Case Report

Ultrasonographic and pathological features of sclerosing stromal tumor: a case report

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Background: We herein report a rare case of a sclerosing stromal tumor (SST) in an adolescent. In this case, the mass displayed a shrinking trend, combined with its ultrasonic manifestations and pathological characteristics and may provide some references for the selection and timing of treatment, to avoid excessive harm to patients.

Case Description: A healthy 17-year-old adolescent female presented to the outpatient department, complaining of abnormal uterine bleeding, but no abdominal pain, bloating, chills, or fever. The patient had no history of malignant tumors, and no relevant family or genetic history. An ultrasound showed an inhomogeneous hypoechoic area (106 mm × 53 mm × 68 mm) in the right ovarian, a clear boundary, an anechoic area inside and blood flow was observed in the mass. At a follow-up regular re-examination, the mass displayed a shrinking trend from 95 mm × 50 mm × 88 mm, 61 mm × 28 mm × 42 mm, 43 mm × 28 mm × 40 mm, 43 mm × 28 mm × 40 mm, to 42 mm × 23 mm × 28 mm. The patient underwent laparoscopic surgery a week later. Based on the immunohistochemistry and morphology results, the posterior ovarian mass was diagnosed as an SST. At one month after operation, there was no obvious abnormality on ultrasound.

Conclusions: The incidence of SST is relatively low. However, due to the low specificity of clinical manifestations, imaging examination and serum tumor markers, the diagnosis of SST mainly relies on pathological examination. Therefore, in clinical practice, the possibility of misdiagnosis is greater, and attention should be paid to the differentiation of ovarian malignant tumors. Surgical resection is recommended, and the effect is good. Surgical methods should be selected individually according to the size of the tumor and the age of the patient.

Keywords: Rare disease; sclerosing stromal tumor (SST); short-term changes; ultrasound; case report

Submitted Jul 06, 2022. Accepted for publication Aug 17, 2022.
doi: 10.21037/gs-22-427
View this article at: https://dx.doi.org/10.21037/gs-22-427

Introduction

An ovarian sclerosing stromal tumor (SST) is a rare benign ovarian sex cord stromal tumor. It is currently believed to originate from undifferentiated stromal cells with multi-directional differentiation potentials in the ovarian cortex (1). According to statistics, SST accounts for 2–6% of ovarian sex cord stromal tumors, and predominately occurs in young women aged 20–30 years (2,3). Previous reports have provided no specificity in terms of the clinical manifestations, imaging features, or serum tumor markers of SST. The common clinical manifestations of SST include irregular menstruation, abdominal pain, and a pelvic mass. Imaging findings are mainly solid or mixed solid-cystic masses with clear boundaries in the ovary (4,5). As the above characteristics cannot be used to distinguish SST from ovarian malignant tumors, clinical diagnosis is difficult and misdiagnosis is frequent; thus, the diagnosis of SST is

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mainly based on a pathological examination.

Surgery is an effective treatment for SST. To date, there are no case reports of postoperative recurrence and metastasis (6,7). Atram *et al.* (8) noted that tumor resection or salpingo-oophorectomy on the affected side is the best treatment for SST. As the disease mostly occurs in young women with reproductive needs, an intraoperative frozen pathological examination must be performed to determine the scope of surgery to avoid excessive physical and mental harm to patients (9,10).

The clinical data of the patient admitted to our hospital was retrospectively analyzed. After careful consideration, the patient did not receive immediate surgical treatment but chose to closely monitor any changes to the lesions under ultrasound. Therefore, different from previous reports that only focused on the clinical manifestations, serological tumor markers and treatment methods of SST, in this case, we observed the development trend of SST, and the trend of SST was gradually shrinking. Combined with the ultrasound findings and pathological features, we considered that the reduction of the mass was related to the fact that the mass was rich in small vesicles. At the same time, the surgical methods are selected according to tumor size and patient age, so this finding helps us to choose the appropriate surgical timing and narrow the surgical scope when we encounter SST with similar ultrasound manifestations. We present the following article in accordance with the CARE reporting checklist (available at https://gs.amegroups.com/article/view/10.21037/gs-22-427/rc).

