# Sclerosing Stromal Tumor of the Ovary

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**OBJECTIVE:** Sclerosing stromal tumor (SST) of the ovary is a rare ovarian disease with prevalence of 1.5 % to 6 % of ovarian stromal tumors. These tumors appear solid and are very vascular and therefore give the impression of a malignant tumor.

**STUDY DESIGN:** Clinical data of five patients with SST are presented. All of the patients underwent surgical treatment. Diagnosis of SST is revealed by final histopathological examinations.

**RESULTS:** Among the five cases encountered the mean age at presentation was 30.2 years. The clinical presentation varied from menorrhagia (1 case), irregular menses (1 case), pelvic pain (2 cases) and asymptomatic mass (1 case). Ca 125 level was elevated in only one case.

**CONCLUSION:** SSTs should be considered in young women with menstrual irregularity who have hypervascular solid and cystic adnexal masses. Though the tumor appears malignant, since it occurs in young women, care should be taken before embarking on radical surgery.

**KEY WORDS:** Stromal tumor, Sclerosing, Adnexal mass

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## Introduction

Sclerosing stromal tumor (SST) of the ovary is a rare benign ovarian stromal tumor with prevalence of 1.5% and 6% of ovarian stromal tumors.[1] It is first described by Chalvardjian and Scully in 1973.[2] This tumor occurs predominantly in the second and third decades and is histologically characterized by the presence of pseudolobulation of cellular areas separated by edematous connective tissue, increased vascularity, and prominent areas of sclerosis. Most patients with this tumor present with menstrual irregularities and pelvic pain.[3] These tumors appear solid and are very vascular and therefore give the impression of a malignant tumor. Morphologically and histologically they have distinct characteristics, which make them different from other stromal tumors.[4] Diagnosis of SST is often made by postoperative pathological examinations. Tumor markers usually remain normal in patients with SST.

The present study describes five cases of sclerosing stromal tumors.

## Material and Method

The present study was approved by the Ethical Committee and Institutional Review Board of Zekai Tahir Burak Women's Health Care Education and Research Hospital, Ankara, Turkey. The present study was conducted at the Gynecologic Oncology Department of the hospital.

Clinical data of five patients with SST are presented. All of the patients underwent surgical treatment. The general attitude of the study center towards suspicious pelvic masses detected by pelvic examination and imaging studies for ovarian malignancy is to perform explorative surgery with frozen investigation regardless of the patient's age and Ca-125 values. So that intra-operative frozen investigation was performed to all patients. As the frozen investigations revealed benign masses radical surgery was not performed. Diagnosis of SST is revealed by final histopathological examinations.

## Results

Among the five cases encountered the mean age at presentation was 30.2 years. The clinical presentation varied from

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menorrhagia (1 case), irregular menses (1 case), pelvic pain (2 cases), and asymptomatic mass (1 case). Ca 125 level was elevated in only one case. Sonographic findings preoccupied malignant tumors. Three of the patients underwent unilateral salpingo-oophorectomy and the other two underwent tumoral excision. Grossly the appearances varied from a solid, partly cystic, hemorrhagic, edematous, nodular (3 cases), firm tumor with yellow flecks (1 case), solid tumor with mixoid areas in patches (1 case). Microscopically, the tumors were characterized by cellular pseudolobules composed of a disorderly admixture of collogen producing fibroblasts (Figure 1).

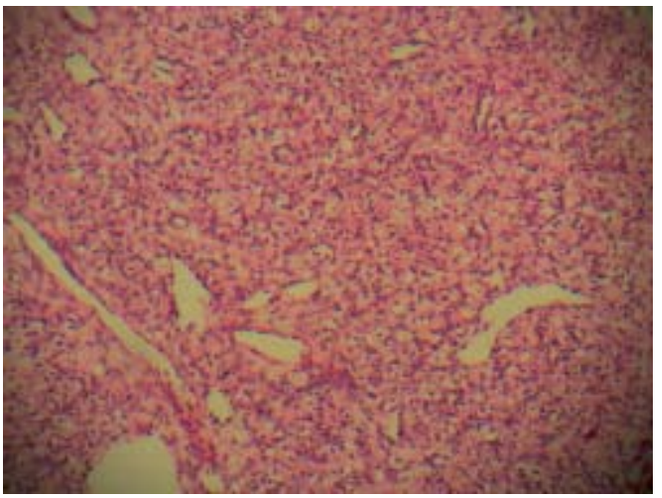

*Figure 1: Sclerosing stromal tumor. Spindle and oval shaped tumor cells with distinct vascularization in fibrous stroma*

