

Sclectrosing Angiomatoid Nodular Transformation of the Spleen, a Rare Cause for Laparoscopic Splenectomy: a Case Report and Literature Review

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Case report

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Abstract

BACKGROUND: Sclerosing angiomatoid nodular transformation of the spleen is one kind of exceedingly rare benign lesions originated from spleen. The incidence and pathogen remain unclear partially due to its rarity and short history. Consequently, preoperative diagnosis become extremely perplex. Splenectomy is a predominated therapy approach as well as diagnosis method for sclerosing angiomatoid nodular transformation of the spleen. We reported a case underwent computed tomography scan, magnetic resonance imaging and ultrasonic scan preoperatively. Examinations conclusion highly suspected sclerosing angiomatoid nodular transformation of the spleen and was confirmed by pathology after laparoscopic splenectomy. We herein reviewed some literatures on imaging characters of to reveal the possibility of preoperative diagnosis when clinicians encounter and investigated whether laparoscopic splenectomy is an appropriate treatment.

CASE PRESENTATION: A 50-year-old female was found with a low dense mass in the spleen by abdominal computed tomography incidentally without any positive symptom. Further enhanced computed tomography scan revealed the lesion with asynchronous enhancement and magnetic resonance imaging showed radio-like rift missing from central. Ultrasonic scan imaging presented hypoechoic gap in the center, pretty similar to magnetic resonance imaging. These radiology imaging features deduced to Sclerosing angiomatoid nodular transformation of the spleen. Laparoscopic splenectomy was performed successfully by our team and histopathological findings convinced the preoperative diagnosis. Gross inspection was coincident to the performance of magnetic resonance imaging.

CONCLUSION: By comprehending imaging character profoundly, sclerosing angiomatoid nodular transformation of the spleen can be a differential diagnosis preoperatively when encounter lesion of spleen. We believe that laparoscopic splenectomy is a reliable and effective therapy option for such rare benign lesion of spleen without risk of recurrence.

Background

Sclerosing angiomatoid nodular transformation (SANT) is a rare benign lesion originated in the spleen with unclear pathogen. It's a very "young" lesion in spleen because of its short history. Martel et al. firstly bring the concept of SANT in the medicine by summarized 25 cases pathology characteristic of SANT in 2004 [1]. With only several hundred cases of SANT have been reported up-to-date, most of physicians, surgeons, and radiologists still lack acknowledge of SANT owing to its rarity. SANT is usually mistaken as splenic inflammatory pseudotumor, hamartoma, or even malignant tumor when encounter spleen lesion in clinic [2]. Splenectomy is rather a method to reaches diagnosed than treatment in most cases because it's very difficult to diagnose SANT preoperatively [3, 4]. Therefore, we reported a case of SANT diagnosed preoperatively and treated by laparoscopy splenectomy in the end. We reviewed literature to investigate the preoperative diagnose of SANT and the appropriate treatment for SANT.

Case Presentation

A 50-year-old Hakka female was found with a low dense mass in the spleen by abdominal computed tomography (CT) during health checkup. She was asymptomatic and didn't have any disease history. There were no positive signs found in abdominal physical examination. Parameters of blood routine, liver and kidney functions tests and serum tumor markers were unremarkable. For further treatment, patient was hospitalized in our department. On the abdominal non-enhanced plain CT scan, a lesion was faintly observed with a vague margin, whose Hounsfield unit was approximate 41, while the surrounding normal splenic tissue were 43 (Fig. 1A). However, it was showed up significantly as a weakly enhanced lobulated-like mass of 57 × 41 × 50 mm with clear boundary on CT plain scan during rapid enhancement period. Compared to the enhancement of surrounding normal spleen (205 HU), the mass presented heterogeneous as well as much lower enhancement with a value of 69 HU (Fig. 1B). The CT value of the mass progressively increased to the peak of 127 HU on portal venous enhancement and delayed enhanced period, but still lower than that of the surrounding spleen (134HU). (Fig. 1C and 1D). Our radiologists proposed diagnosis of SANT differential from other benign lesion in the spleen. Subsequently, she underwent an abdominal magnetic resonance imaging (MRI). The lesion showed an iso-intensity to the normal spleen on T1-weighted (MRI) (Fig. 2A) but a little lower intensity in the T2-weighted (Fig. 2B). The lesion appeared heterogeneous hypo-intensity with lower intensity radial-like rift emission from central on T2-weighted, especially on fat-suppressed (Fig. 2A and 2B). The mass was progressively enhanced during MRI enhanced phases and eventually surpassed the background spleen (Fig. 2C, 2D and 2F). The conclusion of MRI also highly recommended SANT as first diagnosis. Interestingly, radial-like hypochoic gap separating mass also showed up on ultrasonic scan. (Fig. 3)

