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Single Case

Laparoscopic Splenectomy in a Patient with Sclerosing Angiomatoid Nodular Transformation

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Laparoscopic splenectomy · Sclerosing angiomatoid nodular transformation · Spleen

Abstract
Sclerosing angiomatoid nodular transformation (SANT) of the spleen is a specific, tumor-forming, non-neoplastic, vascular lesion with few reported cases worldwide. Herein, we describe the case of a patient who underwent laparoscopic splenectomy for SANT. A 47-year-old woman underwent upper gastrointestinal endoscopy for suspected gastric submucosal tumor. Contrast-enhanced abdominal computed tomography revealed the presence of a gradually enhancing lesion in the splenic hilum. Although we suspected splenic fibrotic hamartoma, malignancy could not be ruled out. Therefore, the patient underwent laparoscopic splenectomy, resulting in the histopathological diagnosis of SANT. Although SANT is a benign tumor, it may be difficult to obtain definitive diagnosis using preoperative imaging alone. Because the long-term natural history of SANT is unknown, we believe that splenectomy could be an appropriate technique for the diagnosis and treatment of SANT.

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Introduction

Sclerosing angiomatoid nodular transformation (SANT) of the spleen is a specific, tumor-forming, non-neoplastic, vascular lesion. It was first reported by Martel et al. [1] in 2004. They reported that SANT was significantly more common in women than in men, with an average age of 48.4 years. Recently, it has been reported that the ratio of occurrence of SANT in men and women was similar [2].

Since its discovery, there have been less than 200 reported cases in Asia, Europe, and the USA. Reports of cases are increasing, but still few. Several hypotheses for the pathogenesis of SANT have been proposed; however, the pathogenesis of SANT has not been determined yet, and the pathogenesis and long-term natural history of SANT remain unknown.

Patients with SANT sometimes present with abdominal pain; however, most cases have no specific clinical symptoms and are often incidentally found during routine imaging. Because SANT rarely presents characteristic image findings, discriminating it from other splenic tumor imaging is often difficult.

Herein, we describe the case of a patient who underwent laparoscopic splenectomy for SANT that was incidentally discovered during a comprehensive medical examination.

Case Presentation

A 47-year-old woman with no significant medical history and family history underwent upper gastrointestinal endoscopy as part of a comprehensive medical examination for suspected gastric submucosal tumor. Contrast-enhanced abdominal computed tomography (CT) revealed the presence of a gradually enhancing lesion (63 × 56 mm) in the splenic hilum (Fig. 1). Abdominal ultrasonography revealed an irregular hypoechoic lesion in the spleen. Although splenic fibrotic hamartoma was suspected, malignancy could not be ruled out. Since the risk of bleeding and dissemination was present in case of malignant tumor, ultrasound-guided needle biopsy was not recommended. We recommended that magnetic resonance imaging (MRI) and 2-[^{18}F]-fluoro-2-deoxy-D-glucose – positron emission tomography (FDG-PET) should be performed, but the patient refused. We also suggested that she undergo CT several months later, but she chose to undergo splenectomy. Therefore, we performed laparoscopic splenectomy after obtaining patient consent. The surgical procedure was as follows. The patient was administered general anesthesia, positioned in the right semi-lateral position, and 4-port laparoscopy was performed to identify the tumor in the splenic hilum (Fig. 2). We cut the greater omentum, opened the omental bursa, and carefully dissected the gastrosplenic ligament. It was difficult to secure the operative area of the upper spleen due to the tumor in the splenic hilum. However, by dissecting the splenic artery and vein at the lower spleen, the operative area of the upper spleen could be secured. We dissected the splenic artery and vein using an autosuturing device. After putting the spleen in a bag, we made a small incision in the umbilicus and removed it. The procedure duration was 134 min, and limited bleeding was observed. The postoperative course was uneventful, and the patient was discharged from the hospital 8 days after surgery.

The resected spleen was 19.0 × 8.4 × 5.6 cm and weighed 240 g; the splenic hilum was occupied by an unencapsulated nodular lesion having dimensions of 6.2 × 4.6 × 5.1 cm. The lesion was firm and grayish to whitish tan in color (Fig. 2).

Immunohistopathological examination revealed extensive fibrous connective tissue and collagen fiber bundles. Furthermore, a nodular cluster of blood vessels, erythrocytes, and slit-

like blood vessel spaces were observed. Three types of vascular components were found: sinusoid veins (CD8+/CD34–/CD31+), cord capillaries (CD8–/CD34+/CD31+), and small veins (CD8–/CD34–/CD31+), which were intermixed and in high numbers. CD68-positive cells were accumulated at the center of the lesion; however, D2–40-positive cells were absent. Therefore, the lesion was diagnosed as SANT (Fig. 3).

