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Multifocal Sclerosing Angiomatoid Nodular Transformation of the Spleen With Splenomegaly and Thrombocytopenia

Chendong He[1], Wei Yang[2,3]

[1]Graduate School of Bengbu Medical College, Bengbu, China
[2]Department of Radiology, Jiangsu Province Hospital of Chinese Medicine, Jiangsu, China
[3]Department of Radiology, Zhejiang Provincial People's Hospital, Hangzhou, China

To the Editor,

Sclerosing angiomatoid nodular transformation (SANT) is a rare but benign vascular disease of the spleen and is usually presented as a solitary mass, and multiple ones are uncommon. Additionally, no obvious clinical symptoms were observed in patients with SANT, and most cases were accidentally found during imaging examinations. Herein, we report an unusual case of multifocal SANT of the spleen with splenomegaly and thrombocytopenia in a 69-year-old female patient. Written informed consent was obtained from the patient.

The female patient presented splenomegaly upon physical examination. Blood routine examination showed a platelet count of 61×10^9/L. Ultrasound examination showed multiple hypoechoic lesions in the spleen and nonhomogeneous internal echo. For further evaluation, a magnetic resonance imaging (MRI, GE Discovery MR750) examination was performed. The abdominal MRI revealed multiple rounded well-defined lesions within the spleen. The largest lesion was approximately 82 × 72 mm in size. The lesions mainly showed peripheral iso-signal on T1-weighted images (T1WI) and T2-weighted images (T2WI), with a slightly increased T2 signal and star-shaped low signal in the center. Compared with out-of-phase imaging, the signal strength of in-phase imaging is lower. The diffusion-weighted image (DWI) with a b-value of 800 s/mm^2 revealed a scattered slightly high signal area and the mixed low signal area. The post-contrast imaging of the lesions showed peripheral enhancement and slightly increased delayed phase diagrams. No obvious enhancement was found in the central stellate part of the lesion (Figure 1. a-g.). Laparoscopic total splenectomy was performed after excluding contraindications. During the surgery, no ascites and no metastatic nodules were found in the abdominal cavity. The spleen was enlarged approximately 16.5 × 12.5 × 7.5 cm in size. Many nodular masses can be seen after the spleen dissection. The lesions were dark-brown in the cross-section, with unclear boundaries, and thickened and grayish-yellow part of the spleen capsule area. Hematoxylin-eosin staining showed that multiple hemangioma-like nodules were separated by fibrous or fibrosclerosing stroma. Collagen fibers and micro-vessels proliferated. Immunohistochemical staining showed three types of blood vessels, namely, dilated sinus vessels showing CD8+/CD31+/CD34−, venules showing CD31+/CD8−/CD34−, and cord capillaries showing CD34+/CD31+/CD8− (Figure 2. a-d.). These immunohistochemical results are the most important points for a clear diagnosis of SANT. The patient was discharged on the 7th postoperative day without any complications, and the computed tomography examination on the 36th postoperative day showed no signs of recurrence. The platelet level returned to average, showing a count of 334×10^9/L.

SANT is a rare benign non-neoplastic proliferative vascular disease of the spleen and was first reported by Aziret et al.[1] in 2004 as a unique pathological entity. Since this initial description, no >250 cases have been reported in English literature. Most patients were female (52.1%), with a median age of 46 years. SANT is usually presented as a single local and well-defined lesion, and multifocal cases were extremely rare. Cao et al.[2] reported that only six multifocal cases were determined up to 2015. Few previous literature works have reported cases of thrombocytopenia in patients with SANT, thus Cao et al.[2] presumed that SANT is the cause of thrombocytopenia because the patient's platelet levels returned to normal level after splenectomy. Additionally, fluctuations in platelet values in our case confirmed this hypothesis. The cause of splenic enlargement is still unclear; however. the authors believe that splenic enlargement may be a compensatory enlargement caused by insufficient hematopoiesis of the spleen.

