

Sclerosing angiomatoid nodular transformation of spleen in a 8-year-old girl

Ammarah Jamal¹, Uzma Siddique², Syed Inam Ur Rehman³, Mustafa Arain⁴, Ahmer Hamid⁵

Abstract

Sclerosing angiomatoid nodular transformation of spleen is a rare, benign vascular lesion with an uncertain pathogenesis. It has been described as a separate entity through specific histopathological characters. It is usually asymptomatic, occurring commonly in adult females. Only a few cases of paediatric cases have been reported which have been commonly symptomatic. This disease has excellent prognosis after splenectomy, which is the only treatment. We report the case of an eight-year-old girl who presented with distended abdomen and history of bleeding from the nose following a road traffic accident. Examination revealed stunted height, decreased weight, tachypnoea, tachycardia, anaemia and a firm, massive spleen. Lab investigations further revealed microcytic anaemia, thrombocytopenia, deranged platelet profile and low vitamin B₁₂ and folate levels. Computed tomography confirmed enlarged spleen. Therefore, a diagnostic biopsy was planned which confirmed sclerosing angiomatoid nodular transformation of spleen. Splenectomy was successfully performed soon after and the child is now healthy with no remissions of previous symptoms.

Keywords: Paediatrics, Oncology, Spleen, Neoplasm, Immunohistochemistry, Child

<https://doi.org/10.5455/JPMA.31769>

Introduction

Sclerosing Angiomatoid Nodular Transformation (SANT) is a rare benign vascular lesion of spleen with an undefined pathogenesis. It was first described in 2004 as a pathologically distinct lesion.¹

Clinically, these patients are often asymptomatic and usually discovered incidentally. SANT has female predisposition, presenting between ages 22 to 77 years,² with only a few paediatric cases.³⁻⁶ Best known treatment for SANT is splenectomy, which has excellent prognosis. We present an exceptional case of an eight-year-old girl

with SANT.

Case Report

An eight-year-old girl presented to us at Civil Hospital, Karachi in October 2016 with a history of uneventful trauma three-and-half-years back, followed, a few months later, by undiagnosed recurrent epistaxis and progressively increasing abdominal distension over the successive years. Episodes of spontaneous epistaxis which recurred every three to four months were relieved variably by ice packs, local pressure and oral tranexamic acid. She once had to be transfused packed red cell and platelets because of severe bleeding and low platelet count. There was no history of bleeding from any other site and past medical as well as family history was otherwise insignificant. On examination her height and weight were found to be below the third centile. In addition, she had tachycardia, tachypnoea and anaemia along with firm splenomegaly, 11.5 cm below left costal margin, moderate hepatomegaly with liver span of 10 cm and single enlarged left axillary lymph node. The possible differential diagnoses considered were storage disorder like Gaucher's disease, myeloproliferative disorders, haemolytic disorders and possibility of benign growth inside the spleen such as hamartoma, haemangioma and lymphangioma.

Investigations revealed severe microcytic hypochromic anaemia and thrombocytopenia, which had been persistent for last three years as well as deranged activated partial thromboplastin time (APTT) and platelet functions. Her reticulocyte count was raised along with vitamin B₁₂ and iron deficiency. Bone marrow trephine showed depleted iron stores, dyserythropoiesis, red cell hyperplasia and megaloblastic changes with hyper segmented neutrophils and no evidence of malignancy or storage disorder. Enzyme assay for Gaucher's (beta-glucosidase level) was normal as was haemoglobin electrophoresis. Important investigations are shown in Table. Computed Tomography (CT) of the abdomen (Figure) revealed massive splenomegaly with a large heterogeneously enhancing area in spleen causing compression and displacement of adjacent structures. The spleen appeared to have smooth and lobulated surface with peripheral

¹⁻²Department of Pediatrics, Civil Hospital, Karachi, Pakistan; ³⁻⁴5th Year Student, Dow University of Health Sciences, Karachi, Pakistan; ⁵Department of Pediatric Oncology, The Indus Hospital, Karachi, Pakistan.

Correspondence: Syed Inam Ur Rehman. Email: inam29694@gmail.com

Table: Lab tests. (N: normal values).

Lab tests	Values
Haemoglobin (g/dl) [n= 12-15]	7
Mean corpuscular volume (fl/ red cell) [n=80-96]	68.7
Hematocrit (%) [n= 37-47]	24.8%
Total lymphocyte count (cells/ μ L) [N=4000-11000]	7700
Neutrophil count (%) [n=40-80]	40
Lymphocyte count (%) [n=20-40]	54
Platelet count (cells/ μ L) [n=150,000-450,000]	115
Reticulocyte (%) [n= 0.5-2.0]	5.2
Prothrombin time (s) [n= 10-14]	12.9
Activated partial thromboplastin time (s) [n= 25-35]	38.5
International normalized ratio [n= 0.8-1.2]	1.23
Iron (μ g/dL) [n= 50-170]	34
Vitamin B ₁₂ (μ g/ml) [n= >200]	102
B- glucosidase enzyme assay (nmol/ml/hr) [n=8.37-37.3]	9

enhancement (Figure: arrowhead) and enhanced lines moving inwards from the periphery (Figure: arrow). Based on CT image and lab test results, Tru-cut biopsy was planned in consultation with interventional radiologists. Histopathological examination revealed large bands of fibrosis surrounding nodules, which varied in shape and contained sinusoids, and small slit-like vessels, lined by endothelial cells containing red blood cells. Patchy lymphoplasmacytic infiltrate and extravasation of blood was also noted. Immunohistochemical studies revealed CD 34+ capillaries, CD 31+ vein and CD 31+/- CD8+ sinusoids. These traits were consistent with SANT. Consequently, open splenectomy was performed which yielded a 13 x 9 cm spleen. This proved to be both diagnostic and therapeutic, with histological findings