To get a precise pathologic diagnosis, laparoscopic splenectomy was successfully operated under patient's authorization. As is shown in Fig. 4.A, the resected specimen appears as a well-defined mass encircled by multiple nodules which are wrapped in transparent shell. And there's a characteristic white stellate colloid in the center of lesion. At low magnification, there are a great number of slit-like, round, or irregular shaped vascular nodules deformed by dense collagen fibrous or fibrosclerotic stroma. Plentiful erythrocytes and stromal cells with mild chronic inflammatory cell infiltration can be observed within the nodules under high magnification. Immunohistochemical staining of specimen presented three typical types of vessels directly pointing to the SANT diagnosis, which are (1) sinusoid type, both CD8 and CD31 positive but CD34 negative vessels; (2) small vein type, CD31 positive but CD8 and CD34 negative vessels; and (3) cord capillary type, both CD34 and CD31 positive but CD8 negative vessels.

There is no any complication and no sign of recurrence so far.

Discussion And Conclusion

SANT is a solitary entity distinguishing from other splenic vascular tumors and firstly defined by Martel et al because of its unique pathology [1]. The incidence of SANT remains unknown due to its rarity. In previous literatures, SANT mainly occurs in middle-aged population with a slightly female inclination and

the mean age at onset was 50 years, which is also supported in our case [5]. However, occurrence of SANT shows a young age trends in recent report [6, 7]. Youngest SANT patient was a 7-month-old infant with splenomegaly [8]. Most cases are found incidentally since most patients are asymptomatic. Majority symptomatic patients attended to clinic with main complaints of abdominal discomfort, others were nonspecific such as flank pain, weight loss, fatigue, night sweat, and mild fever (9).

With increasing cases reported, radiologist and physicians tried to conclude imaging characters of SANT in order to get preoperative diagnoses. On non-enhanced plain CT scan, SANT presented as hypodense mass predominantly, some as equal-density, and partial cases have small central calcification [10]. Further enhanced CT presented solitary well-circumscribed hypodensity lesion comparing to background spleen in artery phased. The contrast between lesion with parenchyma of the spleen was significant in rapid enhanced phased and diminished progressively in portal-venous phased and delayed phased [11]. Karaosmanoglu et al firstly report imaging on MRI, which presented as central hyperintense area simulating hemorrhage on T1-weighted MRI. What's more, they described the enhancement of SANT as progressively centripetal infiltration and the specific pattern was vividly named as "spoke-and-wheel" [12]. In Jianbing Ma's literature, "spoke-and-wheel" pattern in CT or MRI examination can be the key for SANT diagnoses with 58% occurrence rate in total 12 patients [13]. However, only 4 of 12 cases were diagnosed as SANT preoperatively indeed probably due to its rarity. Profound research on correlation between pathological and radiological characters demonstrated that spoke-and-wheel imaging is referenced to a stellate collagen stroma core surrounded by angiomatoid nodules that are separated with proliferative fibrous septa [10]. As is shown in Table1, there is few preoperative diagnoses cases reported and we were the one of several cases. In presented case, SANT was nearly iso-density in both non-enhanced CT and pre-contrast T1-T2 weighted on MRI, and T2 weighted could observed satellite hypodensity septa in center. The multiple phase imaging of CT was consistent with previous report, which was enhanced progressively but remain hypodensity to background. Although spoke-and-wheel was not presented, central satellite hypodensity septa existed lasting the whole enhancement of T1 weighted on MRI. By summarized previous literature, we repute that there are two typical types of enhanced imaging characters on CT and MRI: 1) enhancement started from peripheral part in progressively way presenting as spoke-and-wheel pattern; 2) enhancement evaluated progressively in entity way with or without hypo-intensity septa in center. Our case belongs to the later one. Similar to spoke-and wheel pattern, we suppose that satellite hypo-intensity septa are also corresponded to the colloid scar in center of gross. Another features we learned is that the CT value of SANT remain lower to the background of spleen during multiple phase. We believe that these features are helpful in preoperative diagnosis when spoke-and-wheel sign was absent.