Case presentation

A 17-year-old adolescent female presented to the outpatient department, complaining of abnormal uterine bleeding, but no abdominal pain, bloating, chills, or fever. The patient had regular menstruation, 30 days cycle, 7 days period, moderate volume, red color, no dysmenorrhea, no abnormal leucorrhea, the first menstruation was 12 years old and had no history of malignant tumors, and no relevant family or genetic history. An ultrasound showed an inhomogeneous hypoechoic area (106 mm × 53 mm × 68 mm) in the right adnexal area, a clear boundary, and an anechoic area inside, a resistance index (RI) of 0.44, and possibly from ovary. The mass had well mobility without tenderness. Based on the ultrasound imaging results of the patient, the mass was considered a malignant space-occupying lesion of the ovary. But the serological tumor markers carcinoembryonic antigen, alpha-fetoprotein, CA125, CA199 and CA153 were normal. Besides Serum sex hormone test results were also basically normal. After careful consideration by the patient and her family, an operation was not immediately performed, and the patient chose to undergo re-examinations to monitor the mass.

The patient was reviewed every week, and the ultrasounds revealed that the right adnexal mass displayed a shrinking trend from 95 mm × 50 mm × 88 mm, 61 mm × 28 mm × 42 mm, 43 mm × 28 mm × 40 mm, 43 mm × 28 mm × 40 mm, to 42 mm × 23 mm × 28 mm (see *Figure 1*). At the last 3 re-examinations, no significant change in tumor size was observed. At the last contrast-enhanced ultrasound (CEUS) examination, the hypoechoic area of the right adnexa began to enhance at 18 s after an injection of sonovit, which was earlier than the surrounding ovarian tissue, and showed heterogeneous high enhancement, which began to decrease to low enhancement at 50 s, and then continued to low enhancement. Because the mass displayed a shrinking trend, a borderline ovarian tumor was considered (see *Figure 2*). Borderline tumors generally have low malignant potential and have some of the characteristics of benign and malignant tumors, and have a good prognosis.

The patient underwent laparoscopic surgery a week later. Intraoperatively, we noticed about 20 mL of pale yellow ascites in the pelvic cavity, a normal-sized uterus, and an enlarged right ovary (about 6 cm × 3 cm × 2 cm). A right salpingo-oophorectomy was performed. The lesion was about 2 cm × 1 cm and had an unclear boundary with the surrounding tissue. A frozen section suggested benign lesions. Based on the immunohistochemistry and morphology (hematoxylin and eosin staining) results, the posterior ovarian mass presented as an SST, with a maximum diameter of about 3.5 cm. The immunohistochemistry results were as follows: Calretinin (+ focal), Cytokeratin (CK)7 (–), cluster of differentiation (CD)10 (–), CD99 (+ weak), Epithelial membrane antigen (EMA) (–), inhibina (+ slightly), melan-A (A103) (–), Ki-67 (+, <5%), Smooth muscles actin (SMA) (–), Wilms' tumor (+ focal), CK (pan) (+ slightly), and desmin (–) (see *Figure 3*). At one month after operation, there was no obvious abnormality on ultrasound. Subsequent periodic ultrasound examinations every three months showed no significant abnormalities (see *Figure 4*).

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Declaration of Helsinki (as revised in 2013). Written

