Sclerosing angiomatoid nodular transformation of the accessory spleen
A case report and review of literature
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Abstract
Rationale: Sclerosing angiomatoid nodular transformation (SANT) is a rare case which usually occurs in spleen, but our case occurs in accessory spleen.

Patient concerns: We reported a case of a 27 year old man who was identified with an isodense mass in the left abdominal cavity by ultrasonography without symptoms. Blood routine and tumor markers (alpha fetal protein, carcinoembryonic antigen, CA19-9, CA125) were normal.

Diagnoses: The tumor was firstly diagnosed as left abdominal mass and its origin was considered as small intestinal by the radiologist. The pathological diagnosis was the sclerosing angiomatoid nodular transformation (SANT) of the accessory spleen.

Interventions: Laparoscopic splenectomy is a more reliable method for the treatment of SANT.

Outcomes: Non-enhanced CT showed iso-intensity lesion of the accessory spleen, and enhanced CT showed progressive enhancement. According to the above characteristics, we should consider the diagnosis of spleen SANT. We reviewed some literatures to summarize the characteristics of SANT.

Abbreviations: CT = computed tomography, Hu = Hounsfield unit, MR = magnetic resonance, MRI = magnetic resonance imaging, SANT = sclerosing angiomatoid nodular transformation.

Keywords: accessory spleen, computed tomography, sclerosing angiomatoid nodular transformation.

1. Introduction
Sclerosing angiomatoid nodular transformation (SANT) is a kind of rare benign tumor that occurs in the spleen. Martel et al. [1] first described the disease of SANT in 2004. However, SANT was only a concept of pathological diagnosis, in clinical, the real nature of SANT was not full understood. [2] The pathogenesis of SANT also remained unclear. [3] Chang et al. [4] concluded that SANT was considered as a polyclonal and reactive lesion rather than a neoplasm. Most literatures have provided several information about SANT in spleen, while no literature has reported SANT occurring on accessory spleen. In our case, we report a rare case of SANT that occurs in the accessory spleen.

2. Case report
A 27-year-old male was found with an isodense mass in the left abdominal cavity by abdominal ultrasonography during physical examination. He was asymptomatic and blood routine, tumor markers (alpha fetal protein, carcinoembryonic antigen, CA19-9, CA125) were normal. Subsequently, he underwent an abdominal computed tomography (CT) scan. On the abdominal nonenhanced CT scan, a round isodense tumor with calcification was seen in the right front of left kidney and left front of spleen, and it possessed of the size of 5.9 cm x 5.4 cm and CT value of 38 Hounsfield unit (Hu). The boundary between the lesion and the tail of pancreas, left kidney, and spleen was still clear (Fig. 1A). On contrast-enhanced CT, the lesion was slightly and progressively enhanced (CT values of the arterial, venous, and delayed phases were about 51, 55, 66 Hu, respectively) and its density was lower than that of spleen at the same phases (CT values of the arterial, venous, and delayed phases were about 73–109, 94–107, 70–76 Hu, respectively) (Fig. 1B–D). Moreover, we could find an enhanced edge on contrast-enhanced CT (CT values of the arterial, venous, and delayed phases were about 71, 72, 76 Hu, respectively) (Fig. 1B: red arrow). The tumor was first diagnosed as left abdominal mass and its origin was considered as small intestinal by the radiologist.

2.1. Surgical and pathological findings
The electric knife cut the skin into the abdomen, and liver, gallbladder, spleen were normal. During the operation, the mass was located in the lower pole of the spleen measuring 6.0 cm x 6.0 cm with complete capsule. Later, the mass was resected by ultrasonic knife. The mass was off-white and gray red, medium texture. Lesion consisted of spindle cells, which was arranged as
nodules. There were splenic sinus structures around the nodules and sinus-like blood vessels in the central of nodules. The pathological diagnosis was the SANT of the accessory spleen (Fig. 2).

