rare non-neoplastic vascular sclerosing mass-like lesion of the spleen with uncertain etiology, presents most commonly as an asymptomatic splenic mass found incidentally on imaging studies but may cause abdominal pain or discomfort, leading to splenectomy, as the risk of rupture and malignancy cannot securely be ruled out. Diagnosis is made postoperatively on histology.

Case report: We report a case of 64-years old man, with incidentally detected splenic lesion on imaging who underwent elective splenectomy and was diagnosed postoperatively on histological examination. The post-operative course was uneventful and the patient was discharged on POD9.

Conclusion: SANT of the spleen although rare, is to be considered as a differential diagnosis of splenic vascular lesions. Further research should focus on clinical and radiological characteristics and consideration of observation with serial imaging as an alternative approach. Open or laparoscopic splenectomy is still known to be curative as it is both therapeutic and diagnostic.

Keywords: Sclerosing angiomatoid nodular transformation (SANT), incidental, splenectomy

INTRODUCTION

Splenic masses may be identified during the workup of symptoms presenting as abdominal pain or discomfort, a common presentation in the surgical department or incidentally detected during imaging which alone does not always result in a definitive diagnosis. Often long-lasting symptoms lead to a variety of diagnostic procedures, which in turn lead to incidental findings. These lesions may need to be followed serially, or if concerning, splenectomy should be considered. Splenic neoplasms generally arise from the lymphoid or vascular elements of the spleen. Sclerosing angiomatoid nodular transformation (SANT) is a benign vascular lesion first defined by Martel in 2004 and is one such very rare incidental finding. SANT consists of altered red pulp trapped by non-neoplastic stromal proliferation. There is often a central scar. It usually occurs in the middle-aged group with a female predominance. Ultrasound shows a hypoechoic lesion. CT and MRI studies may show a central scar, enhancing capsule, and radiating bands corresponding to fibrosis. The lesion may have $^{18}$F-fluorodeoxyglucose (FDG) avidity on positron emission tomography (PET) scan. Although SANT often displays characteristic radiologic findings, differentiation from other benign and malignant lesions may be challenging, moreover risk of spontaneous rupture and possible malignancy of the suspicious lesion cannot be ruled out, therefore splenectomy is performed.
We report a case of 64-years old man, with incidentally detected splenic lesion on imaging who underwent elective splenectomy and was diagnosed postoperatively on histological examination.

CASE REPORT
A 64-year-old non-smoker and non-drinker man, native of Rajasthan, reporting no health complaints, underwent abdomen-pelvic ultrasonography as a part of a routine health check-up presented to the outpatient department. The patient reportedly had hypertension and was on treatment with nebivolol 5mg/dl, telmisartan 40mg/dl, and amlodipine 5mg/dl. The patient had an operated history of TURP in July 2022. At presentation pulse rate of the patient was 78/min, blood pressure of 150/92mmHg, respiratory rate of 16/min, and spo2 was 98% on room air. On clinical abdominal examination- no abdominal distension was seen, the abdomen was soft, non-tender and no palpable mass felt. USG of the abdomen and pelvis demonstrated spleen with partially defined hypoechoic mass/area of approximately 40x36mm in the parenchyma and Post TURP status of the prostate. CECT Abdomen and pelvis were performed and demonstrated a benign lesion in the body of the spleen with centripetal enhancement, likely to be a haemangioma. (Figure.1) Based on history, examination, USG abdomen, and CECT abdomen findings diagnosis of splenic haemangioma was kept. The patient underwent elective open splenectomy. Intraoperatively there was approximately 3.5*3.5cm white firm mass present in the spleen. On the cut surface- the lesion showed few hemorrhagic points in the background of a white firm mass (Figure.2). Intraoperative and postoperative period was uneventful and the patient was discharged on 9th POD in stable condition. The diagnosis was made postoperatively on histological examination which on gross examination demonstrated a grey-brown specimen of spleen measuring around 10*7.5*4 cm with a 3.5x3cm hard area which on the cut surface shows a well-demarcated grey-white area measuring around 3.5cm in diameter surrounded by normal spleen tissue with small hemorrhagic areas. On microscopic examination, multiple sections from the grey-white area examined showed multiple nodules having increased vascularity and separated by fibrous stroma, suggestive of sclerosing angiomatoid nodular transformation.

Figure.1 CECT Abdomen showing benign lesion of the spleen with centripetal enhancement
Figure 2 Intra-operative photograph of spleen

Figure 3 Histopathological examination of sclerosing angiomatoid nodular transformation (SANT)

DISCUSSION

The term SANT, first appeared in the literature in 2004 reported by Martel, who was able to collect 25 cases of SANT. [2] The occurrence of SANT is very low, with only a few hundred cases worldwide. Although the histological features were described years before in a few case reports by other authors under different names such as splenic hamartoma, cord capillary haemangioma, and multinodular haemangioma, there was no consensus on the nomenclature for this clinicopathological pattern.[7] SANT are benign, nodular vascular proliferations of splenic red pulp with considerable sclerosis. The etiology is still unclear. Numerous theories about the pathogenesis of SANT have evolved. It was been proposed by Martel that SANT represents an unusual transformation of the red pulp in response to an exaggerated stromal proliferation.[2] Diebold suggested that metabolic changes and passive congestion of red pulp leading to damage to sinusoidal endothelial cells culminate in a repair process resulting in the deposition of inflammatory cells, histiocytes, and fibrosis.[8] Chang speculated that it might be associated with vascular insufficiency and the subsequent vascular proliferation for repairing, indicating it's a polyclonal reactive lesion rather than a genuine neoplasm.[9]

According to the literature, central calcification and the "spoke wheel" pattern were usually considered typical manifestations in CT imaging, but these signs were not present in all patients.[10] In our case, CT reveals centripetal enhancement. Differential diagnosis with only pre-operative imaging is difficult because SANT is a rare entity, moreover, there is a realistic risk of rupture in cases of large spleen masses. Recently, Pelizzo reported a case of SANT with spontaneous rupture and intraperitoneal hemorrhage in a 9-week-old female infant. [11] Open or laparoscopic Splenectomy appears to be curative as it is both
therapeutic and diagnostic, avoiding the risk of spontaneous rupture and suspicion of malignancy. \[12\] Whether it is even necessary, especially for asymptomatic and small cases, remains unclear. Awareness of intra and postoperative complications of splenectomy is necessary before surgery is recommended. Splenectomy has a high risk for intra and postoperative hemorrhages. \[13\] The major long-term risk is an overwhelming post-splenectomy infection with an increased risk of fulminant infection. Severe and acute life-threatening cases of SANT are extremely rare. \[11\] Further research is required for clinical and radiological evaluation and to study the course of SANT, whether it is possible to observe a small and asymptomatic vascular lesion or surgical resection is always necessary.

**CONCLUSION**

Unawareness of this recently recognized rare differential of splenic vascular lesion and often misdiagnosed as haemangioma on pre-operative imaging poses a dilemma for surgeons whether to perform splenectomy or observe, more so in small and asymptomatic incidentally detected lesions. Knowledge of clinical and radiological characteristics can be helpful in preoperative diagnosis, avoiding the need for postoperative histological examination. Open or laparoscopic splenectomy is still known to be curative as it is both therapeutic and diagnostic.

**REFERENCES**


