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A Rare Case of Sclerosing Angiomatoid Nodular Transformation of the Spleen Managed with Laparoscopic Splenectomy

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ABSTRACT

Sclerosing angiomatoid nodular transformation (SANT) is a rare, benign splenic lesion characterized by distinct histopathological and immunohistochemical features. It often presents as an incidental finding but can occasionally cause symptoms. Imaging studies like contrast-enhanced CT scans can suggest a diagnosis, but definitive confirmation requires histopathology. Surgical management via laparoscopic splenectomy offers excellent outcomes, with low morbidity and rapid recovery. We present the case of a 22-year-old female with a symptomatic SANT lesion, successfully treated with laparoscopic splenectomy. Postoperative recovery was uneventful, and the patient remains asymptomatic on follow-up.

INTRODUCTION

SANT lesion of spleen has been described in various case reports as different entities and recently in 2004 Martel et al gave the terminology and description of the disease[1]. With uniform terminology and description in place multiple case series and reports have come up describing the lesion and upto 140 cases have been reported [14].

They usually present in 5th -6th decade of life with slight 2:1 female preponderance with most of them being detected incidentally [1]. These are solitary well circumscribed benign lesions of spleen with 'benign' features in imaging and are a part of the differential of benign vascular splenic lesions [9]. Surgical intervention in form of splenectomy is therapeutic and histopathology reveals characteristic architecture. Immunohistochemistry provides the evidence of three types of blood vessels like the normal red pulp of the splenic parenchyma and this immunohistochemical profile along with the histopathological features clinches the definitive diagnosis [5]. This has led to hypothesise the etiopathogensis of this tumor as red pulp of spleen being trapped and having a hamartomatous response in response to a non malignant stromal proliferation [6].

We present a similar case of a young girl who presented with symptomatic benign appearing lesion in the spleen. A preoperative differential of SANT was kept in this patient and she underwent successful laparoscopic splenectomy with

uneventful postoperative recovery. Histopathology and immunohistochemistry confirmed the diagnosis and patient is in regular follow up.

CASE IN MENTION

A 22 year old female presented to asian institute of gastroenterology with insidious onset dull aching , continuous pain in the left upper quadrant of the abdomen for the past 1 month with no radiation, referral, and aggravating factors. No history of trauma, fever, altered bowel habits, early satiety, or vomiting was ellicited. Physical examination was unremarkable and routine blood investigations including complete blood count, liver function tests and renal function tests were within normal limits. Ultrasound abdomen visualised a focal lesion in spleen suspicious of a hemangioma and patient further underwent contrast enhanced CT abdomen **(Figure 1)** which revealed a well defined hypodense , exophytic lesion of 6 X 5.3 cm in the hilar area of spleen extending in the gastrosplenic area showing progressive enhancement with central non enhancing area.

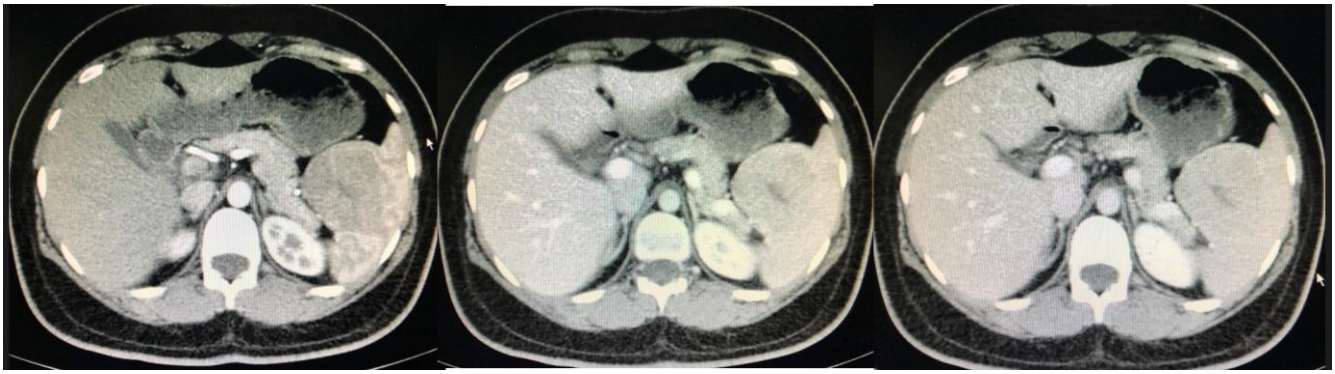

Figure 1 - Contrast enhanced CT abdomen revealing progressively enhancing splenic lesion with central non enhancing area. (Arterial phase, portal phase and venous phase respectively)

Contrast enhanced MRI abdomen revealed a well defined lobulated lesion of 6.3X 6.1 cm in lower pole of spleen which was isointense in both T1 and T2 weighted images with central hyperintense focus in T1 and hypointense in T2 weighted images. The lesion had similar enhancement as that of splenic parenchyma in early and late arterial phases and progressive mild enhancement in extracellular phases. The spleen was normal in size in all of the imaging. A working diagnosis of a benign splenic tumor was kept and the patient was taken up for splenectomy after vaccination ,preanesthetic workup and preoperative optimisation.