3. Discussion
SANT is a benign tumor of the spleen and usually occurs in middle-aged female. SANT is usually asymptomatic or just along with abdominal pain. In our case, the patient was a young male without any symptom and the primary site was accessory spleen. Accessory spleen is relatively common, which is known as healthy spleen tissue isolated from spleen and can be seen in 10% to 30% of patients at autopsy. It is clinically important in some patients, although usually asymptomatic and incidentally discovered. Previous reports about SANT all occurred in the spleen. Our article summarized the characteristics of some SANTs of spleen in Table 1. Nonenhanced CT or magnetic resonance (MR) scan show a round mass with hypodensity and clear boundary. On enhanced CT or MR scan, many SANT lesions are exhibited as a “spoked wheel” mode. This “spoked wheel” has been shown on detailed radiological and pathological findings, which is equivalent to the central stellate fiber interstitial fibrillar nodules isolated nodules. Karaosmanoglu et al described the SANT’s magnetic resonance imaging (MRI) findings at first. They mentioned in their article that the “spoked wheel” images of lesions on T2-weighted images were similar to CT multiphasic imaging. Subsequently, some studies have suggested that the MRI features and multiphase CT imaging of “spoked wheel” of SANT may be useful for the diagnosis of SANT. Our patient only underwent CT examination, which showed progressive enhancement, although it
### Table 1
Some SANT characteristics.

| Ref. | Age, y/Gender | CT | MR | Cl. Feature | Blood routine | Treatment | Follow-up/recurrence |
|------|---------------|----|----|-------------|---------------|-----------|---------------------|
| Atas et al [5] | 33/M | – | – | Equal | Normal | Splenectomy | 1 y/No |
| Huang et al | 27/M | Hypodense | – | – | – | Laparoscopic splenectomy | – |
| Demirci et al | 43/F | Hypodense | Inhomogeneous | Hyperdense | Mildly hypodense | – | – |
| Bushati et al | 46/F | – | – | Hypodense | Mildly hypodense | – | – |
| Saito et al | 53/F | – | – | – | Hypodense | Progressive | – |
| Martinez Martinez et al, 2017 | 57/F | Hypodense | – | – | – | – | – |
| Nagai et al [3] | 37/M | Hypodense | Progressive | Hypodense | Inhomogeneous hyperdense | – | – |
| Tian-Bao Wang et al, 2016 | 27M | Hypodense | Progressive | Hypodense | Inhomogeneous hyperdense | – | – |
| Murthy et al [13] | 56/M | Hypodense | – | – | – | Laparoscopic splenectomy | – |
| Cao et al [2] | 38/M | Hypodense | Progressive | Hypodense | Progressive | – | – |
| Hock Tai Gavin Lim et al, 2015 | 39/M | Hypodense | Homogeneous | Equal | Mildly hypodense | Progressive | – |

CT = computed tomography, MR = magnetic resonance.

Size, cm | Cl. Feature | Blood routine | Treatment | Follow-up/recurrence |
|-----------|-------------|---------------|-----------|---------------------|
| 8 | Intermittent abdominal pain for 1 y | Normal | Splenectomy | 1 y/No |
| 4 | Maffucci syndrome, 4 years of recurrent abdominal pain, | Normal | Laparoscopic splenectomy | – |
| 2 | Having a superficial spreading malignant melanoma localized at his left instep 7 years ago | Normal | Laparoscopic splenectomy | – |
| 6 | Abdominal discomfort | Mild anemia | Laparoscopic splenectomy | – |
| 4 | Occasionally found | Normal | Laparoscopic partial splenectomy | – |
| 7 | Heartburn and a 1-month history of epigastric abdominal pain and left hypochondrium pain | Normal | Laparoscopic splenectomy | – |
| 2 | Primary hyperaldosteronism shows rapid growth after adrenalectomy | Normal | Laparoscopic splenectomy | – |
| 10 | Abdominal pain | Mild anemia | Laparoscopic splenectomy | 2 y/No |
| 3.5/5.5/7.6 | Left upper quadrant pain and weakness | Elevated gamma-glutamyl transpeptidase and alanine transaminase levels | Laparoscopic splenectomy | – |
| 4 | Occasionally found | Elevated gamma-glutamyl transpeptidase and alanine transaminase levels | Laparoscopic splenectomy | – |