Gray-scale ultrasonography usually shows a heterogeneously hypoechoic or isoechoic mass with hypoechoic rim compared to the surrounding splenic parenchyma. The lesion appeared "spoke-and-wheel" enhancement pattern as well on contrast sonography [14, 15]. Our case presented as heterogeneously majority isoechoic mass with hypoechoic radio-like linear area in center, which is similar to T2-weighted on MRI (Fig. 3 and Fig. 2A).

Positron emission tomography CT (PET-CT) is a powerful examination mainly applying in malignant tumor follow-up. SANT presented slight FDG accumulation, similar to tumor metastasis. John et. concluded that SANT can be a differential diagnosis in the patient with malignant tumor history [16, 17].

Needle biopsy possesses excellent accuracy in preoperative diagnosis despite accompanying with the risks of bleeding and needle path metastasis. Literature shows that bleeding risk of spleen needle biopsy can be retained in very low level under the ultrasonic guidance [16, 18].

Laparoscopic splenectomy is a prevalent therapy approach for variety spleen benign lesions, in which SANT accounted for 1.7% [4]. In our case, laparoscopic splenectomy was implemented in consideration that SANT is a benign mass and laparoscopic surgery has been surgery mature as well as regular in our department. In a series study with 37 cases, laparoscopic partial splenectomy demonstrated a great advantage in the sequent complication of surgery compare to total splenectomy [19].

Pathology is gold standard for SANT diagnosis. The gross appearance of resected spleen is coincided with spoke-wheel pattern in CT and MRI imaging, which presented as a well-circumscribed mass with a stellate white stroma at the center [10]. Under microscope, classic pathological characters are as follow: i) multiple angiomatoid nodules are overspread in the horizon separated by dense collagen fibrosis and fibroid rims. ii) Individual nodule is composed of slit-shaped, round, or irregularly shaped lumens lined by swollen endothelial cells. iii) numerous red blood cells with scattered inflammatory cells are crowded in the inflammatory cells. iv) Three typical types blood vessels are presented with immunohistochemical staining cord capillary-like type with CD34⁺CD31 co-expression and absence of CD8⁻, sinusoids-like type (CD34⁻/CD8⁺/CD31⁺), and small veins-like type (CD34⁻/CD8⁻/CD31⁺) [1, 20]. Those features mentioned above can be found in our case. Additionally, some researchers detected plenty IgG4-positive plasma cells that and proposed SANT as an IgG4 related disease [21, 22]. But the hypothesis was not approved by advanced studies, in which Cheng et al revealed that SANT is a reactive lesion originated polyclonally rather than a neoplasm [23]. What's more, no cases associated with IgG4 disease has been reported up to date.

In summary, SANT is potential to be diagnosed preoperatively when imaging features are profoundly comprehended. Laparoscopic splenectomy is a reliable and effective therapy option for SANT treatment or diagnosis.

Abbreviations

Sclerosing angiomatoid nodular transformation: SANT; computed tomography: CT; Hounsfield unit: HU; magnetic resonance imaging: MRI; fine needle aspiration: FNA; Ultrasonography: US;

Declarations

Ethics approval and consent to participate

The Ethics Committee of The Second Medical College of Jinan University, Shenzhen People's Hospital approved this study. The patient signed an informed consent form for surgery.

Consent for publication

Not applicable.

Availability of data and materials

The datasets used or analyzed in the current study are available from the corresponding author on reasonable request.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

TCW, YZ, and GZP conducted the surgery. TCW and YZ designed the report. YJY collected patient's clinical data and information. YJY and GZP wrote the paper. All authors read and approved the final manuscript.