Intraoperatively a single mass lesion of 5X5cm at the hilum of spleen with extensive vascularity was found. No evidence of disease spread, either local or distant was evident. Patient underwent laparoscopic splenectomy with a 10mm camera port at the umblicus, 10mm working ports in epigastrium and left lumbar region and 5mm retraction port in the supra-umblical region right of midline **(Figure 2)**. Extraction of the specimen was done through the pfannelstein incision. Cut section revealed a un-encapsulated well circumscribed lesion with multiple peripheral large nodules giving a bosselated appearence with a central stellate scar with radiating septa not reaching upto the periphery **(Figure 3)**.

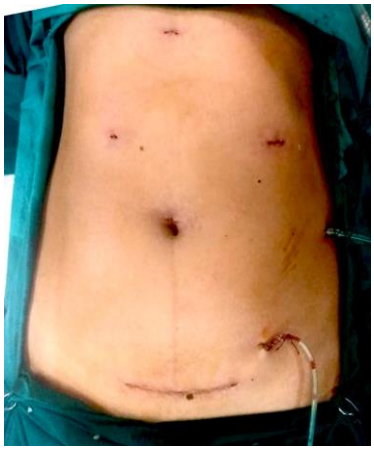

Figure 2- postoperative photograph depicting the port insertion sites,

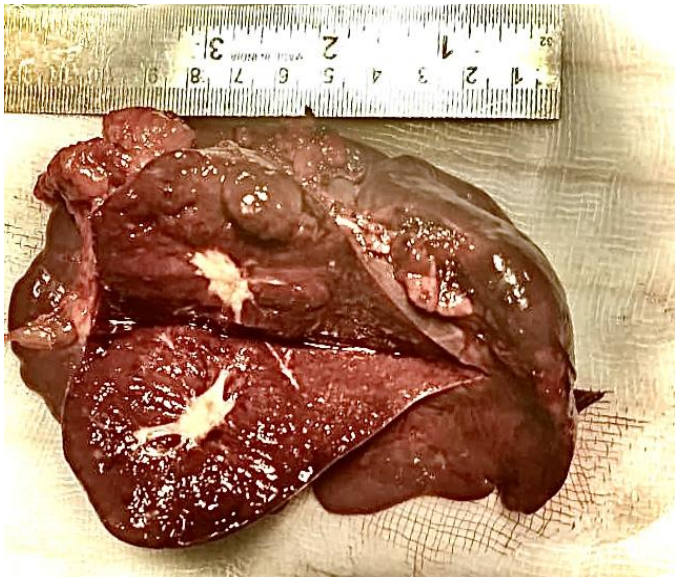

Figure 3- spleen measures ~12 cm and contains ~5.5 cm well circumscribed non encapsulated tumor at the hilar end with reddish brown peripheral nodules arranges around a stellate central scar.

Histopathology revealed a well circumscribed non encapsulated bosselated mass near the hilum of 5X5X4.5cm which is firm in consistency. Cut section show red brown nodules with a central fibrotic scar. Microscopic picture reveals multi nodular pattern of arrangement composed of vascular spaces surrounded by dense hyalanised collagen. The nodules contain vascular spaces of varying sizes lined by endothelial cells with interspersed hemosiderophages. No evidence of atypia in endothelial lining, no mitosis noted, no nuclear atypia or necrosis. Sparse inflammation with few lymphocytes, plasma cells and occasional giant cell reaction were seen. Immunohistochemistry with 4 markers were done and the tissue was SMA positive, with vessels positive for CD34 and CD31. Ki67 was 25% **(Figure 4 and 5).**