Discussion

SANT of the spleen was first described by Martel et al. [1] in 2004 as a specific, tumor-forming, non-neoplastic, vascular lesion. The characteristics of SANT are as follows: (1) having clear border with surroundings; (2) having multinodular hemangioma-like structures; (3) the presence of collagen fiber hyperplasia that forms the septum between nodules; and (4) the proliferation and coexistence of the three vascular components that make up the red pulp of the spleen, namely, cord capillaries (CD34+/CD8–/CD31+), sinusoids (CD34–/CD8+/CD31+), and small veins (CD34–/CD8–/CD31+).

Although the pathogenesis and long-term natural history of SANT remain unknown, several hypotheses for the pathogenesis of SANT have been proposed, such as association with Epstein-Barr virus infection, transformation of red marrow anomalies due to stromal growth, and the final stage of hamartoma/inflammatory pseudotumor [3–6]. Recent studies have reported that SANT proliferation may be related to immunoglobulin G4 (IgG4) [6, 7]. However, there was no association with IgG4 and Epstein-Barr virus infection in our case.

Martel et al. [1], in a series of 25 cases, have reported that SANT was significantly more common in women than in men, with an average age of 48.4 years. Recently, Cipolla et al. [2], in a series of 152 cases, have reported that the ratio of occurrence of SANT in men and women was similar (1:1.08) and that the average age was 45.4 years. Although patients with SANT sometimes present with abdominal pain, most cases have no specific clinical symptoms and are often incidentally found during routine imaging. Nonetheless, other reported symptoms include vomiting, presence of abdominal mass, cytopenia, abdominal pain, fever, anemia, and weight loss [4, 8, 9].

A radially drawn "spoke-wheel pattern" has been described as a characteristic SANT imaging finding during contrast-enhanced ultrasound and contrast-enhanced MRI [10]. Studies have reported that in the early phase of enhanced CT, the nodule's intensity gradually enhances from the tumor edge to the center and that the inside of the tumor can be radially visualized in the late phase [11–13]. However, it was reported that radial visualization was seen in only 22% cases [11]. Therefore, discriminating it from other splenic tumors via imaging is often difficult. Although there have been reports that SANT can be preoperatively diagnosed via ultrasound-guided needle biopsy [14], this is not commonly practiced because of the risk of bleeding and dissemination, particularly when it is malignant. In our case, although the inside of the nodule was enhanced, it could not be radially visualized in the late phase of enhanced CT. In addition, lack of a proper imaging examination may have made it difficult to obtain a definitive diagnosis. We should have encouraged her to undergo an MRI and FDG-PET.

In a series of 25 cases, Tani et al. [15] reported that there was an enlargement of a tumor in most cases that were followed up (up to 7 years). In all cases, splenectomy was performed for diagnostic purposes because malignancy could not be ruled out. Although there are no reports of postoperative recurrence, the long-term natural history of SANT remains unknown.

In conclusion, the patient was a 47-year-old woman with no clinical symptoms and was diagnosed with SANT based on the presence of the abovementioned three vascular components. Although SANT is a benign tumor, it is difficult to establish a definitive diagnosis via preoperative imaging findings alone. Because the long-term natural history of SANT is unknown, we believe that at present, splenectomy can be an appropriate technique for the diagnosis and treatment of SANT.

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Statement of Ethics

Informed consent was obtained from the patient for publication of the case report and accompanying images. Ethics approval was not needed for this paper.

Disclosure Statement

The authors have no conflicts of interest to disclose.

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Author Contributions

T. Kusano drafted the manuscript and is the surgeon who operated on the patient. C. Ryu and T. Matsuo supervised the editing of the manuscript. T. Kusano and H. Hayashi diagnosed the SANT of the spleen in this paper. All authors read and approved the final manuscript.

References

1 Martel M, Cheuk W, Lombardi L, Lifschitz-Mercer B, Chan JK, Rosai J. Sclerosing angiomatoid nodular transformation (SANT): report of 25 cases of a distinctive benign splenic lesion. Am J Surg Pathol. 2004 Oct;28(10):1268–79.

2 Cipolla C, Florena AM, Ferrara G, Di Gregorio R, Unti E, Giannone AG, et al. Sclerosing angiomatoid nodular transformation: laparoscopic splenectomy as therapeutic and diagnostic approach at the same time. Case Rep Surg. 2018 May;2018:7020538.

3 Weinreb I, Bailey D, Battaglia D, Kennedy M, Perez-Ordoñez B. CD30 and Epstein-Barr virus RNA expression in sclerosing angiomatoid nodular transformation of spleen. Virchows Arch. 2007 Jul;451(1):73–9.