In cellular pseudolobuler sections edematous connective tissue with low cell density and fibrous connective tissue with high cell density were mixed. The results of immunohistochemical stainings were positive for vimentin and smooth muscle actin. Based on these histomorphologic findings the diagnosis of sclerosing stromal tumor was made in all cases. The postoperative course was uneventful for all cases, and they were discharged on the fifth day after the operation and the report of final histology. All of them have been followed on an outpatient basis for one year, with no adnexal pathology. Ca-125 returned to normal soon after the operation and remained in the normal range in the one patient with an elevated Ca-125. Table 1 shows the clinical data of the cases.

## Discussion

SST of the ovary is a rare tumor derived from the sex cord stroma. The vast majority of tumors in the thecoma-fibroma group are readily subcategorized based on relatively distinct clinical and histological characteristics. Major subcategories include thecoma, fibroma-fibrosarcoma, and SST.[3]

This relatively rare tumor differentiates itself histologically and clinically from both thecomas and fibromas. The presence of pseudolobulation of cellular areas separated by edematous connective tissue, increased vascularity and prominent areas of sclerosis are histologic characteristics. Clinically, SSTs mostly occur in the second and third decades of life, whereas other types of stromal tumors are most common in the fifth and sixth decades. Most patients with SSTs present with menstrual irregularities and pelvic pain as it was seen in our case series. Although some patients with SST have shown increased hormone levels including estrogen, androgen, and testosterone, SST usually causes no change in tumor markers.[5] Although there are reported cases with elevated Ca-125 levels in the literature,[3,5-6] no specific tumor marker has been identified for SSTs to date. Terauchi et al.[5] suggested that the expansion of the tumor may cause mild peritonitis, resulting in the increase in peritoneum derived Ca-125. Consistent with

*Table 1: Clinical data of the cases with sclerosing stromal tumor*

| Case number | Age | Symptom | Gross Appearance | Treatment | Ca-125 | Follow-up (U/mL) |
|---|---|---|---|---|---|---|
| 1 | 38 | menorrhagia | R, 7x5x4 cm solid, partly cystic, edematous | tumoral excision | 21 | NS, 1 year |
| 2 | 28 | irregular menses | L, 8x5x6 cm solid, partly cystic, gelatinous | tumoral excision | 18 | NS, 1 year |
| 3 | 28 | pelvic pain | R, 11x9x5 cm solid, partly cystic, gelatinous, nodular | RSO | 10 | NS, 1 year |
| 4 | 27 | pelvic pain | R, 12x6x5 cm solid, firm tumor with yellow flecks | RSO | 72 | NS, 1 year Ca-125, WNL |
| 5 | 30 | asymptomatic mass | L, 8x8x7 cm solid, partly cystic tumor with mixoid areas | LSO | 12 | NS, 1 year |

***R:** right, **L:** left, **RSO:** Right Salpingo-Oophorectomy, **LSO:** Left salpingo-oophorectomy, **NS:** No Symptoms, **WNL:** Within Normal Limits,*

the literature only one of the patients in our case series was presented with an elevated serum Ca-125 level.

Lee et al.[7] analyzed seven cases of SSTs, which showed a typical sonographic appearance. According to their findings, on sonograms SSTs were solid and cysticand contained multiple round or cleftlike cysts. Ascites was rare. On transvaginal color Doppler sonograms, SSTs were very hypervascular in the peripheral solid area and internal cystic space and showed low-impedance flow. They concluded that SSTs should be considered in young women with menstrual irregularity who have hypervascular solid and cystic adnexal masses. It is often difficult to differentiate SST from a malignant tumor with imaging techniques because ST is observed as a mixture of solid and cystic parts on ultrasonic and MRI images.[5] In all of our five cases, sonographic findings preoccupied suspicious malignant tumors with solid appearance and extended size. As the general attitude of the study center towards suspicious pelvic masses detected by pelvic examination and imaging studies is to perform explorative surgery with frozen investigation regardless of the patient's age and Ca-125 values, intraoperative frozen investigation was performed. Fortunately, none of them underwent extensive surgery.

