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Sclerosing Stromal Tumor of Ovary: Presenting a Unilocular Cyst

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Abstract

Sclerosing stromal tumors (SSTs) are benign neoplasm of sex cord stromal tumor of ovary. The tumors occur predominantly in young female usually in the 2nd–3rd decades. Incidence is 2–6% of ovarian stromal tumors. It is usually unilateral. Other sex cord stromal tumors of ovary comprised fibromas, thecomas, granulosa cell tumor, Leydig cell tumor, and SST. Due to rarity of SST, most of the time it is not always possible to predict the presence of SST by clinical and radiological findings. The case is being presented for its rarity as the patient presented with unilocular cyst and minimal ascites.

Keywords: Pseudolobulation, Sclerosing stromal tumor, Sex cord stromal tumor

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Introduction

Sclerosing stromal tumors (SSTs) are a type of benign neoplasm of stromal component of ovary. SSTs account 2–6% of ovarian stromal tumors. More than 80% of SSTs occur during the 2nd–3rd decades with an average age of 27 years.[1] The SSTs are associated with estrogen or androgen secretion or both.[2] Most of the reported cases of SST are unilateral. All SSTs are managed by surgical enucleation and no recurrence has been reported after surgery.[3]

Case Report

A 16-year-old girl presented with menstrual irregularities, lower abdominal pain, and mild distension of abdomen for 5 months in the outpatient Department of Obstetrics/Gynaecology in KCGMCH, Karnal.

On physical per abdominal examination, a soft to firm vague mass was noted. The mass was non-tender. On radiological examination, a right adnexal solid unilocular cystic pelvic mass was revealed of size 13 × 11 cm, with unremarkable right fallopian tube, left ovary and left fallopian tube, uterus and cervix, and minimal ascites. All other routine hematological investigations, biochemical tests, tumor markers, and serum hormonal assay were within limits.

A right-sided salpingo-oophorectomy was performed and specimen was sent for histopathological examination in the Department of Pathology, KCGMCH, Karnal. The ascitic fluid was tapped and sent for cytological examination.

Pathological Findings

Grossly – on examination, a right side ovarian mass was submitted, measuring 11 × 9 × 3.5 cm, weighing 525 g, with attached right fallopian tube measuring 4.5 cm in length. The mass was gray-white, lobulated, and encapsulated. External surface was smooth and showed prominent vascular markings [Figure 1]. On cut section, a unilocular cyst was identified. Cyst was filled with mucinous material. The cyst lining was smooth. The wall was thickened and measured 1 cm. No papillary projections or hard solid areas were seen. Focal areas of necrosis and hemorrhage were identified [Figure 2].

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Microscopic Examination

Multiple sections examined showed a tumor showing pseudolobulations with cellular nodules separated by less cellular area of collagenous tissue and edematous connective tissue [Figure 3]. The cellular nodule showed admixture of fibroblasts and round vacuolated cells, some of which had signet cell appearance [Figure 4]. Nodules also showed some prominent thin walled staghorn-shaped dilated vessels [Figure 5]. Some focal areas showed myxomatous changes. The fallopian tube showed normal histology.

Smears prepared from ascitic fluid showed only mesothelial cells. No malignant cells were identified.

A diagnosis of SST of the right ovary was made.

Discussion

SST of ovary is a relatively infrequent neoplasm and represents only 2–6% of primary ovarian tumors.[4] The most common symptoms of SST include menstrual irregularities, lower pelvic pain, and abdominal distension, related to ovarian mass size. Virilization and anovulation may be present as they are associated with estrogen and androgen production.[2] SSTs are mostly hormonally inactive, if hormonally active, they are usually androgenic and occur most frequently during pregnancy. The etiology of the origin of SST is still not well established. According to a study by Damajanov *et al*., SSTs are derived from pluripotent immature stromal cells from cortex of ovary on the basis of ultrastructural features.[5] Tiltman