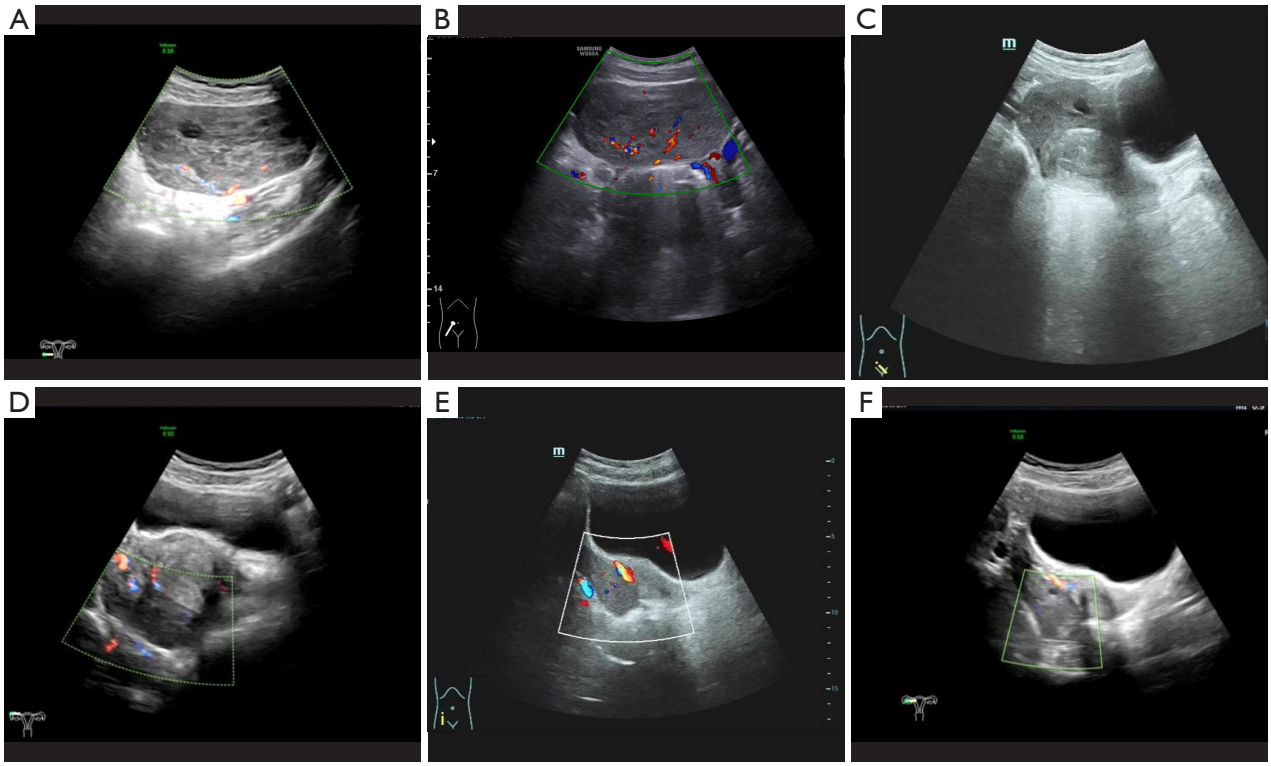

Figure 1 Results of routine ultrasound examinations during follow-up. The right adnexal mass was observed to shrink over time. The ultrasound times for the images above were as follows: 1 (A), 9 (B), 25 (C), 40 (D), 55 (E), and 70 d (F).