Imaging examinations is one of the essential methods in SANT diagnosis. MRI evaluation revealed iso-signal to a low signal lesion on T1WI and T2WI, whereas enhanced scan showed peripheral

Corresponding author: Wei Yang, Department of Radiology, Jiangsu Province Hospital of Chinese Medicine, Jiangsu, China
e-mail: youngwei0713@163.com

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ORCID iDs of the authors: C.H. 0000-0002-2216-7245; W.Y. 0000-0002-7216-3808.

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enhancement and low signal intensity in the center of the lesion. This kind of enhancement pattern was described as a "spoke wheel"[3,4], which reflects the central stellate fibrous matrix with fibrous septa that separates the nodules. These typical radiological findings in the present case were consistent with the pathological results of the resected specimens. The low signal of DWI was due to susceptibility artifacts caused by hemosiderin deposition.[5] Low signal intensity on T2WI and the signal intensity on the in-phase image are lower than those on the out-of-phase image, which also indicates hemosiderin deposition.[6] However, histopathological

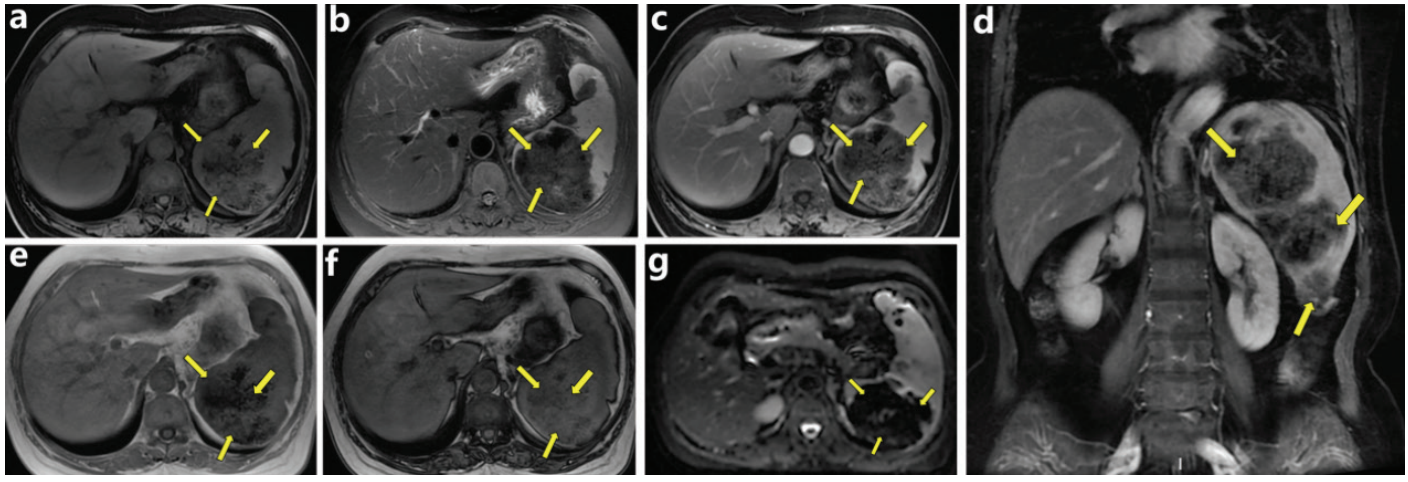

FIG. 1 a-g. MRI finding of the spleen. a-g. Axial T1WI showed heterogeneous isointense and hypointense tumor areas (arrows) (a). Axial T2WI showed a heterogeneous hypointense tumor with a hyper intense central scar (arrows) (b). Axial post-contrast T1 fat-saturated MRI showed peripheral enhancement and lack of central filling (arrows) (c). Coronal post-contrast T1 fat-saturated MRI shows peripheral enhancement (arrows) (d). In-phase axial MRI (e). Out-of-phase axial MRI (f). Compared without-of-phase imaging, the signal strength of in-phase imaging is lower. DWI (b = 800) showed the scattered slightly high signal area and the mixed low signal area (g).