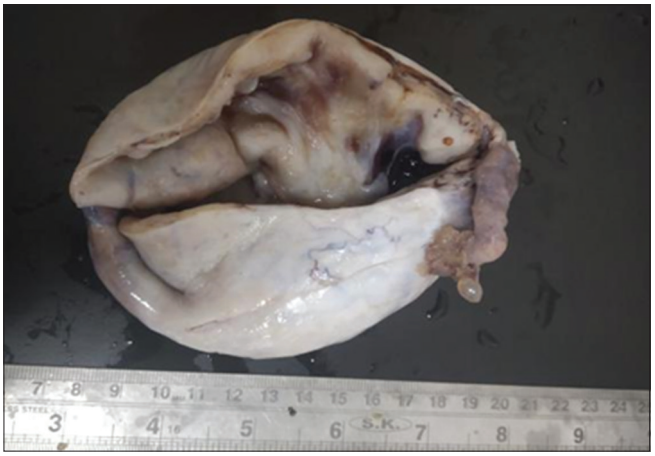

Figure 1: Gross specimen of ovary with thickened wall along with fallopian tube

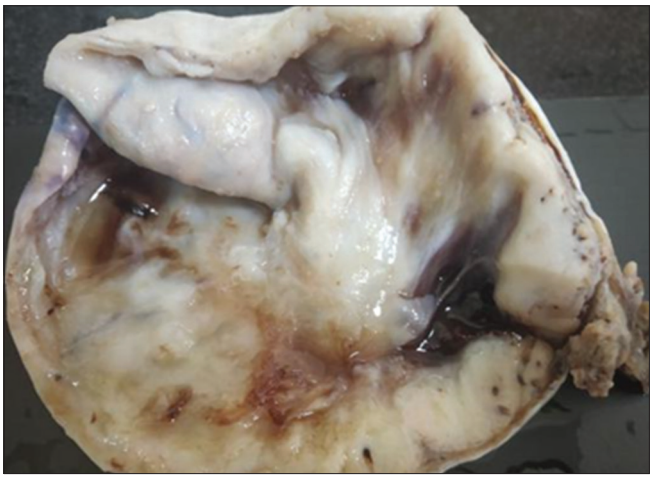

Figure 2: Cut section of ovary showing unilocular cyst, thickened wall, and focal areas of hemorrhage

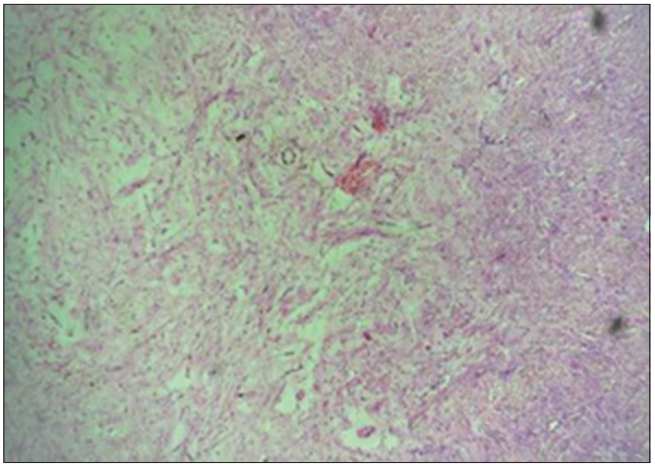

Figure 3: Both tumors with cellular and hypocellular areas (H and E stain, ×20)

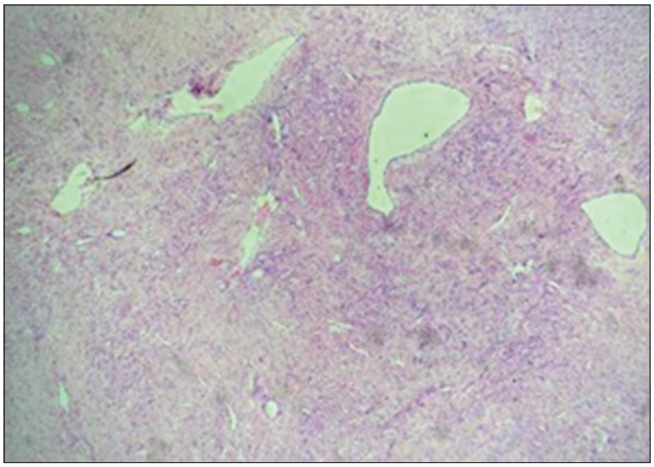

Figure 4: Spindle cells and round cells with clear cytoplasm (H and E stain, ×40)