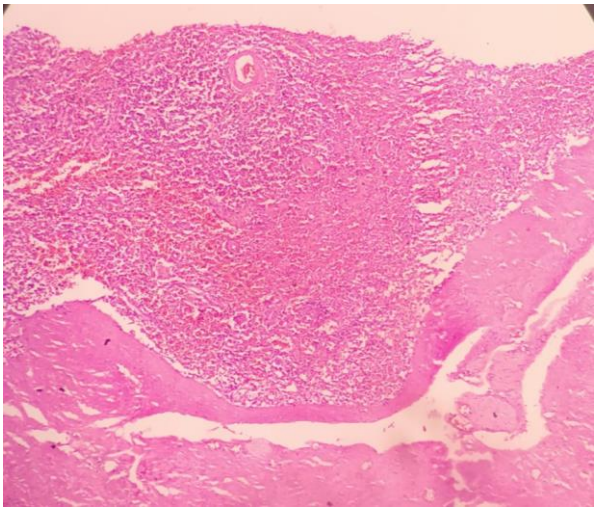

Figure 4- 400X magnification shows incomplete nodule formation with homogenous collagen at the inferior end

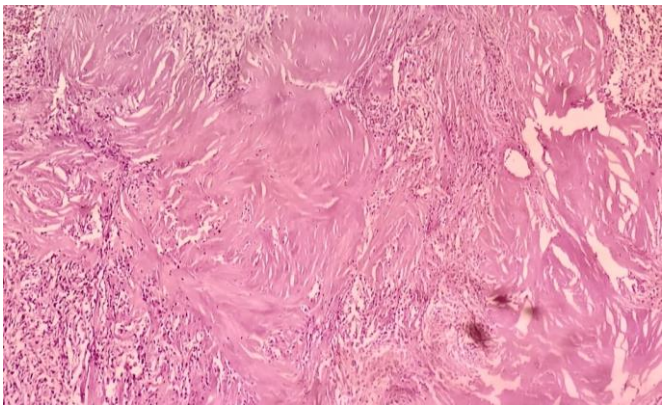

Figure 5- 400X magnification in the stellate scar shows multiple homogenous collagen nodule formation

Postoperatively patient was allowed oral diet on postoperative day 1 and drain was removed on post operative day 3 after measuring drain fluid amylase levels which were within 3 times the upper normal laboratory value. She developed post splenectomy thrombocytosis for which she was started on ecosprin as per the institutional protocols. Patient was asymptomatic at the time of discharge on post operative day 8 with healthy wound and accepting oral diet well with normal bowel functions. On 1month follow up patient is doing well, has no complaints and the wound is healthy.

DISCUSSION

Several unique solitary splenic vascular lesions were described in literature as case reports and termed variously as multinodular hemangioma [4], splenic hemangioendothelioma [3], splenic hemartoma [10] or cord capillary hemangiomas [2]. Martel et al studied 25 such cases and proposed the term sclerosing angiomatoid nodular transformation (SANT) for this unique lesion in 2004 [1]. These lesions are peculiar to spleen and present as a solitary, well circumscribed lesion with female preponderance in 5th - 6th decade of life. They have a typical immunostaining pattern and have no reported recurrence after excision or associated malignant changes to date.They most commonly present as incidental findings

but association with synchronous and metachronous malignancies have been found in upto 20% of the patients. Pain abdomen is the second most common presentation followed by splenomegaly [1,9].

Imaging reveals a solitary well defined non infiltrating 'benign appearing lesion' in spleen. Many cases are detected as a lesion with on ultrasound abdomen with non specific features of a heterogenous well defined hypoechoic mass with internal vascularity on color Doppler [12]. Further characterisation of the tumor requires Contrast enhanced cross sectional imaging like CT or MRI which reveal a characteristic progressive enhancement in the periphery and radially along the density of angiomatoid nodules between the radial fibrous scars sometimes giving a spokewheel appearence [8]. This enhancement is heterogenous in the beginning in the arterial phase starting from the periphery in the angiomatoid tissues but progressing to complete homogenous enhancement of the lesion in the delayed phase when even the fibrous tissue gets filled up with contrast in most but not all cases [9,11]. T2 weighted MRI images reveal a relatively hypointense lesion compared to rest of the splenic parenchyma. The fibrous scar is not always appreciated in the cross sectional imaging [9]. These tumors are FDG PET avid signifying abundance of hypermetabolic cells most probably due to inflammatory cells in the interstitium. But uptake of technetium sulphur colloid indicated absence of reticuloendothelial cells [11].

Grossly they are a well demarcated but un-encapsulated nodular lesions with multiple dark brown nodules with a star shaped central scar with radiating septa [5]. Microscopically these dark brown nodules are angiomatoid lesions consisting of central compressed slit like vascular spaces lined by plump endothelial cells with spindle and ovoid cells lying in the interstitium along with variable inflammatory cells and erythrocytes [1]. These nodules are surrounded by collagen sheets and fibro-sclerotic stroma which forms the central scar and associated septa. Features like necrosis, mitotic figures, or significant nuclear atypia are rare.

