

Spindle cell hemangioma of the spleen

A case report

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Abstract

Rationale: Spindle cell hemangioma (SCH) is considered a benign vascular lesion. It typically develops as a solitary nodule or multiple masses located in the dermal or subcutaneous layers of the distal extremities. To the best of our knowledge, there are no prior reports of SCH in the spleen.

Patient concerns: A 41-year-old male was admitted to our hospital with recurrent headaches, nausea, and vomiting persisting for 5 days. Ultrasound, computed tomography, and magnetic resonance imaging revealed multiple space-occupying lesions in the spleen, and the biggest lesion was 4.8 cm × 5.4 cm in size.

Interventions: The patient underwent laparoscopic splenectomy.

Diagnosis: A diagnosis of spindle cell hemangioma of the spleen was made based on the histopathology.

Outcomes: No evidence of local recurrence or distant metastases was observed over 4-year follow-up.

Lessons: Splenic SCH may exhibit relatively high proliferative activity and be comorbid with epithelioid hemangioendothelioma or angiosarcoma, raising the possibility of malignant potential. However, the patient remained alive and disease-free 4 years after the operation. The nature of SCH in deep soft tissues requires further study.

Abbreviations: CD = cluster of differentiation, CT = computed tomography, SCH = spindle cell hemangioma, SMA = smooth muscle actin.

Keywords: case report, spindle cell hemangioma, spleen, vascular lesions

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Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Written informed consent was obtained from all participants. Ethical approval was obtained from the Ethics Committee of the First Affiliated Hospital, School of Medicine, Zhejiang University, China, in accordance with the ethical guidelines of the 1975 Declaration of Helsinki.

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1. Introduction

Spindle cell hemangioma (SCH), first identified by Weiss and Enzinger in 1986, was initially characterized as a borderline vascular neoplasm and classified as spindle cell hemangioendothelioma.^[1] SCH is now considered a benign vascular tumor or malformation and can occur at any age in males and females. The lesion (s) may develop as a solitary nodule or as multiple masses located in the dermal or subcutaneous layers of the distal extremities.^[2,3] Cases involving deep soft tissues are rare. The present case represents the first documented case of splenic SCH.

2. Case report

A 41-year-old male was admitted to our hospital with recurrent headaches, nausea, and vomiting persisting for 5 days. Cerebral magnetic resonance imaging and electroencephalography did not indicate any abnormalities. Abdominal computed tomography (CT) revealed multiple space-occupying lesions in the spleen. The patient's medical history and physical examination were unremarkable aside from chronic hepatitis B viral infection. Laboratory tests indicated a total serum bilirubin level of 30 μM/L and a direct serum bilirubin level of 11 μM/L. The serum total protein, aminotransferase, and platelet count were within the normal ranges.

Ultrasound examination indicated an irregular hyperechoic area with indistinguishable boundaries in the spleen. Unenhanced CT indicated a large, mixed-density mass in the spleen. On a contrast-enhanced CT scan, the tumor presented a gradual light-to-moderate level of contrast enhancement; the boundary between the tumor and healthy tissues was clear in the portal

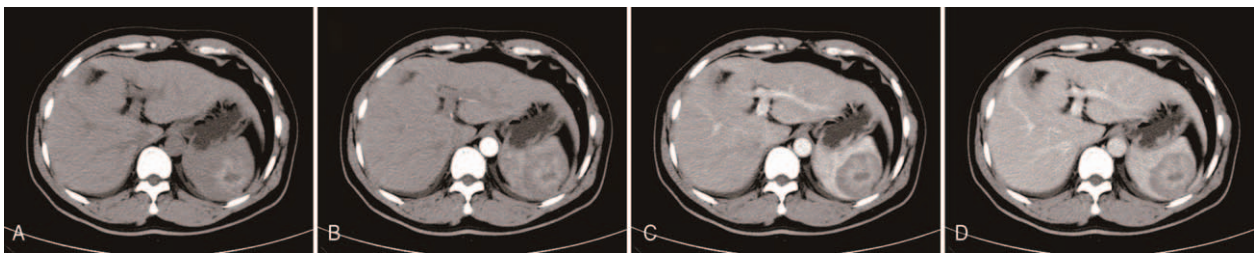


Figure 1. (A) An unenhanced computed tomography (CT) image of a large, mixed-density mass in the spleen. (B–D) On contrast-enhanced CT, the tumor exhibited a light-to-moderate level of contrast enhancement, and the boundary between the tumor and healthy tissues was distinct in the portal and delayed phase.

and delayed phases (Fig. 1). Magnetic resonance imaging revealed abnormal signal density in the splenic parenchyma with heterogeneous hypointensity on T1- and T2-weighted images measuring approximately 4.8 cm × 5.4 cm. Multiple small, round, low-density lesions and splenomegaly were observed on T1-weighted sagittal images (Fig. 2).