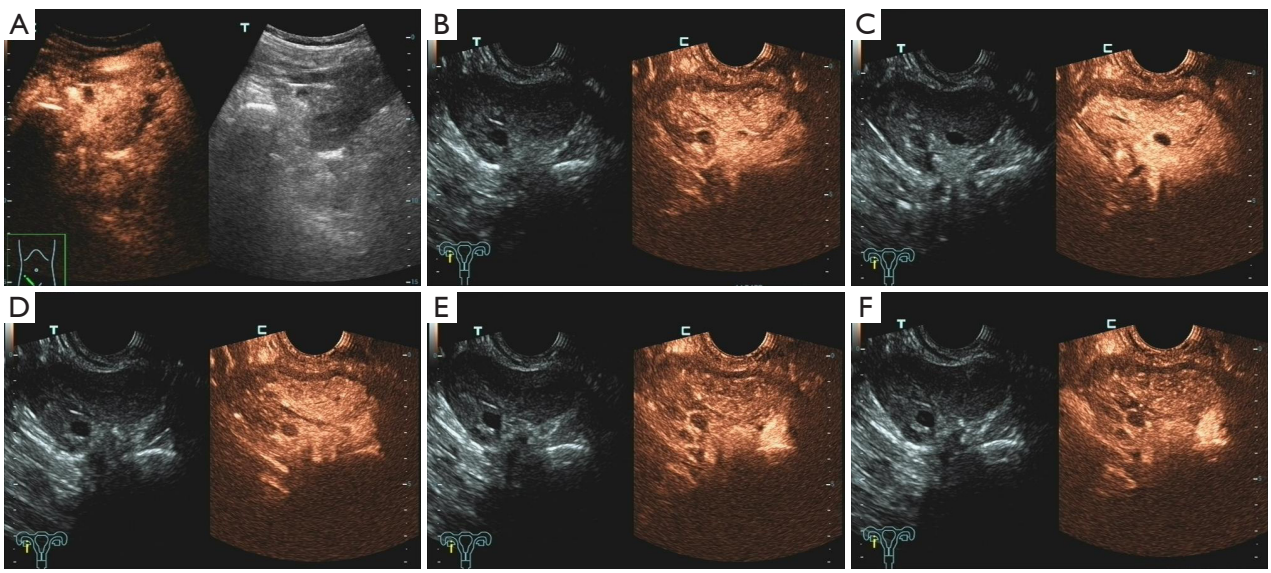

Figure 2 CEUS findings of the ovarian mass at the following different time points: 23 (A), 24 (B), 25 (C), 26 (D), 40 (E), and 52 s (F). CEUS, contrast-enhanced ultrasound.