4 Murthy V, Miller B, Nikolousis EM, Pratt G, Rudzki Z. Sclerosing angiomatoid nodular transformation of the spleen. Clin Case Rep. 2015 Oct;3(10):888–90.

5 Cafferata B, Pizzi M, D'Amico F, Mescoli C, Alaggio R. Sclerosing angiomatoid nodular transformation of the spleen, focal nodular hyperplasia and hemangioma of the liver: a tale of three lesions. Pathol Res Pract. 2016 Sep;212(9):855–8.

6 Kashiwagi S, Kumasaka T, Bunsei N, Fukumura Y, Yamasaki S, Abe K, et al. Detection of Epstein-Barr virus-encoded small RNA-expressed myofibroblasts and IgG4-producing plasma cells in sclerosing angiomatoid nodular transformation of the spleen. Virchows Arch. 2008 Sep;453(3):275–82.
7 Nagai Y, Satoh D, Matsukawa H, Shiozaki S. Sclerosing angiomatoid nodular transformation of the spleen presenting rapid growth after adrenalectomy: report of a case. Int J Surg Case Rep. 2017;30:108–11.
8 Corrado G, Tabanelli V, Biffi R, Petralia G, Tinelli A, Peccatori FA. Sclerosing angiomatoid nodular transformation of the spleen during pregnancy: diagnostic challenges and clinical management. J Obstet Gynaecol Res. 2016 Aug;42(8):1021–5.
9 Kakisaka T, Kamiyama T, Yokoo H, Orimo T, Wakayama K, Tsuruga Y, et al. Hand-assisted laparoscopic splenectomy for sclerosing angiomatoid nodular transformation of the spleen complicated by chronic disseminated intravascular coagulation: a case report. Asian J Endosc Surg. 2014 Aug;7(3):275–8.
10 Thacker C, Korn R, Millstine J, Harvin H, Van Lier Ribbink JA, Gotway MB. Sclerosing angiomatoid nodular transformation of the spleen: CT, MR, PET, and ^{99}(m)Tc-sulfur colloid SPECT CT findings with gross and histopathological correlation. Abdom Imaging. 2010 Dec;35(6):683–9.
11 Lewis RB, Lattin GE Jr, Nandedkar M, Aguilera NS. Sclerosing angiomatoid nodular transformation of the spleen: CT and MRI features with pathologic correlation. AJR Am J Roentgenol. 2013 Apr;200(4):W353-60.
12 Raman SP, Singhi A, Horton KM, Hruban RH, Fishman EK. Sclerosing angiomatoid nodular transformation of the spleen (SANT): multimodality imaging appearance of five cases with radiology-pathology correlation. Abdom Imaging. 2013 Aug;38(4):827–34.
13 Subhawong TK, Subhawong AP, Kamel I. Sclerosing angiomatoid nodular transformation of the spleen: multimodality imaging findings and pathologic correlate. J Comput Assist Tomogr. 2010 Mar-Apr;34(2):206–9.
14 Gutzeit A, Stuckmann G, Dommann-Scherrer C. Sclerosing angiomatoid nodular transformation (SANT) of the spleen: sonographic finding. J Clin Ultrasound. 2009 Jun;37(5):308–11.
15 Tani M, Yamamoto H, Yamada M, Yamamoto M, Harada H, Zaima M. Laparoscopic splenectomy in a patient with sclerosing angiomatoid nodular transformation (SANT). J Jpn Surg Assoc. 2018;79(6):1299–304. DOI: 10.3919/jjsa.79.1299. Japanese.

