

Sclerosing stromal tumor: a rare ovarian neoplasm

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Abstract

Sclerosing stromal tumor (SST) is an extremely rare and distinctive sex cord stromal tumor, which occurs predominantly in the second and third decades of life. SSTs make up 2-6% of ovarian sex-cord stromal tumors. Due to the solid and distinct vascular structure of the tumor, it can be mistaken as a number of malignant ovarian tumors. As this specific neoplasm is very rare, it is not always possible to diagnose the tumor preoperatively with clinical and ultrasonographic findings. Furthermore, histopathological and immunohistochemical analysis does not always confirm the diagnosis. In this case report, clinical findings, histopathological features, and macroscopic appearance during laparoscopy of an SST are presented in a 20-year-old woman with pelvic pain. SST should be considered among the differential diagnosis of women with adnexal masses.

Keywords: Benign ovarian neoplasm, laparoscopy, sclerosing stromal tumor

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Introduction

Sclerosing stromal tumor (SST) is a very rare, benign ovarian tumor, which was first described in 1973 by Chalvardjian and Scully (1). To date, less than 208 cases have been reported worldwide (2). SST is categorized as being one of the sex cord stromal ovarian neoplasms. It can be differentiated from other stromal tumors clinically as well as pathologically (3). Between 2% and 6% of all the sex cord stromal tumors are SST (4). These tumors are commonly seen in patients who are in their second or third decades (3). Pelvic pain, menstrual irregularities and abdominal mass are the most common symptoms and findings. Solid structures in the macroscopic examination of the tumor may be mistaken for malignancy. This may lead to unnecessary radical surgery (5). SSTs are usually unilateral, and well demarcated and recurrences are not reported (3). Histopathologic and immunohistochemical analyses confirm the diagnosis.

The purpose of this case presentation was to show the macroscopic view and the laparoscopic excision of an SST in a 20-year-old woman. To the best of our knowledge,

this is the first video article to describe a laparoscopic SST operation.

Presentation of case

A 20-year-old virgin woman attended our outpatient gynecology clinic with the complaint of lower abdominal pain for six months. During the physical examination, an abdominopelvic mass was detected at the right lower abdominal area. A unilateral, heterogenous, cystic mass, originating from the right adnexal area was visualized with ultrasonography. No pathological laboratory findings were reported. All the tumor markers were in the normal ranges. On magnetic resonance imaging a heterogeneous, smooth, contoured mass with fat-intensity areas and solid components was observed in the right adnexal area with measurements of 60x50 mm. Dermoid cyst was considered as a differential diagnosis. The patient was referred to gynecologic oncology. Since malignancy was not primarily considered, laparoscopic cystectomy was planned by the gynecology team. A 10 mm trocar was inserted into the abdominal cavity by direct entry technique from the umbilicus, and a pneumoperitoneum was created. Two



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lateral trocars were placed on bilateral lower quadrants, and one suprapubic trocar was placed in the same plane as one lateral trocar. During the operation, a 60x50 mm sized, multi-lobulated mass with a smooth and intact external surface, apparently originating from the right ovary, was observed. When cut, the internal surface of the mass was grey white to yellowish in color and was solid with a rubbery consistency and contained small cystic spaces. The mass was attached to the ovarian cortex very tightly, and there was a dense blood supply to the mass. It was hard to separate the mass from the ovarian cortex. During the operation, multiple contaminated, whitish viscous tissue pieces, the largest being 4.5x3x2 cm, and the smallest being 1x0.5x0.3 cm were sent for frozen section examination. The result was reported as sex cord stromal tumor (fibroma?), although the definite diagnosis would have to wait for paraffin section examination. The tumor was totally excised and the operation ended. A total operative time of 45 minutes and estimated blood loss of 150 mL were recorded. No intraoperative surgical complications were observed. On postoperative day 1, the patient was discharged from the hospital uneventfully. Histopathologically, the definite result was reported as "SST" (Supplementary Video 1).

The patient was examined one, six and 12 months postoperatively. During the follow-up examinations, the patient reported that her inguinal and abdominal pain had completely resolved. During the follow-up period recurrence was not observed.

Written informed consent was obtained from the patient for publication of this video article and any accompanying images.

Discussion

Approximately 8% of all primary ovarian neoplasms are ovarian sex cord stromal tumors (6). Granulosa cell tumors, fibrotecomas, Sertoli-Leydig cell tumors, steroid cell tumors, and SSTs are categorized as ovarian sex cord stromal tumors (6). Commonly, ovarian sex cord stromal tumors are seen in a single ovary, but rarely they can be detected bilaterally. The youngest patient reported in the literature was 4-years old (7). In our case, the patient was 20 years old and had a unilateral ovarian cyst.

Frequently seen symptoms include menstrual irregularities and pelvic pain (3). There may be masculinization or anovulation due to estrogen and/or androgen secretion (3). In our case there was no clinical virilization and hormone levels and tumor markers were normal.

Macroscopically, SST is a solid, often yellowish mass, varying in size from 3 to 17 cm. SSTs also tend to be well differentiated and usually present with edema and cystic components. The tumor consists of cellular areas with pseudolobular structures surrounded by edematous and collagenous

stroma. Hemangiopericytoma-like capillary-rich fields can be detected in these cellular areas (8). Lobule structures consist of two types of cells; spindle-shaped cell secreting collagen and Theca-like cells containing lipids, eosinophilic cytoplasm with vacuoles, and with small dark nuclei with a definite nucleolus (9). It has been reported that inhibin and calretinin are important immunohistochemical markers that help in the diagnosis of ovarian sex cord stromal tumors (5). In our specimen, positive immunohistochemical staining for inhibin and calretinin led us to believe that the tumor originated from stroma.

Differential diagnosis of the SST is essential. Frozen section is crucial for making a distinction between SST and malignant ovarian tumors, because of the similarity in their macroscopic appearance (9). An SST may easily be mistaken for a fibroma or thecoma, both clinically and histopathologically (9). The pattern of the tumor and the patient age will help to differentiate SST from other tumors. Massive ovarian edema may be present with SSTs. In order to eliminate this confusion, compressed ovarian tissue can be identified by palpation of the stroma in the massive ovarian edema (10). SSTs can be treated successfully with unilateral salpingo-oophorectomy or enucleation. There is no local or distant metastasis reported in the literature (10).

Conclusion

As SSTs are rarely encountered, a preoperative clinical and ultrasonographic diagnosis can be challenging. SST should be considered in the differential diagnosis of patients presenting with unilateral, solid cystic, and complex ovarian masses. This tumor has a benign course and good prognosis with conservative surgery.

Supplementary Video 1. Laparoscopic excision of "sclerosing stromal tumor: a rare ovarian neoplasm"



<https://www.doi.org/10.4274/jtgga.galenos.2021.2021.0097.video1>

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