CASE REPORT

Sclerosing Angiomatoid Nodular Transformation of Spleen in a 3-year-old Child

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Background: Reports of sclerosing angiomatoid nodular transformation in the pediatric population are rare. ***Case characteristics:*** The 3-year-old child was injured in a car accident and then diagnosed with sclerosing angiomatoid nodular transformation. The patient underwent exploratory surgery with partial splenectomy including total excision of the tumor mass. ***Outcome:*** Neither recurrence nor metastasis occurred over 20 months of follow-up. ***Message:*** Sclerosing angiomatoid nodular transformation can occur in early childhood.

Keywords: *Abdominal pain, Neoplasms, Splenectomy.*

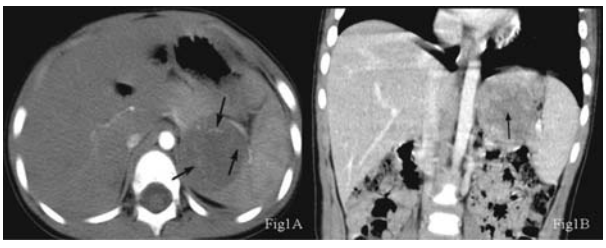
Sclerosing angiomatoid nodular transformation (SANT) of the spleen is a rare benign vascular lesion. Since the first description of this disease by Martel, *et al*. [1] in 2004, several more cases have been described. Most cases of SANT have been incidentally found in adults who were either completely asymptomatic or had vague abdominal complaints [2]. We searched the MEDLINE database and reviewed articles related to SANT published in English between January 1, 2004 and July 31, 2012. Reports of SANT in the pediatric age group are rare, with only three cases reported to date and the youngest case in an 11-year-old patient [3,4]. Herein, we present the pathological and imaging evidence used to diagnose SANT in a 3-year-old child.

Case Report

A 3-year-old boy was admitted to our hospital with complaints of abdominal pain and vomiting 2 hours after being involved in a car accident. On examination, his vital signs were within normal limits. Abdominal examination revealed upper abdominal tenderness with mild muscle spasm. His medical history prior to the car accident and family history were unremarkable.

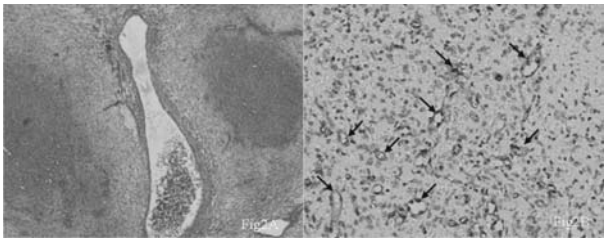
The patient's blood tests, including full blood count, urea and electrolytes, liver and renal functions, were within normal limits. Plain radiograph of abdomen ruled out perforation Because the patient had a closed abdominal injury, suspicion of parenchymatous intra-abdominal organ hemorrhage was maintained and computed tomography (CT) of the abdomen was performed (***Fig.* 1**). Plain CT images revealed a circular lesion with low density and a well-circumscribed border. There was neither a cystic change nor necrosis or calcification in the lesion. Contrast-enhanced CT scans indicated an uneven nodular enhancement on the edge of the lesion and vascular tissue-like bundles from the edge to the enhanced center. In the portal and delayed phases, the lesion enhanced centripetally with a wheel-like appearance. In the delay phase, most of the lesions and spleen parenchyma had similar densities, and a star-like lesion could be partially seen in the center. Carcinoemboroynic antigen and α-feto-protein levels were tested after the detection of tumor mass.

The patient underwent exploratory surgery with partial splenectomy including total excision of the tumor mass. Gross examination revealed a well-demarcated, solitary lesion, measuring 4×5×3 cm. The solitary lesion was connected to the spleen without a capsule and was located on the surface of the spleen with a prominent



(*a*) (*b*)

Fig. 1. (*a*) *The arterial phase showed nodular enhancement around the lesion and many enhanced separations coming from the edge to the center (arrow); (b) The portal vein phase showed a star-shaped, low-density area (arrow) in the center of the mass, which had the appearance of a wheel with spokes.*



(*a*) (*b*)

FIG. 2. (*a*) *The fibrous tissue was loose, including thick-walled blood vessels, but lacked prominent collagen staining (HE staining, ×200); (b) Angiomatoid modules were CD34 positive (HE, ×400).*

nodular and lobulated shape (***Fig.* 2a**). The tumor mass was chiefly composed of connective tissue.