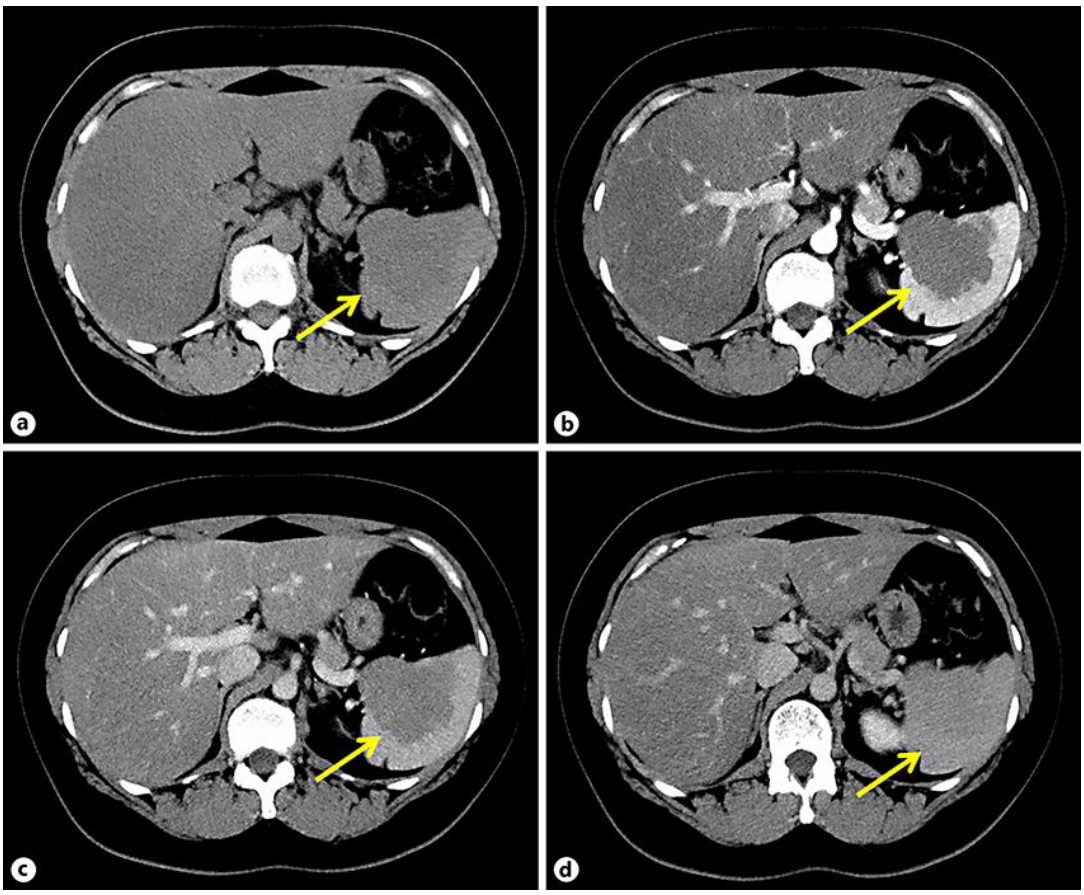

Fig. 1. Dynamic computed tomography of the spleen. A gradually enhancing mass (65 × 65 × 55 mm) is observed in the spleen (arrow). No radial visualization of the inside of the tumor is observed. **a** Simple phase. **b** Arterial phase. **c** Portal vein phase. **d** Equilibrium phase.