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Tables

Table 1
Summary of preoperative SANT diagnosis in previous reports

Author	Case number	Age	Diameter	Preoperative Diagnosis	Type of examination	treatment
Jianbing Ma	4	21–62*	30–85*	SANT	CT,MRI	splenectomy
Keiso Matsubara	1	64	4.5 cm	SANT	CT,MRI,PET-CT	Follow up for 5 years, splenectomy eventually
Punit Sharma	1	56	3.9 cm	SANT	PET-CT,FNA	Follow up
Present case	1	50	4.5 cm	SANT	CT,MRI	splenectomy
*no mention in detail						

Figures

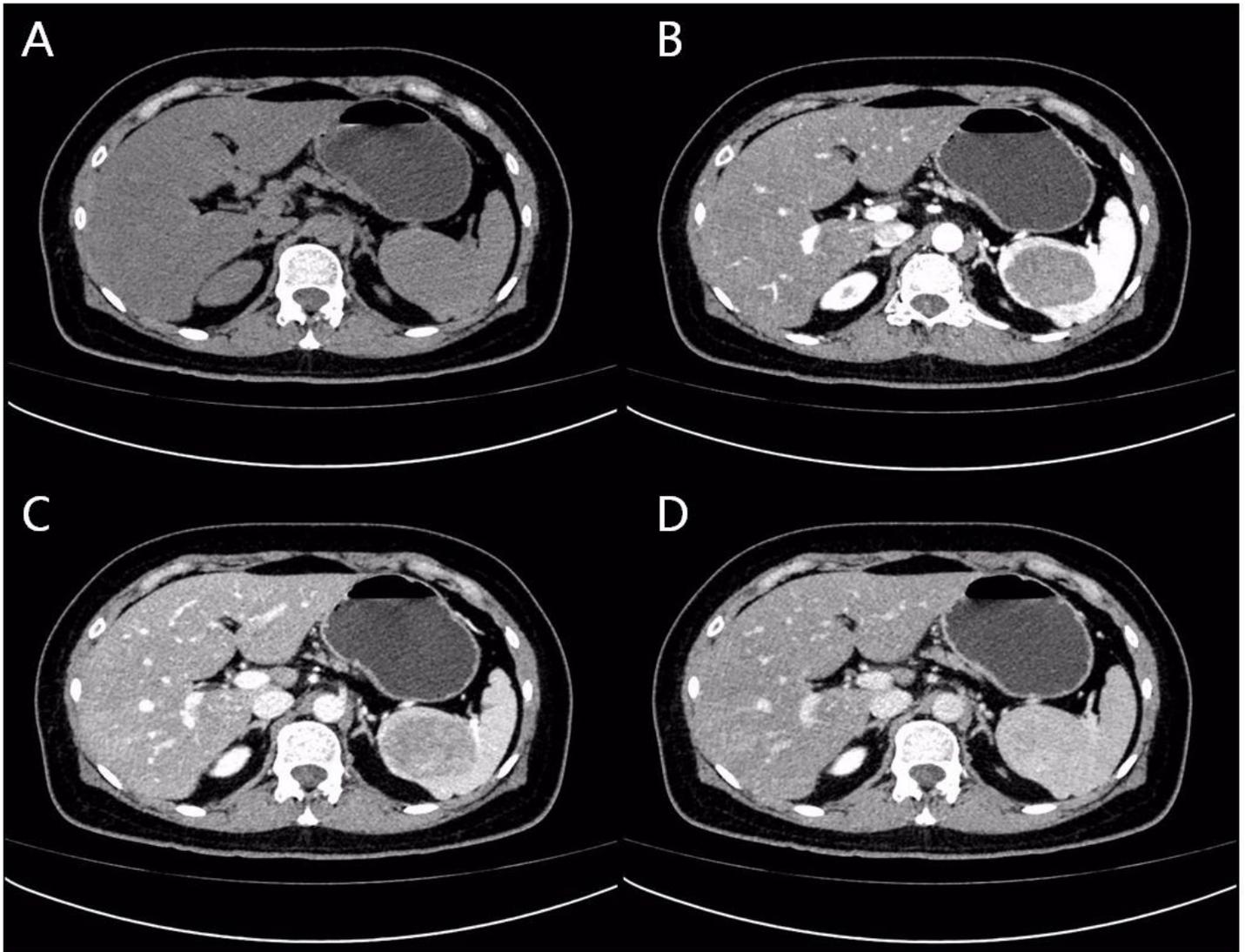


Figure 1

SANT showed asynchronous enhancement comparing to parenchyma of spleen on CT scan multiple phase. A, non-enhanced CT scan B, artery enhanced period CT scan C, portal venous enhanced period CT scan D, delayed enhanced period CT scan.