The patient was diagnosed with splenic tumors, and laparoscopic splenectomy was performed. During intraoperative exploration, nodular liver cirrhosis, scanty ascites, and gastroesophageal varices were identified. The spleen was swollen to approximately 25 cm × 15 cm × 10 cm, and small white nodules were visible on the surface. The spleen was removed, and

macroscopic examination identified several nodules on the cut surface. Microscopically, spindle cells with slight heterogeneity and mitoses were arranged in an irregular fascicular or mesh pattern. Fissure-like blood vessel lumens lined with flat endothelial cells were observed among the spindle cells (Fig. 3A). A limited number of cells, presumed to be endothelial, with vacuolated cytoplasm were interspersed among the solid cells (Fig. 3B). Cytologic atypia and mitoses were inconspicuous. Immunohistochemistry revealed cluster of differentiation (CD) 34 (Fig. 4A) and CD31 (Fig. 4B) positivity in the endothelial cells lining the vessel spaces, indicating phenotypic differentiation of the vascular endothelium. Immunostaining for CD8, pan-

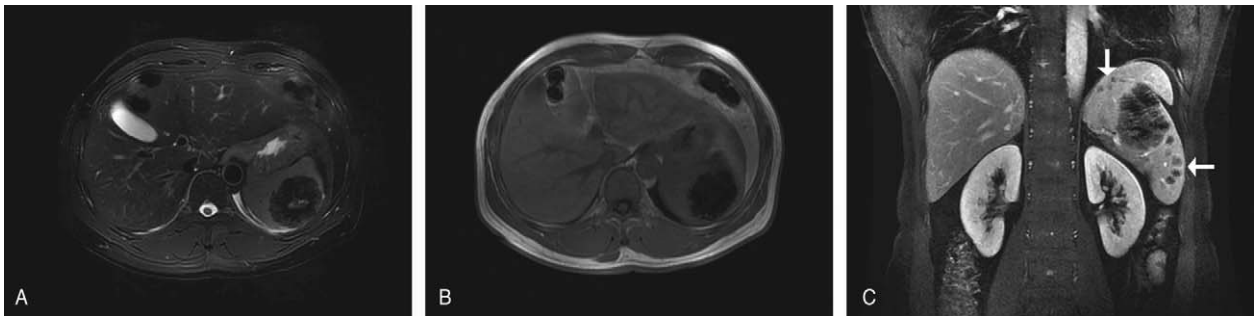


Figure 2. (A and B) Magnetic resonance imaging indicated an abnormal signal density in the splenic parenchyma with heterogeneous hypointensity on T1- and T2-weighted images measuring approximately 4.8 × 5.4 cm. (C) Multiple small, round, low-density lesions (arrow) and splenomegaly were observed on T1-weighted sagittal images.

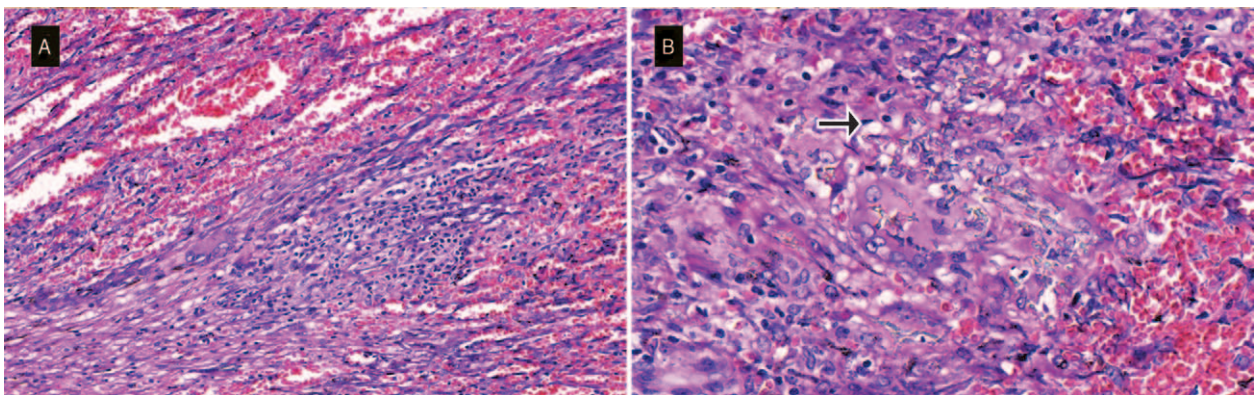


Figure 3. (A) Spindle cells were arranged infiltratively in irregular fascicular or mesh patterns. Fissure-like blood vessel lumens lined with flat endothelial cells were observed among the spindle cells (hematoxylin and eosin staining, original magnification × 200). (B) A few presumed endothelial cells with vacuolated cytoplasm (arrow) were interspersed among the solid cells (hematoxylin and eosin staining, original magnification × 400).