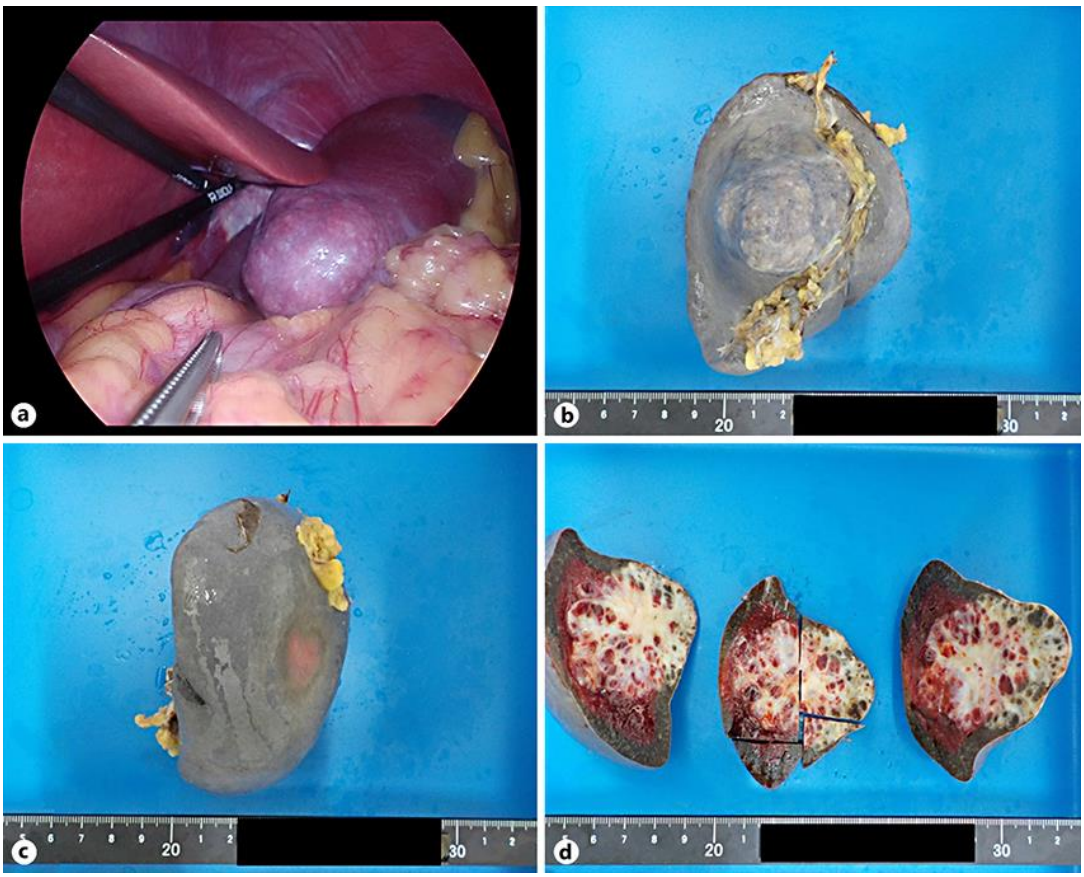

Fig. 2. Laparoscopic and macroscopic findings of spleen. **a** A swollen tumor is observed at the splenic hilum with no gastric invasion of the tumor. The tumor was grayish in color. **b** A swollen tumor is observed at the splenic hilum, and there is no tumor exposure. **c** There is no tumor exposure on the dorsal side of the spleen. **d** In this section of the spleen, a well-defined grayish tumor is observed.

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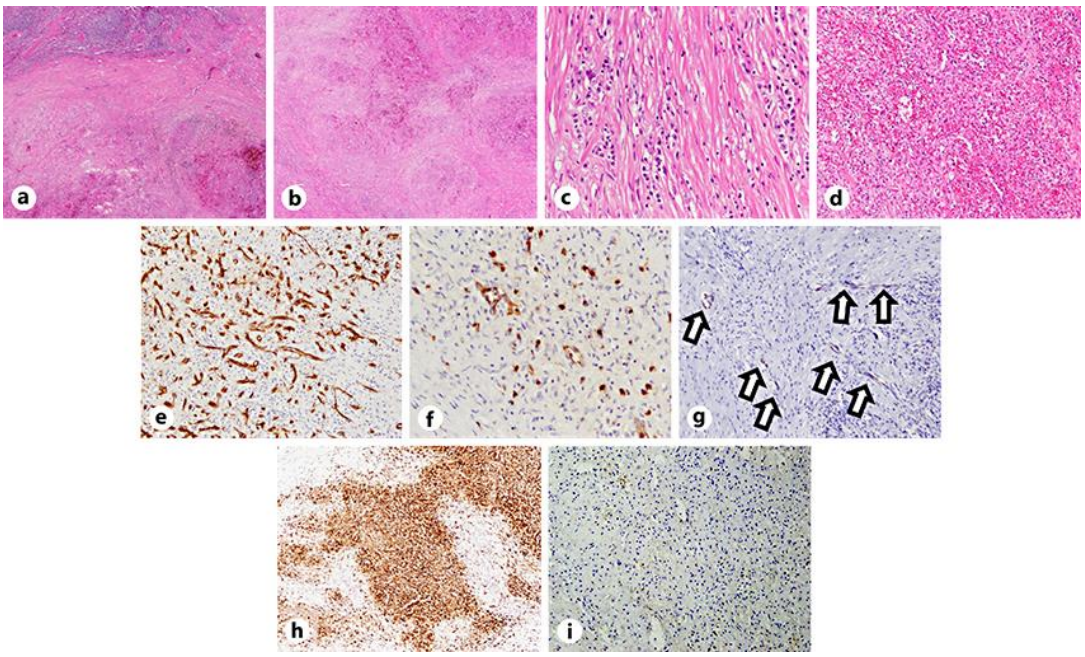

Fig. 3. Histopathology and immunohistochemistry of splenic tumor tissue. **a** Nodular lesions are covered with thick bundles of collagen fibers. Magnification, ×20 . **b** Multinodular structure of SANT. Magnification, ×20. **c** Infiltration of plasma cells in the thick collagen fibers. Magnification, ×200. **d** The slit-like blood vessels, spindle cells, and circular cells are mixed and shown proliferation in the nodule. Magnification, ×100. **e** Capillary blood vessels positive for CD34 are observed under a microscope. Magnification, ×100. **f** A sinusoid positive for CD8 is observed under a microscope. Magnification, ×100. **g** A vein positive for CD31in fibrotic tissue is observed under a microscope (arrows). Magnification, ×100. **h** CD68-positive cells were accumulated at the center of the lesion. Magnification, ×40. **i** D2–40-positive cells were absent at the center of the lesion. Magnification, ×100.