Review Article

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Sclerosing Angiomatoid Nodular Transformation of the Spleen in Children: A case report and review of the literature

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Introduction

Sclerosingangiomatoid nodular transformation (SANT) is a rare and benign primary vascular lesion of the spleen with of the spleen unknown aetiology and pathogenesis [1]. Its diagnosis is often incidental and it's characterised by numerous angiomatoid nodules in fibrous tissue, within the red pulp of the spleen [2,3]. The neoplasm has shown a predilection for adult females [4].

SANT was first reported by Martin et al. in 2004 as a distinct benign vascular lesion of the spleen, with a well-circumscribed and multinodular angiomatoid appearance, with a characteristic immunostaining pattern, although, prior to this other authors have identified this characteristic lesion of the spleen on a different name such as splenic hamartoma, cord capillary hemangioma, haemangioma-endothelioma, sclerosed haemangioma sand multinodular hemangioma [5,6].

Since it was first described, there has been an increasing knowledge of SANT in the published literature with a number of case reports, case series and reviews, and majority were in the middle aged adult and possible co-existent with other medical condition [7]. Although, SANT has been noted to have a characteristic histology and radiological findings, yet difficult to make a definitive diagnosis without requiring a surgical removal and moreso in children where this lesion is uncommon with unknown incidence and prevalence [8].

In this paper we report a case of SANT in a child and review the literature of all cases published in children less than 18 years old from 2004 till date to better understand its pattern, characteristics and association in children and hopefully enhance early diagnosis and management of SANT in this group of patient with a different physiology.

Case Report

A 14-year-old boy presented to the ophthalmologist for a routine review and was found to have papilloedema. At the time, he was completely asymptomatic and was referred to our tertiary hospital for further evaluation. Apart from a past medical history of previous bruising and short sightedness, he has been otherwise well. Of note significant family history glaucoma On examination, there were of scanty bruises with massive splenomegaly. Other systemic examinations were unremarkable.

Investigations revealed pancytopenia with normal coagulation studies and liver function test. His C-reactive protein was slightly raised at 40-60 range, with a normal erythrocyte sedimentation rate. His lactate dehydrogenase, lipid profile and ferritin were normal. In addition, all his Microbiology investigations, including his extended viral and parasitology screens were all negative.

His serial abdominal ultrasound scans showed an increasing splenic mass from 17cm to 27cm within 4 weeks. Abdominal CT revealed a massive splenomegaly of 26.4cm, with an enhancement pattern within it and the enlarged splenic vein with at least one discrete focal lesion. There are some small perihilar splenic lymph nodes and no radiological evidence of disease elsewhere. A suspicion of primary splenic lymphoma was entertained (**Figure 1,2**). A bone marrow aspiration showed a normocellular marrow with no morphological evidence of malignancy or hemophagocytic lymphohistiocytosis.

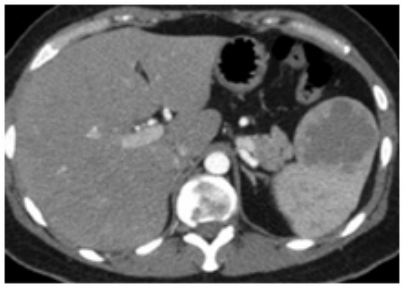

Figure 1: CT image of the spleen showing an enhancement pattern within it and the enlarged splenic vein and at least one discrete focal lesion.

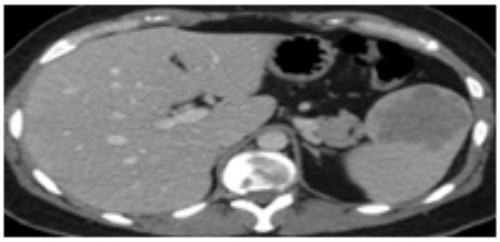

Figure 2: CT image shows predominantly hypodense mass with lobulated contours. Also note rim-style contrast enhancement of external borders of lesion.

The patient underwent splenectomy for diagnostic and therapeutic purposes with normalisation of his full blood count. The gross weight of the spleen was 1012 gram and subsequent histological analysis revealed a prominent, nodular, 15mm mass with slit-like or irregular shaped vascular spaces that were typical of SANT. Immunohistochemistry was positive for CD8, CD31+ and CD34+. We also found CD240 to be positive, however, Stains for S100, Glut 1, S100, CD1A, desmin, Alk 1, HHV8 and CK8/18 were all negative. Lymphoid markers show a normal staining pattern in the splenic parenchyma. (**Figure 3**).