Immunohistochemistry reveals the presence of 3 types of blood vessels in the tumor which mimics the normal red pulp of the splenic parenchyma and hence it is been suggested that these tumors are a hamartomatous transformation of splenic red pulp in response to an exagerrated non neoplastic stromal proliferation [6] and thus represent a kind of a inflammatory pseudotumor [7]. These 3 types of blood vessels are (1)Well formed capillaries well organised forming lobular pattern that are CD34 +, CD31+, CD8-, (2) vessels mimiking splenic sinusoids with open channels that areCD34-CD31+CD8+ and (3) small veins arranged in mesh like pattern which are CD34-CD8-CD31+ [1]. These characteristic immunohistochemical staining pattern differentiate these tumors from their more common vascular counterparts namely hemangiomas, littoral cell angiomas, and hamartomas which have a single kind of blood vessel.

Definitive preoperative diagnosis is difficult to achieve and USG guided biopsy although proposed by some authors [12], is not well accepted in view of associated periprocedural risks and unknown malignant nature of the tumor under investigation. Thus splenectomy is well accepted at the diagnostic and therapeutic modality of choice. Most studies quote KI-67 index of around 5% with case reports finding upto 10%. In our patient the KI-67 index was unusually high of 25% but no study to this date have report malignant changes or recurrence. Instances where patient refused surgery are present and follow up imaging after 4-5 months revealed no change in dimensions [13].

CONCLUSION

SANT is a recently described benign entity of the spleen with characteristic imaging features on contrast enhanced cross sectional imaging. Many patients are taken up for surgical intervention with differential diagnosis of more common benign lesions of spleen like hamartomas and hemangiomas but histopathology and subsequent immunohistochemistry establishes the diagnosis. They are benign tumors and no recurrence of malignant changes have been reported to this date although longer follow up is warranted for this newly described lesion in view of occasional reports of high proliferative index and FDG positivity. Role of conservative management has not been proposed till date as definitive preoperative diagnoses is difficult to achieve.

REFERENCES

1. 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; 28:1268–1279.
2. Krishnan J, Danon A, Frizzera D. Use of anti-factor VIII-related antigen (F8) and QBEN10 (CD34) antibodies helps classify the benign vascular lesions of the spleen. Mod Pathol. 1993;6:94A.
3. Kaw YT, Duwaji MS, Knisley RE, et al. Hemangioendothelioma of the spleen. Arch Pathol Lab Med. 1992;116:1079–1082.
4. Rosai J. Rosai and Ackerman 's Surgical Pathology. 9th ed. Edinburgh:Mosby, 2004:2035.
5. Dinesh Pradhan, Sambit K. Mohanty, Sclerosing Angiomatoid Nodular Transformation of the Spleen.Arch Pathol Lab Med 2013;137.
6. Awamleh AA, Perez-Ordonez B. Sclerosing angiomatoid nodular transformation of the spleen. Arch Pathol Lab Med. 2007;131(6):974–978.
7. Diebold J, Le Tourneau A, Marmey B, et al. Is sclerosing angiomatoid nodular transformation (SANT) of the splenic red pulp identical to inflammatory pseudotumor?: report of 16 cases. Histopathology. 2008;53(3):299–310.
8. Karaosmanoglu DA, Karcaaltincaba M, Akata DCT and MRI findings of sclerosing angiomatoid nodular transformation of the spleen: spoke wheel pattern. Korean J Radiol 2008; 9(l):S52–S55.
9. Rachel B Lewis, Grant E, Lattin Jr, Meenakshi Nandedkar, Nadine S. Aguilera. Sclerosing Angiomatoid Nodular Transformation of the Spleen: CT and MRI Features With Pathologic Correlation.AJR:200. 2013
10. Silverman ML, LiVolsi VA. Splenic hamartoma. Am J Clin Pathol. 1978; 70:224–229.
11. Thacker C, Korn R, Millstine J, Harvin H, Van Lier Ribbink JA, Gotway MB. Sclerosing angiomatoid nodular transformation of the spleen: CT, MR, PET, and 99mTc-sulfur colloid SPECT CT findings with gross and histopathological correlation. Abdominal Imaging, 2009;35(6): 683–689.
12. Gutzeit A, Stuckmann G, Dommann-Scherrer C: Sclerosing angiomatoid nodular transformation (SANT) of the spleen: Sonographic finding. J Clin Ultrasound 2009;37: 308-311.
13. 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.
14. Wang TB, Hu BG, Liu DW, Gao ZH, Shi HP, Dong WG. Sclerosing angiomatoid nodular transformation of the spleen: A case report and literature review, 2016;12(2): 928–932.