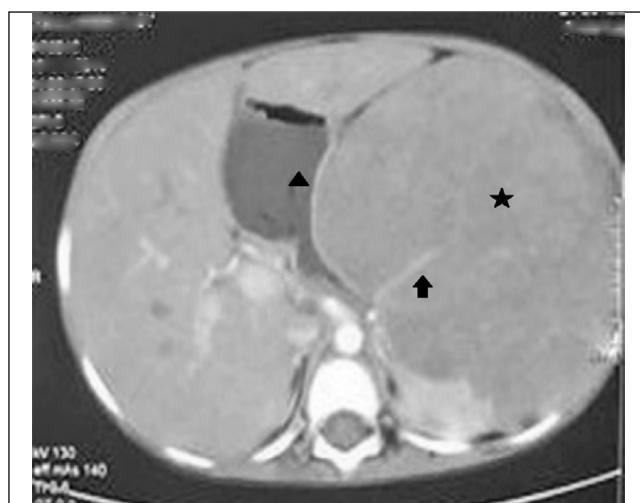


Figure: CT abdomen (axial view). Star: Spleen; Arrow: enhanced line moving inwards from the periphery; Arrowhead: peripheral enhancement.

reconfirming the diagnosis of SANT.

With two years of follow-up, the patient has normal lab parameters and no residual sequela. Oral consent from the mother to publish has been obtained.

Discussion

SANT is a rare benign vascular lesion described as a multinodular lesion composed of endothelium-lined vasculature of varying qualities surrounded by fibrous tissue.¹ Since it was first described as a distinct entity in year 2004, there are no more than 150 cases reported from around the world, including less than five in paediatric age. Although pathogenesis for SANT is unknown, several researchers propose that SANT can be an inflammatory pseudotumour³ or even a vascular malformation.^{5,7} Therefore, SANT is included among the differential diagnoses of non-lymphoid lesions of splenic vasculature.

Most cases of SANT are asymptomatic, diagnosed incidentally during investigations for other associated medical conditions. Children are more often symptomatic than adults, most commonly due to smaller abdominal space.⁸ Patient may present with abdominal mass, pain, raised erythrocyte sedimentation rate, cytopenia, leukocytosis and fever.^{1,8} Our patient conformed to the literature by presenting with abdominal mass and pain along with cytopenia but, in addition, she had certain other clinical and laboratory features which have not been reported in literature yet. These include massive growth of spleen and liver, enlarged axillary lymph node, deranged APTT and platelet functions, increased reticulocyte count and, iron and vitamin B₁₂ deficiency. Iron and vitamin B₁₂ deficiencies can be explained by malnutrition and reticulocytosis may express bone marrow activation in response to anaemia, but we could not justify hepatomegaly, derangement in APTT and platelet functions.

Results of abdominal CT scan in our patient were consistent with the literature, which can show some distinctive features of SANT.⁹ In one study, MRI and CT scan revealed solitary mass with peripheral enhanced radiating lines, with a few patients having rim enhancement of the lesion. This is explained histopathologically by peripheral angiomatoid nodule in the periphery of the lesion which is separated by fibrous columns that radiate outwards from a fibrous focus, characteristically called a "spoke-wheel" appearance.

Macroscopically, SANT's morphology is described as a non-encapsulated and well circumscribed mass,¹⁰ relatively firm as compared to the rest of the spleen with multiple nodules on the surface of the lesion with white tissue in between. Cut section studies commonly show a white fibrous central stellate scar projecting towards the periphery from the centre, with reddish brown nodules in between.¹⁰ Microscopically, the nodules are slit-like or irregularly shaped surrounded by fibrotic stroma. Immunohistochemical staining reveal CD 8 +, CD31 + and CD34 - sinusoids; CD8 -, CD31 + and CD34 + capillaries; and CD8 -, CD31 + and CD34 - veins. These features were identified on Tru-cut biopsy sample taken from our patient. SANT's unique immunohistological profile and lack of atypia make core biopsy a very sensitive and specific test. There are concerns over percutaneous biopsy of the lesion due to a risk of bleeding⁴ but in our experience, it provided prompt diagnosis which reassures that careful planning and a multidisciplinary team can minimise the risk of bleeding in such patients.

This case is one of the few cases of SANT occurring in children with no more than five cases reported earlier.³⁻⁶ In comparison to other cases, our patient presented with multiple symptoms which may have been helpful in ascertaining its pathogenesis. It's possible that early age of presentation of SANT may be a manifestation of more aggressive form of the disease as compared to adults where its detection is usually incidental.⁴ Retarded growth as seen in our patient may point to an inflammatory etiology.^{3,6} Due to limited resources we were not able to assess IgG4+ plasma cell count; however, in other cases it was increased and that may point towards an immunological aetiology. This might have explained chronic lymphadenopathy and chronic low-grade fever present in our patient.

Our case was exceptional considering that the child had a recent history of road traffic accident followed by complaints of recurrent epistaxis, abdominal pain and distension. This varied from normal presentation of SANT which is asymptomatic and incidental. SANT is also said

to have slight female disposition,² which is supported by our case.

Conclusion

This manuscript reports a case of SANT in female child who was diagnosed through percutaneous biopsy and was successfully treated by performing splenectomy.

Disclaimer: None.

Conflict of Interest: The authors declare that they received non-financial support from Genzyme in form of lab test for Gaucher disease.

Funding disclosure: None.

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