J Genet Syndr Gene Ther 2018, Volume 9
DOI: 10.4172/2157-7412-C1-019

Sclerosing Angiomatoid Nodular Transformation of the Spleen: An Unusual finding in Paediatric Population

Godwin Oligbu[1,2], Indula Bopitiya[3], Praveen Saroey[1], Atra Ayad[4,5]
[1]Department of Paediatrics, St Georges Hospital London, UK
[2]Paediatric Infectious Disease Research Group, St Georges University of London, UK
[3]Department of Medicine, Imperial College, London, UK
[4]Department of Paediatric Haematology, St Georges Hospital, London, UK.
[5]Paediatric Oncology, Royal Marsden Hospital, London, UK

Background: Sclerosing angiomatoid nodular transformation (SANT) is a rare and benign primary vascular lesion of the spleen with unknown aetiology and pathogenesis its diagnosis is often incidental and is characterised by numerous angiomatoid nodules, in fibrous tissue, within the red pulp of the spleen. The neoplasm has shown a predilection for adult females with cases in the paediatric population being exceptionally rare.

Case summary: We present an asymptomatic 14-year-old boy with incidental examination findings of scanty bruising and massive splenomegaly. Blood tests revealed pancytopenia with normal coagulation studies, raised C-reactive protein however normal erythrocyte sedimentation rate. Abdominal CT and ultrasound scans showed increasing splenomegaly (the largest size being 27cm) with one discrete focal lesion, small perihilar splenic lymph nodes and an enlarged splenic vein (Figure 1). The patient underwent splenectomy for diagnostic and therapeutic purposes with good long-term outcome and normalisation of the full blood count. Subsequent histological analysis revealed a prominent, nodular, 15mm mass with 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. We also found D240 to be positive. The boy was also diagnosed with congenital retinoschisis and Scheuermann's disease of the spine.

Conclusions & Significance: 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 D240 immunotyping. Although uncommon, SANT should be included in the differential diagnosis of children presenting with splenomegaly and a well circumscribed, hypovascular lesion on CT imaging. Long term follow up of this cohort of adult and paediatric patients is required to better understand the epidemiology of this condition.

godwin.oligbu@nhs.net