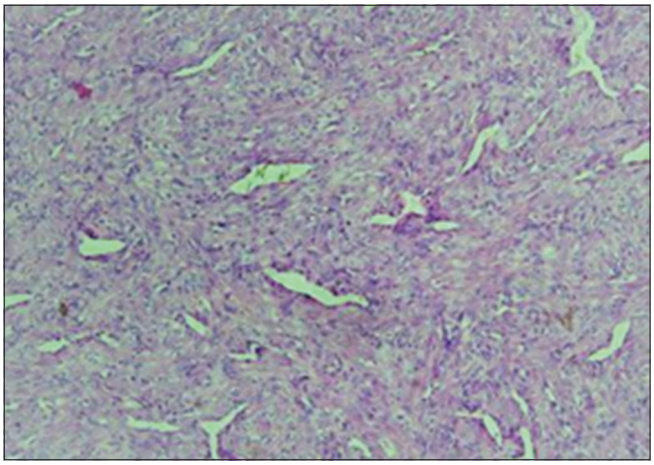

Figure 5: Dilated staghorn-shaped vessels (H and E stain, ×20)

and Haffajee postulated that SST and thecoma share common morphological features and antigenic determinants such as SMA and Vimentin.[6] Other serum biomarkers and biochemical essays remain normal but CA-125 was elevated in few cases because of repeated probing during per abdomen palpation.[4,7] Sonographic features include a multilocular cystic mass of heterogeneous or hypoechoic echogenicity with irregular thick septae. Early and strong peripheral enhancement is a key characteristic feature in distinguishing between SSTs and other types of sex cord stromal tumors.[8] On magnetic resonance imaging (MRI), they are known to be mainly heterogeneous solid tumors showing intermediate signal intensity with hyperintense foci corresponding to stromal edema on T2-weighted imaging (WI) and striking enhancement on contrast-enhanced T1WI.[9] Histologically, it is characterized by pseudolobulation pattern with cellular areas and hypocellular areas. Cellular areas comprise spindled fibroblast cells and oval to polygonal lipid containing vacuolated cells.[10] Some of the clear cell show signet ring morphology with edematous edema, maybe mistaken for signet ring cell of Krukenberg tumor of ovary, as they occur in the 6^{th}–7^{th} decades of life, with bilateral preponderance. Histologically, Krukenberg tumors lack pseudolobulation pattern.[4] Cellular areas of SST show many thin walled blood vessels mimicking a vascular tumor.[11]

SST has to be differentiated from other sex cord tumors. SSTs usually present in younger age female, that is, 2^{nd}–3^{rd} decades of life, whereas other ovarian sex cord stromal tumors occur in older age

Table 1: Differentiating features of sex cord stromal tumors of ovary[12]

Parameters	*Sclerosing stromal tumor*	*Fibroma*	*Thecoma*	*Steroid cell tumor*
Age	80% under 30 years	10% under 30 years	Average age 63 years	25% under 30 years
Function	Almost always absent	Absent	Typically estrogenic	Typically androgenic
Gross variegation	Yes	No	No	No
Pseudolobulation	Yes	Rare	Rare	No
Prominent ectatic vessels	Yes	Rare	Rare	Rare
Two cell types	Yes	No	Only in luteinized form	No
Hyaline plaques	No	Common	Common	No
Behavior	Benign	Almost always benign	Almost always benign	Sometimes malignant

group, that is, 5th–6th decades of life.[1] The other sex cord stromal tumors of ovary comprised of fibromas, thecomas, granulosa cell tumor, Sertoli-Leydig cell tumor, and SSTs [differentiating features are mentioned in the Table 1].

- SSTs show positivity for SMA, vimentin, calretinin, ER +, PR+, and alpha-inhibin.[6]
- SSTs are negative for S-100 and epithelial markers.[6]

Conclusion

Since, SSTs are a rare preprodence in ovarian neoplasm, it is not always possible to predict the presence of this tumor preoperatively on the basis of clinical and radiological investigations. However, MRI/CT scan leads over USG in prediction of SST. SST should be considered in young females with an ovarian mass and related symptom. Hence, the tumor is benign and treated successfully by surgical unilateral salpingo-oophorectomy. In this case, SST having a unilocular cyst and minimal ascites is a rare presentation. Histopathological diagnosis remains the gold standard for its diagnosis over the clinical and radiological investigations. IHC can be used for confirmation of the diagnosis.

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