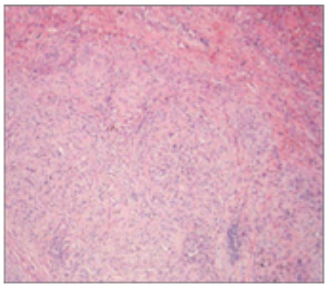

Figure 3: Microscopic image of the excised splenic lesion showing multiple angiomatoid nodules with prominent numbers of macrophages and scattered lymphocytes and plasma cells.

This teenager was also diagnosed with congenital retinoschisis and Scheuermann's disease of the spine. This patient recovered well without any post- operative complications during the post-surgical period, and no evidence of recurrence was observed on a 10 months of follow-up.

Discussion

Our case described a rare finding of SANT in the paediatric population with histological characteristics of slit-like or irregular shaped vascular spaces that was typical of SANT. Immunohistochemistry was positive for CD8-, CD31+ and CD34+, which was in keeping with results found in previous cases [2,9]. In addition, we also found CD240 to be positive. Martel et al. had earlier reported three distinct types of blood vessels similar to the normal composition of splenic red pulp. The first were well-formed cord capillaries in an organized lobular arrangement that were CD34+/CD8−/CD31+. The second type of vessel were consistent with splenic sinusoids and were CD34−/CD8+/CD31+, which now characterized SANT [5].

SANT in children is exceptionally rare and review of the existing literature in children less than 18 years old revealed 4 patients, with a median age of 10.5 years (range: 3-17 years), of which 75% (n=3) males [2,3,10,11]. (**Table1**). This was considered to be a female predominant disease in the adult population, however 3 of the 4 cases (75%) in this review in children less than 18 years were of male preponderance, similar to that reported in multifocal SANT, although our sample size is too small to make any firm conclusion, with the lowest age at diagnosis been 3 years, this further support the rarity of this lesion in children. Evidently, SANT has been regarded as an incidental findings with only very few cases been symptomatic [7,12,13]. In the review by Falk G et al. of 97adults diagnosed with SANT, only 18% (n=18/97) cases presented with abdominal pain, as compared with children where, 50% (n=2/4) presented with abdominal pain [14]. Suggesting that children are more likely to be symptomatic. This could be due to their small abdominal cavity relative to the body mass in children and early onset pain from pressure. In addition, as most cases were just at the time of puberty and rapid growth in children the impact of pubertal hormones is questionable, especially with rapid increase in size of the spleen from 17cm to 27cm within 4 weeks and with the reported case of glucocorticoid concentration following adrenalectomy contributing to rapid growth of SANTin a 37 year old man [15].

Table 1: Clinical characteristics of published SANT cases in children less than 18years from January 2004 to June 2017.

Author	Cases	Age	Sex	Presentation	Findings	Co-exiting condition	Follow up (Months)
Zhang S [10] 2015	1	3	Male	Incidental findings following injury	The lumen of the vessels in the angiomatoid nodules was small and contained small lacuna and sinusoid structures with vein- like and capillary-like vessels	None	20
Kuybulu [3] 2009	1	11	Female	Incidental finding on physical examination	Well circumscribed confluent vascular/angiomatoic nodules with mixed-type inflammatory cells	Short Stature	12
Vyas [11] 2011	1	11	Male	Left flank pain since 2 months	well-circumscribed unencapsulated lesion with bulging cut surface and central fibrotic scar	None	36
Bamboat [2] 2010	1	17	Male	Abdominal pain for 6 months	Lobulated 4 cm fibrotic mass with hemorrhagic areas	None	7

One important dilemma is the possible association with other condition. Two children (including the case), had co-existent of a medical condition, congenital retinoschisis and Scheuermann's disease in the case and a short stature in one of the reviews [3]. Its significance in patient with SANT is yet to be elucidated. However, worrying is the co-existent of malignancy in few cases reported in adults, though SANT is a benign lesion with no reported risk of malignant transformation. It is therefore paramount that clinicians thoroughly evaluate children with SANT for possible co-existent with other diseases [5,6,16].

This case was followed up for 10 months with no concerning features, including all cases in the reviews [2,11]. Although fatality from SANT is rear, 2 cases were reported in middle-aged adult, both had a co-existent of malignancy and the cause of death was due to sepsis and complications of cancer respectively [5].

Conclusion

We identify common features, in the presentation, diagnosis and management of this rare condition in paediatrics, as well as documenting our own unique findings including normal erythrocyte sedimentation rate and CD240. Although uncommon, SANT should be included in the differential diagnosis of children presenting with splenomegaly and a well circumscribed, hypovascular lesion on CT imaging [2,3].

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GO conceptualized and designed the study. GO and IB reviewed the literature, analysed the data, wrote the first draft, and co-ordinated the production of the manuscript. GO had full access to all the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis. All authors were involved in the interpretation of the data and writing the report; all authors approved the final version.

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