Immunohistochemical expression of tumor cells may be helpful for accurate diagnosis. Immunohistochemistry of desmin, vimentin and smooth muscle actin is useful in distinguishing SSTs from thecomas and fibromas.[3,6] It is suggested that SST is derived from a population of a muscle specific actin positive elements from the theca externa, namely the perifollicular myoid stromal cell.[8] Similarly, immunohistochemical findings of our cases showed positive staining for vimentin and smooth muscle actin.

In conclusion, SSTs of the ovary are rare benign tumors of the fibroma-thecoma group with appearance of solid and very vascular features giving the impression of malignant tumors. The clinical and microscopic features of our patients were similar to those reported in the literature. The present case series suggested that it is necessary to keep the possibility of SST in mind in selecting surgical techniques for potentially malignant tumors in young women. This means including intraoperative pathologic diagnosis in the surgery.

## Overin Sklerozan Stromal Tümörü

**AMAÇ:** Overin sklerozan stromal tümörü nadir görülen over tümörlerindendir. Prevalansı over stromal tümörleri içerisinde %1.5-6'dır. Solid ve damarsal yapıları gelişmiş tümörler olmalarından dolayı, malign tümor izlenimi verirler.

**GEREÇ VE YÖNTEM:** Malignite şüphesiyle operasyona alınarak frozen inceleme sonucunda benign kitleler tespit edilen ve final patolojisi sklerozan stromal tumor olan 5 hastanın klinik bulguları sunulmuştur.

**BULGULAR:** Tanı konduğunda hastaların ortalama yaşı 30.2 idi. Klinik presentasyonlar ise bir vakada menoraji, bir vakada düzensiz uterin kanamalar, 2 vakada pelvic ağrı ve bir vakada da asemptomatik pelvic kitle idi Ca-125 düzeyi sadece bir vakada yükselmişti.

**SONUÇ:** Düzensiz menstrüel siklusları olan genç kadınlarda, solid alanlar içeren hipervasküler adneksiyel kitle ile karşılaşıldığında, ayırıcı tanıda sklerozan stromal tumor düşünülmelidir. Tümör malign kitle izlenimi vermesine rağmen, radikal cerrahiye girişmeden once dikkatli davranılmalıdır.

**Anahtar Kelimeler:** Stromal tümor, Sklerozan, Adneksiyel kitle

## References

1. Peng HH. Chang TC. Hsueh S. Sclerosing stromal tumor of ovary. Chang Gung Med J 2003;26:444-8.
2. Chalvardjian A. Scully RE. Sclerosing stromal tumors of the ovary. Cancer 1973;31:664-70.
3. Youm HS. Cha DS. Han KH. et al. A case of huge sclerosing stromal tumor of the ovary weighing 10 kg in a 71-year-old postmenopausal woman. J Gynecol Oncol 2008; 19:270-4.
4. Bildirici K. Yalçın OT. Özalp SS. et al. Sclerosing stromal tumor of the ovary associated with Meigs' syndrome: a case report. Eur J Gynaecol Oncol 2004;25:528-9.
5. Terauchi F. Onodera T. Nagashima T. et al. Sclerosing stromal tumor of the ovary with elevated CA125. J Obstet Gynaecol Res 2005;31:432-5.
6. Jung NH. Kim T. Kim HJ. et al. Ovarian sclerosing stromal tumor presenting as Meigs' syndrome with elevated CA-125. J Obstet Gynaecol Res 2006;32:619-22.
7. Andrade LA. Gentilli AL. Polli G. Sclerosing stromal tumor in an accessory ovary. Gynecol Oncol 2001;81: 318-9.