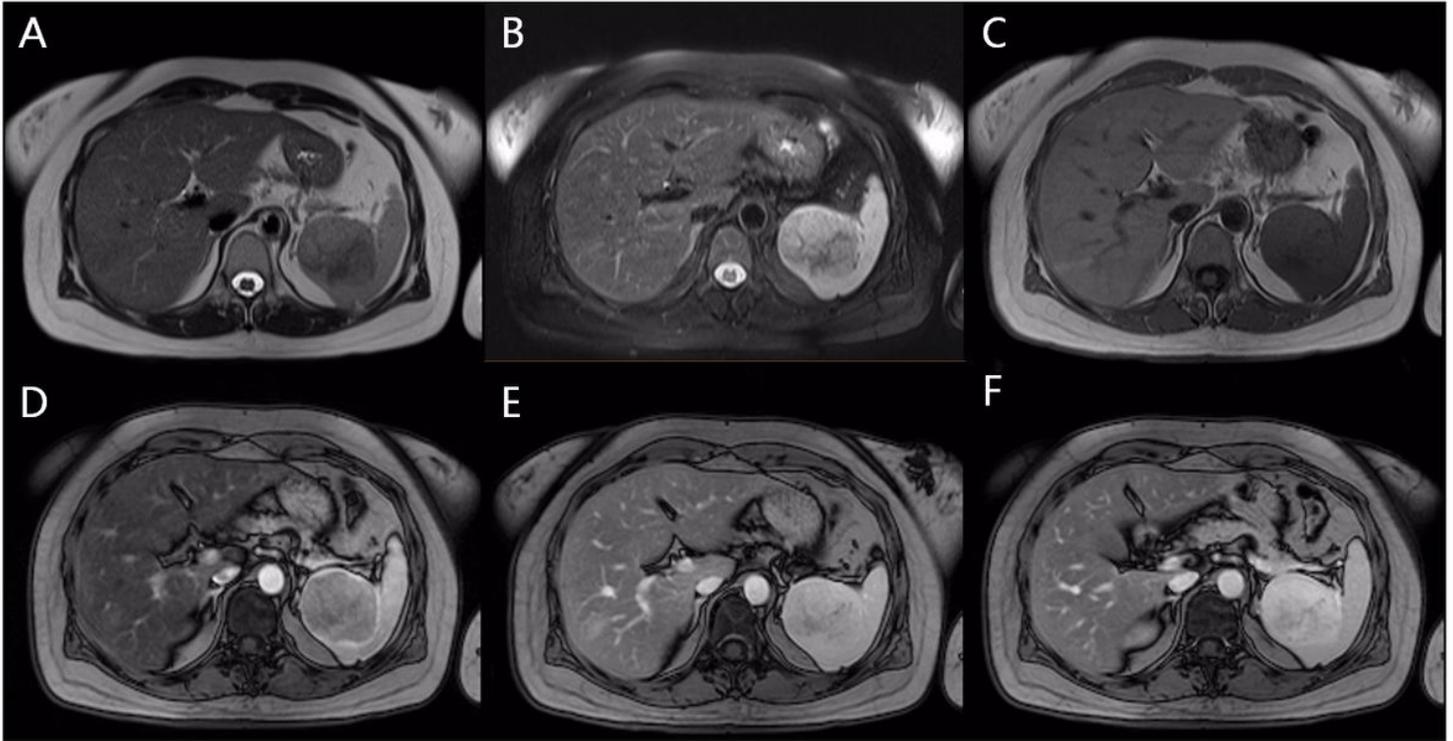


Figure 2

SANT presented radio-like hypointensity rift splitting in central on MRI multiple phases. A, T2-weighted MRI on fat-saturate; B, T2-weighted MRI on fat-suppress ; C, T1-weighted MRI of precontrast; D-E, T1-weighted MRI after injection