Figure 3. (A) 3D imaging, (B) 2D imaging: The main artery of the tumor was from the splenic artery.
had not yet reached the obvious “spoked wheel” pattern. Furthermore, calcification was also observed in previous literature, which is consistent with our case.[13] In addition, according to the patient’s CT angiography images (Fig. 3A, B), we can find that the main feeding artery of the lesion mainly comes from the splenic artery, and it is generally known that the feeding artery of the spleen is the splenic artery, which can provide a diagnostic idea. However, the final diagnosis is still immunohistochemistry but not imaging. The histomorphology and staining profile taken together finally confirmed the diagnosis of SANT.[14] So far, laparoscopic splenectomy is a more reliable method for the treatment of SANT.[15] We summarize the case reports in Table 1; most used splenectomy or laparoscopic splenectomy, and the prognosis, follow-up are well without recurrence.

4. Conclusion

On the basis of these characteristics of CT, we conclude that if we find an isodense lesion of the accessory spleen on nonenhanced CT, and show progressive enhancement on enhanced CT, meantime, the main nutritional artery is the splenic artery, diagnosis of SANT in accessory spleen should be taken into account. To summarize, we think that our report may constitute a significant step forward for diagnosis of SANT in accessory spleen and it may provide more therapeutic and prognostic information for clinical management.

Author contributions

Writing – original draft: Miao Niu, Qinhe Zhang.
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References

[1] Martel M, Cheuk W, Lombardi L, et al. Sclerosing angiomatoid nodular transformation (SANT): a distinctive benign splenic lesion. Am J Surg Pathol 2004;28:1268–79.
[2] Cao Z, Wang Q, Li J, et al. Multifocal sclerosing angiomatoid nodular transformation of the spleen: a case report and review of literature. Diagn Pathol 2015;10:95.
[3] Nagai Y, Satoh D, Matsuoka H, et al. 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.
[4] Chang KC, Lee JC, Wang YC, et al. Polyclonality in sclerosing angiomatoid nodular transformation of the spleen. Am J Surg Pathol 2016;40:1343–51.
[5] Azas H, Bulus H, Akkurt G. Sclerosing angiomatoid nodular transformation of the spleen: an uncommon cause of abdominal pain. Euroasian J Hepatogastroenterol 2017;7:89–91.
[6] Morele KJ, Morote B, Silverman SG. CT features of the accessory spleen. AJR Am J Roentgenol 2004;183:1653–7.
[7] Karaozumoglu DA, Karcaaslanbca M, Akata D. CT and MRI findings of sclerosing angiomatoid nodular transformation of the spleen: spoke wheel pattern. Korean J Radiol 2008;9(suppl):S52–5.
[8] Lewis RB, Lattin GE Jr, Nandedkar M, et al. Sclerosing angiomatoid nodular transformation of the spleen: CT and MRI features with pathologic correlation. AJR Am J Roentgenol 2013;200:W353–60.
[9] Lee D, Wood B, Formby M, et al. F-18 FDG-avid sclerosing angiomatoid nodular transformation (SANT) of the spleen: case study and literature review. Pathology 2007;39:181–3.
[10] Bampot ZM, Masiakos PT. Sclerosing angiomatoid nodular transformation of the spleen in an adolescent with chronic abdominal pain. J Pediatr Surg 2010;45:E3–6.
[11] Chikkappa MG, Morrison C, Lowe A, et al. Case report and magnetic resonance images of sclerosing angiomatoid nodular transformation (SANT) of the spleen. BMJ Case Rep 2009;2009:pii: bcr07.2009.2131.
[12] Metin MR, Evrimler S, Cey N, et al. An unusual case of sclerosingangiomatoid nodular transformation: radiological and histopathological analyses. Turk J Med Sci 2014;44:530–3.
[13] Murphy V, Miller B, Nikoloussis EM, et al. Sclerosing angiomatoidnodular transformation of the spleen. Clin Case Rep 2015;3:888–90.
[14] Wang HL, Li KW, Wang J. Sclerosing angiomatoid nodular transformation of the spleen: report of five cases and review of literature. Chin Med J (Engl) 2012;125:2386–9.
[15] Kim KH, Lee S, Youn SH, et al. Laparoscopic splenectomy for sclerosing angiomatoid nodular transformation of the spleen. J Korean Surg Soc 2011;80(suppl 1):S59–62.