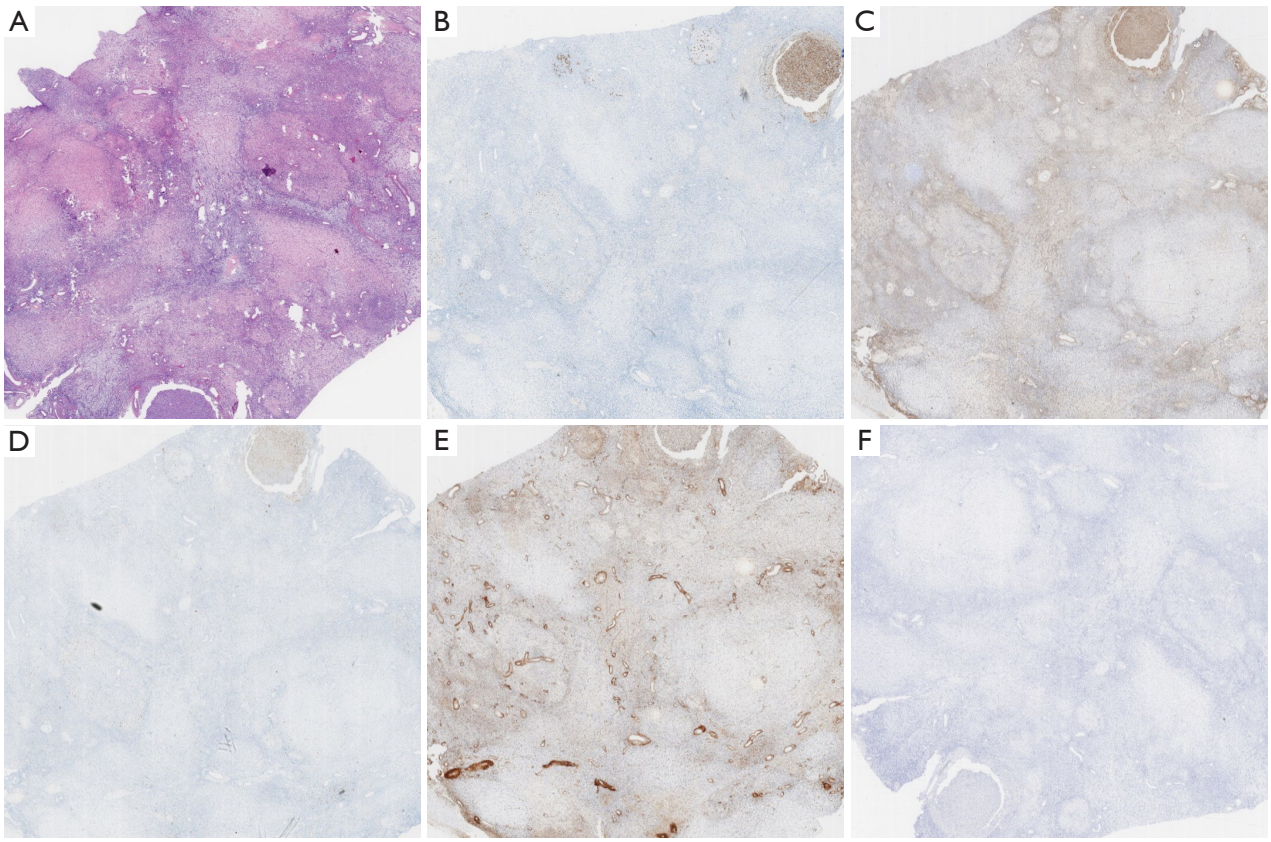

Figure 3 Immunohistochemistry and morphology results (×400); haemotoxylin and eosin staining of tumor after treatment (×400). (A) A histological analysis revealed the pseudo-lobular structure and sclerotic zone of the collagen fibers. Immunohistochemically, the tumor cells were positive for calretinin (B), CD99 (C), inhibina (D), Wilms' tumor (E), and CK (pan) (F). Based on these findings, the patient was diagnosed with a sclerosing stromal tumor.

informed consent was obtained from the patient's guardians for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

Discussion

OSST was first identified by Chalvardjian and Scully in 1973 and was classified as an ovarian sex cord stromal tumor in 1999 by the World Health Organization (11,12). In this article, we reported an extremely rare case of a 17-year-old adolescent female with an SST. The ultrasonic manifestations of an SST are closely related to its pathological features, which typically include a complete capsule, internal follicle area, collagen fibers, or pseudo-lobules formed by loose and edematous tumor cells (13). The RI of the blood flow in this case was always low during the review process, which was consistent with relevant literature reports at home and abroad. A low RI value is associated with the histopathological features of multiple thin-walled small vessels between pseudo-lobule cells (14); thus, SSTs are easily misdiagnosed as ovarian malignancies.

The ultrasound findings of the tumor were retrospectively analyzed, and the tumor displayed a heterogeneous hypoechoic pattern with multiple thin strip-like hyperechoic and sporadic anechoic patterns (see *Figure 1A*). As *Figure 1B-1F* show, the thin strip-like hyperechoic and anechoic areas inside the tumor gradually decreased or even disappeared. As *Figure 1F* shows, the tumor appeared hypoechoic. The microscopic cystic area in the solid tumor was considered the pathological basis for the uneven internal echo and scattered thin strip hyperecho indicated by the ultrasound images. Because of this structure, the difference in the acoustic impedance between the 2 sides of the interface was large, resulting in a strong reflection of the acoustic beam at the interface