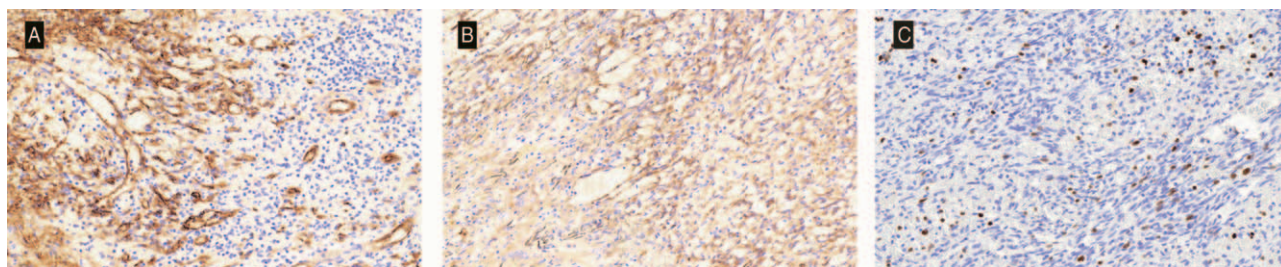


Figure 4. Immunohistochemical analysis revealed CD34 (A) and CD31 (B) positivity in the endothelial cells lining the vessel spaces, suggesting phenotypic differentiation of the vascular endothelium. The spindle cells did not express CD34 (A) or CD31 (B). The proportion of proliferative cells (Ki-67 positive) was 15% (C).

cytokeratin, soluble protein-100, desmin, CD117, epithelial membrane antigen, CD21, and CD23 was negative. The spindle cells showed very focal positivity for smooth muscle actin (SMA) but no expression of CD34 (Fig. 4A) or CD31 (Fig. 4B). The proportion of proliferative cells (Ki-67 positive) was 15% (Fig. 4C). A diagnosis of SCH of the spleen was made based on the histopathology.

The patient was discharged from the hospital 6 days after surgery. No evidence of local recurrence or distant metastases was observed over 4-year follow-up.

3. Discussion

SCH, first identified by Weiss and Enzinger in 1986, was initially believed to be a borderline vascular neoplasm and was classified as spindle cell hemangioma, because regional lymph node metastasis was observed in 1 case and local recurrence in several cases.^[1] Further investigation demonstrated that the observed metastasis resulted from radiation-induced sarcomatous transformation of SCH. Local recurrence was reassessed as multifocal origin or contiguous spread along the vessel. Subsequently, SCH was reclassified as a benign vascular tumor or malformation.^[4]

Histologically, SCH exhibits combined features of nodular Kaposi sarcoma and cavernous hemangioma and was correspondingly divided into 2 groups. One group, composed of extensive spindle cells and focal round endothelial cells (due to vacuolated cytoplasm), consists of solid cellular areas similar to Kaposi sarcoma. The other comprises cavernous vascular areas resembling cavernous hemangioma, consisting of thin-walled dilated vessels. The cavernous vessels are lined with flat endothelial cells and may contain organized phlebolith.^[1,4] Immunohistochemical analysis of the endothelial cells and vacuolated cells revealed expression of endothelial markers including CD34 and factor VIII-related antigen. The spindle cells exhibit divergent positive immunostaining for vimentin, desmin, and SMA.^[5] All SCH lesions showed low expression of the immunohistochemical proliferation markers Ki67 and proliferating cell nuclear antigen, consistent with the indolent clinical course of the tumor.^[6] Cavernous hemangioma and the nodular stage of Kaposi sarcoma should be considered in the differential diagnosis. Cavernous hemangioma is distinguished from SCH by the absence of solid spindle-cell areas. Kaposi sarcoma lacks epithelioid endothelial cells with vacuolization and cavernous vascular structures.^[2,3]

Clinically, SCH is predominantly found in young adults, and strikes both genders with a similar frequency.^[2,3] It tends to occur in the subcutaneous or dermal layers of the distal extremities, especially the hands. Rare cases presenting in

noncutaneous regions have also been described, including the lip, tongue, tonsils, hypopharynx, esophagus, ileum, colon, anal canal, and liver.^[7]

The clinical manifestation of splenic vascular tumors is nonspecific. An accurate diagnosis often requires pathological examination. In this case, the tumor was a typical SCH of the spleen, consistent with the abovementioned pathological characteristics, except for the rate of Ki-67 positivity of 15%. To the best of our knowledge, this is the first documented case of splenic SCH. It is notable that there have been 2 cases of splenic vascular tumor associated with SCH. Suster reported a case of spindle and epithelioid hemangioma of the spleen in a child.^[8] Spindle and epithelioid hemangioma is considered a variant of epithelioid hemangioma.^[9] Fanburg et al reported a low-grade angiosarcoma with scattered SCH-like foci in the spleen, which may represent malignant transformation of SCH to an angiosarcoma.^[10]

In conclusion, splenic SCH may exhibit relatively high proliferative activity and be comorbid with epithelioid hemangioma or angiosarcoma, raising the possibility of malignant potential. However, the patient remained alive and disease-free 4 years after the operation. The nature of SCH in deep soft tissues requires further study.

Author contributions

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