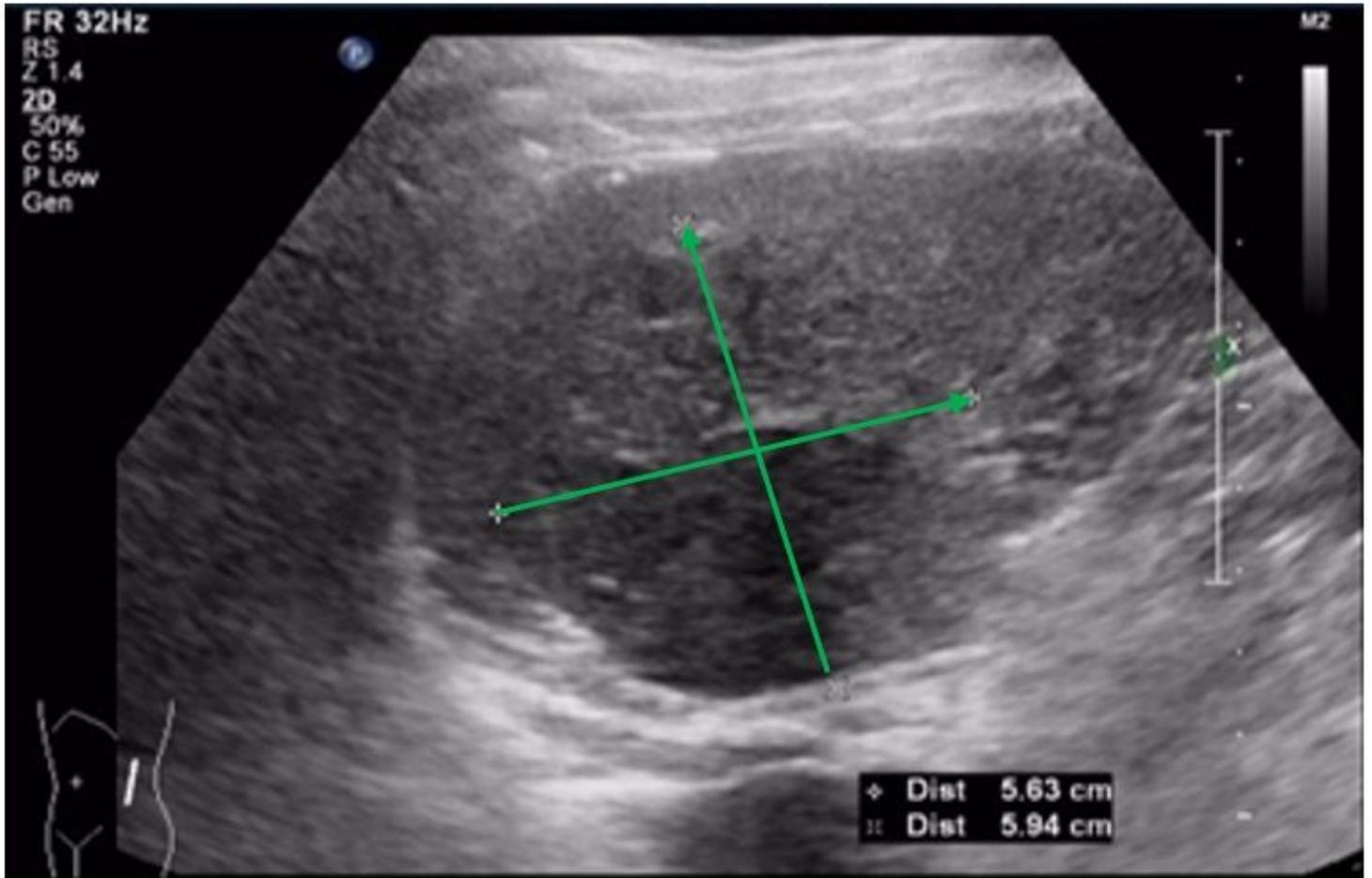


Figure 3

Imaging of SANT on US

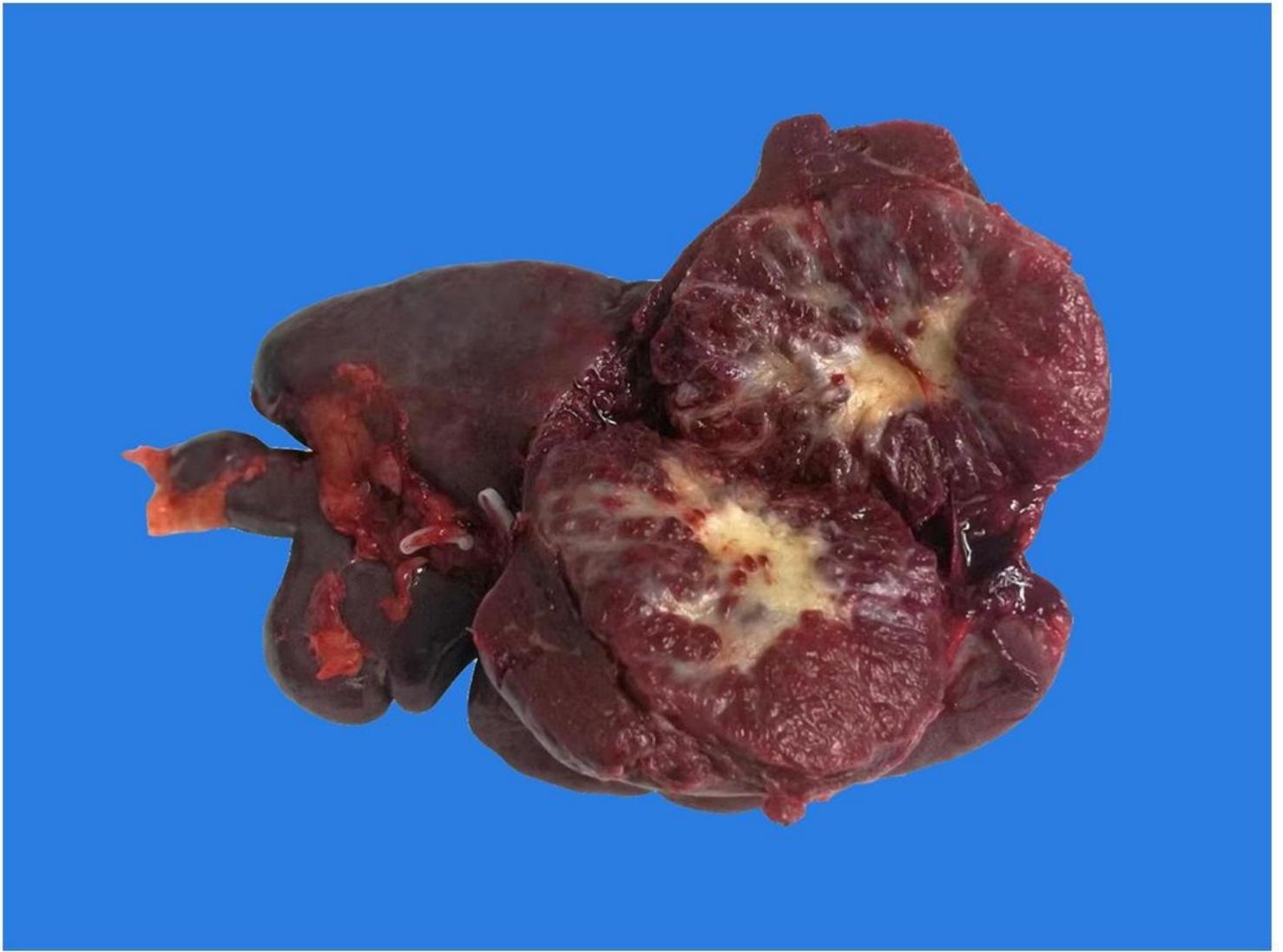


Figure 4

Macroscopic examination of the postoperative specimen. The resected lesion measured 5.8*4.6*4.3cm in size