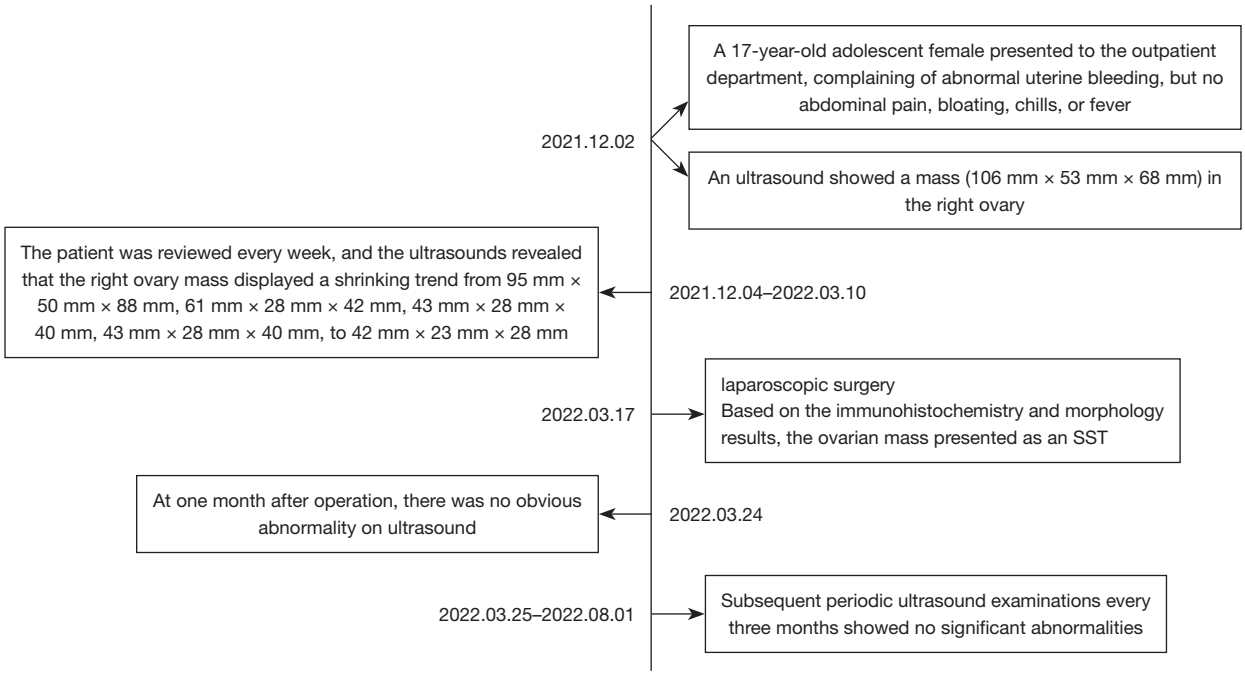

Figure 4 Timeline of interventions and outcomes in this case.

and the formation of the small thin strip-like hyperecho in the ultrasound images (15). During the review, the tumor volume gradually decreased from the anterior upper uterus to the periovarian region, which was thought to be related to the gradual absorption of the cystic region of the tumor from the ultrasound image changes.

As *Figure 1F* shows, the thin strips of hyperechogenicity within the tumor disappeared, which might explain why there was no reduction in tumor volume during the 3 subsequent reviews. Some scholars have identified the following enhancement features of SST (16): (I) obvious peripheral enhancement in the early stage and gradually increasing centrality in the late stage; (II) uniform enhancement in the early stage, which is sustained in the late stage; (III) early mild enhancement, late continuous enhancement, but the intensification is not as strong as above two.

Torricelli *et al.* (16) noted that the first SST enhancement method is similar to the hepatic cavernous hemangioma enhancement method with specificity. Similar enhancements of the lesion may suggest a diagnosis of SST. Additionally, the pattern of tumor enhancement is closely related to the proportion of the tumor components, and the enhancement of tumor margins is associated with the abundance of blood vessels between the pseudo-lobule cells in the tumor tissue. A few scattered tumor cells in the edema area showed mild to moderate enhancement, or delayed enhancement. There was no enhancement in the area of cystic necrosis. In this case, the tumor showed rapid and uneven hyperenhancement, reflecting a high proportion of pseudo-lobular components in the tumor. However, due to the absence of CEUS during the follow-up period, the cause of tumor reduction could not be further analyzed from the contrast-enhanced features.

Conclusions

It has been reported that the malignant rate of ovarian tumors in adolescent women is high, and accounts for about 31% (17). However, for young patients with menstrual disorders, when a malignant tumor is suspected, the possibility of SST should be considered to avoid over diagnosis and treatment. In this case, a retrospective analysis suggested multiple thin strips with high echo in the tumor. The solid part of the tumor was in a large number of small cystic areas, and continuous absorption was observed in the cystic area of the tumor, which reduced the volume of the tumor, which in turn was conducive to the timing of surgery, narrowed the scope of operation, and reduced the harm to the patient. Additionally, after CEUS was added

during the partial follow-up, we were better able to assess changes in the tumor components and we improved the treatment efficiency by analyzing changes in the tumor enhancement patterns.

Acknowledgments

Funding: None.

Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at https://gs.amegroups.com/article/view/10.21037/gs-22-427/rc

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://gs.amegroups.com/article/view/10.21037/gs-22-427/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Declaration of Helsinki (as revised in 2013). Written informed consent was obtained from the patient's guardians for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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(English Language Editor: L. Huleatt)

Cite this article as: Qian X, Wang X, Xu Z, Zhang S. Ultrasonographic and pathological features of sclerosing stromal tumor: a case report. Gland Surg 2022;11(9):1555-1561. doi: 10.21037/gs-22-427