The histopathologic findings revealed that some fibrous tissue had hyaline or myxoid degeneration and some nodules had merged to form irregular patterns. The thick-walled vessels contained myxoid areas with infiltration of inflammatory cells. 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, which were more prominent in the center of the nodule. Plasma cells, eosinophilic leukocytes, and lymphocytes were scattered in the fibrous stroma, primarily infiltrating the junction between the fiber and blood vessels. Small amounts of hemorrhage and hemosiderosis were present in the interstitial tissues and nodules (***Fig.* 2a**). Immuno-histochemical staining of the small blood vessels within the angiomatoid nodules were positive for CD34 (***Fig.* 2b**) and CD31, but negative for glucose transporter 1 (GLUT1), Desmin, S100, neuron-specific enolase (NSE), CD21, anaplastic lymphoma kinase (ALK), and Epstein-Barr virus (EBV). Smooth muscle actin (SMA)-positive venous channels were found in between the epithelioid endothelial cells. Irregular staining of vimentin and SMA was observed in the stroma. Some cells in the lesion were CD68+ and CD8+, and the Ki-67+ percentage was <5%. The observed histomorphology and staining profile supported the diagnosis of SANT.

The patient recovered well without any post-operative complications during the post-surgical period, and no evidence of recurrence was observed during 20 months of follow-up.

DISSCUSSION

Martel, *et al*. [1] named the condition sclerosing angiomatoid nodular transformation (SANT) and reported 25 cases; SANT has since been gradually accepted as a distinct condition. The pathogenesis remains unclear. Some people consider it a bleeding hemangioma, with necrosis, inflammatory pseudotumor nodules, fibrous tissue collagen, and nodules-like angiomatoid [5]. Immunohistochemical staining of specimens from our patient was negative for GLUT1, suggesting SANT to be a vascular malformation rather than a hemangioma. Some researchers also believe that SANT is inflammatory pseudotumor-like [4] and related to EBV infection[6]. The differential diagnosis of SANT includes splenic hamartoma, for which the abnormal structure is mainly composed of splenic red pulp without fibrous tissue dividing capillaries into hemangioma sample nodules. Splenic hemangioma, which mainly consists of capillary or spongy vessels, could appear as focal infarction, hemorrhage, and fibrosis without hemangioma nodule formation and be CD34+, CD31+, and CD8–. In 2010, Hou, *et al*. [5] reported 10 cases. SANT is typically considered more common in female adult patients than in males. A literature search revealed four cases of SANT in children (3 boys and 1 girl). Many patients with SANT are asymptomatic, and usually the condition is identified incidentally during physical examination [7]. About half of SANT patients present with abdominal pain or other non-specific symptoms. Abdominal pain and bloating have been reported in pediatric cases [3,4]. Li, *et al*. [8] first reported CT imaging of SANT, showing the tumors were low-density with calcifications and the condition progressively enhanced. In the delayed phase, the lesion had the same density as the adjacent tissue without a clear boundary between the spleen parenchyma. The differential diagnoses include Gaucher's disease, splenic hamartoma, hemangioma, sarcoid, and a low-grade lymphoma and were difficult to rule out completely before operation.

Asymptomatic patients with benign vascular splenic lesions can be treated with conservative management, because SANT is benign and there has been no report of malignant transformation. In addition, surgical resection or laparoscopic resection of SANT results in a good prognosis. In all reported cases of SANT, surgical resection was both diagnostic and therapeutic. No recurrence or metastasis was observed over a follow-up duration up to 9 years in the 25 cases reported by Martel, *et al*. [1]. With follow-up durations of 7 months to 3 years [3,4], all previously reported pediatric patients remained asymptomatic and healthy. Neither recurrence nor metastasis was observed in the present case over a follow-up of 20 months.

Pediatricians and pediatric surgeons need to be aware about the possibility of SANT occurring in asymptomatic pediatric patients, though rarely.

Contributors: SZ: drafted the manuscript and performed some clinical studies; WY: contributions to the conception and design of the work and diagnosis of the disease HX: performed clinical examinations, especially during the follow-up period; ZW: helped to diagnose this disease.
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