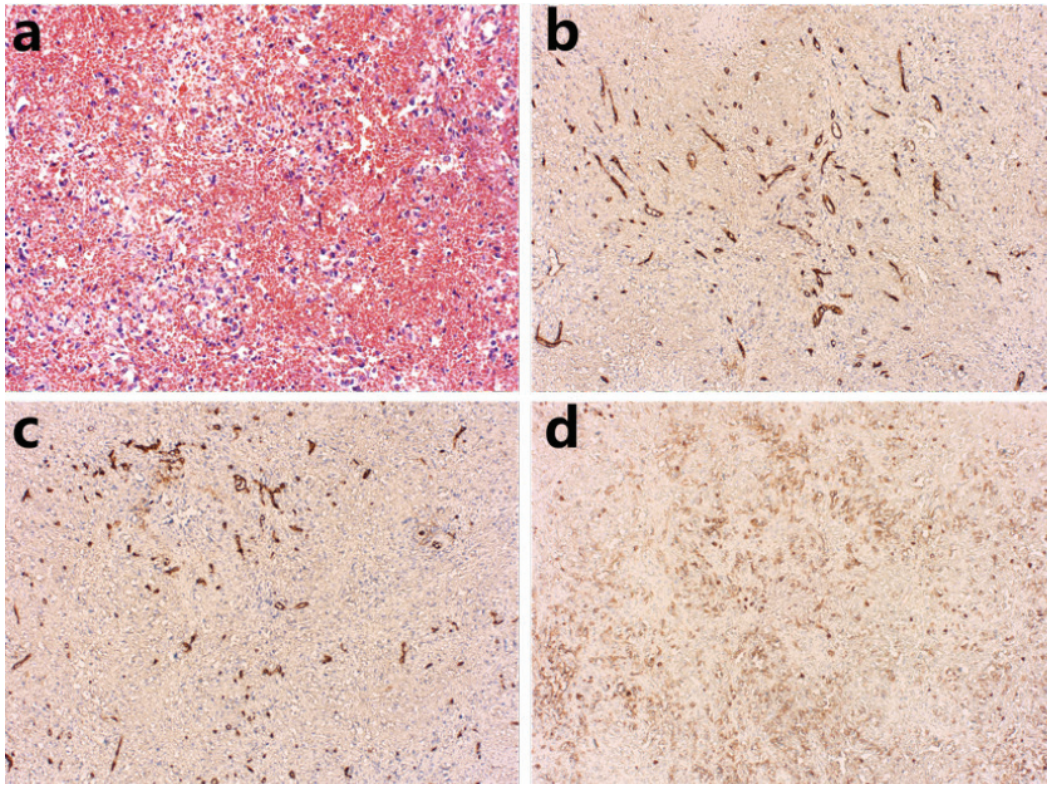

FIG. 2 a-d. Histopathological and immunohistochemical analysis of the mass. a-d. Histopathological findings: Hematoxylin-eosin staining showed that multiple hemangioma-like nodules were separated by fibrous or fibrosclerosing stroma. The proliferation of both collagen fiber and micro-vessels was observed (H&E stain, 200×) (a). Immunohistochemistry staining: different vessels in the nodules had distinct immunophenotypes, CD31 was positive in three blood vessels (b, 40×), CD34 in capillaries (c, 40×), and CD8 in dilated sinusoid like vessels (d, 40×).

findings are still the gold standard for SANT diagnosis. The main differential diagnoses include hemangiomas and littoral cell angiomas, which are composed of a single-type blood vessel rather than multiple types and without the unique SANT structural characteristics. The blood vessels usually express both CD31 and CD34 but do not express CD8. Numerous lesions in the spleen are typical coastal cell hemangioma, which is unique since almost all cases express the histiocytic marker CD68.

In conclusion, the clinical manifestation of SANT lacks specificity. SANT diagnosis can be strongly based on the sign of progressive enhancement with a spoke wheel pattern, but the final diagnosis still needs the support of pathological results. The long-term natural history of SANT is unclear, thus splenectomy may be a suitable technique for current SANT treatment.[1]

Patient Consent for Publication: Written informed consent was obtained from the patient.

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Conflict of Interest: The authors has no conflict of interest to declare.

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