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## CASE REPORT

# SCLEROSING STROMAL TUMOR OF OVARY IN YOUNG FEMALE: A CASE REPORT

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**ABSTRACT**

Sclerosing stromal tumor (SST) is an extremely rare benign tumor of the ovary that is derived from the sex cord stroma. It affects mostly young females in their second and third decade of life. Sclerosing stromal tumor of ovary most commonly presents as an abdominal mass with menstrual irregularities and pain in abdomen. Preoperative diagnosis of the tumor is difficult as it often mimics a malignant tumor. Oophorectomy is usually performed in many cases of young women, and diagnosis of SST is made based on post-operative pathological examination. Here, we report a case of SST of the ovary in an 23-year-old young female who came to JNMCH for treatment of primary infertility and abnormal uterine bleeding, she was diagnosed on the basis of histopathological report when she underwent cystectomy. Accurate preoperative imaging helped to perform minimally invasive surgery for SST.



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## INTRODUCTION

Ovarian Sclerosing stromal tumors are extremely rare tumor of ovary account for about 5-8% of all ovarian malignancies (Young, 2005). Till now <200 cases of these tumors have been studied in literature. SSTs are benign, ovarian neoplasm first described by Chalvardjian and Scully in 1973 (Chalvardjian *et al.,* 1973). These tumors usually are unilateral but rarely can be bilateral and predominantly affects young women in their second and third decade of life (Ozdemir *et al.,* 2014; Kaygusuz *et al.,* 2013). Most common clinical presentation of SST are menstrual irregularities, pain in abdomen and or lump in abdomen (Blake *et al.,* 2013). Some cases may present with anovulatory infertility and features of hirsutism. Earlier SST were considered to be inactive tumors but now several reports state that these are active tumor that produce the hormone (estrogen, androgen). These tumors synthesis dehydroepiandrosterone which on further steroid conversion causes irregular menstrual cycles, amenorrhea, infertility, precocious puberty, hirsutism (Kaygusuz *et al.,* 2013). The hormonal levels and sign and symptoms become normal and get resolved respectively following excision. SST cannot be diagnosed precisely in preoperative period on the basis of clinical and ultrasonography finding, histopathological examination and IHC are required for the confirmation of diagnosis (Rao *et al.,* 2018).

## CASE REPORT

A 23 year old female presented to JNMCH OPD with the chief complain of prolonged cycles and inability to conceive for 2 years. During her cycles she had bleeding throughout the month and she used to take 4-5 pads per day and bleeding stopped only after taking oral medication. There was no history of chronic cough with expectoration, ATT intake. Patient was a known case of Hypothyroidism and on regular medication (tab eltroxin x 125 microgram). Vital assessed showed a pulse rate of 90/min, blood pressure 124/84 mmHg and respiratory rate of 18/min. Per abdominal examination revealed a solid globular mass ≈14weeks, firm in consistency in right iliac region extending uptosuprapubic adnexa, firm in consistency, tender, felt separately from the uterus. Ultrasonographic findings were suggestive of right adnexal mass having solid and cystic component may be malignant. Patient underwent laparotomy and her postoperative period was uneventful.

### Per-operative findings

A large right ovariansolid cystic mass was found which was about 8x10 cm and highly vascularized. Cyst Enucleation followed by ovarian tissue reconstruction was done and tissue was sent for Histopathological examination and frozen section analysis. Mild ascitic yellow colored fluid was found in the

peritoneal cavity, sent for ADA and cytology. Rest adnexa was found normal. Abdomen was closed and drain put in the pelvis and patient was shifted out of the OT in satisfactory condition.

**Histo pathological examination:** Ovarian mass received grossly measured (10.5x 6x 5.3) cm, cut section shows solid areas with cystic changes. Solid areas appeared firm, homogenous, grey white in color with hamorrhagic foci.

**Microscopic examination**

Section shows lobular pattern of growth having hypo and hyper cellular areas separated by interlobular fibro collagenous tissue along with spindle cells and lipid containing round to oval cells resembling lutenized cells. Marked vascularity was seen in the form of hemangiopericytoma like foci. Focal areas with edematous changes and hyalanisation scleroses seen. Histo pathological features were indicative of sex cord stromal tumor of the ovary.

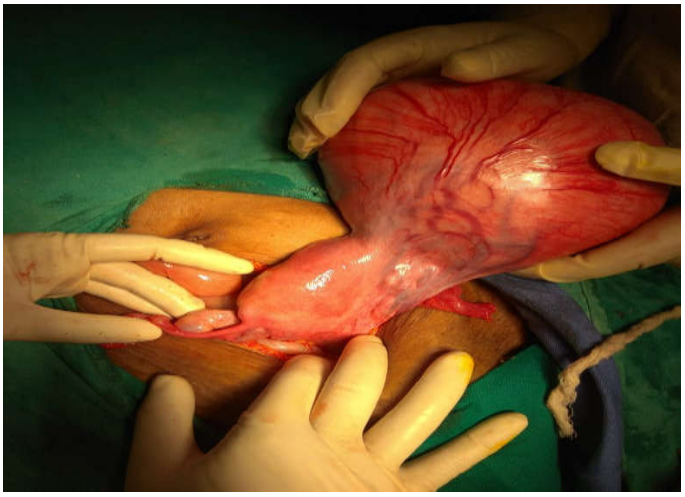
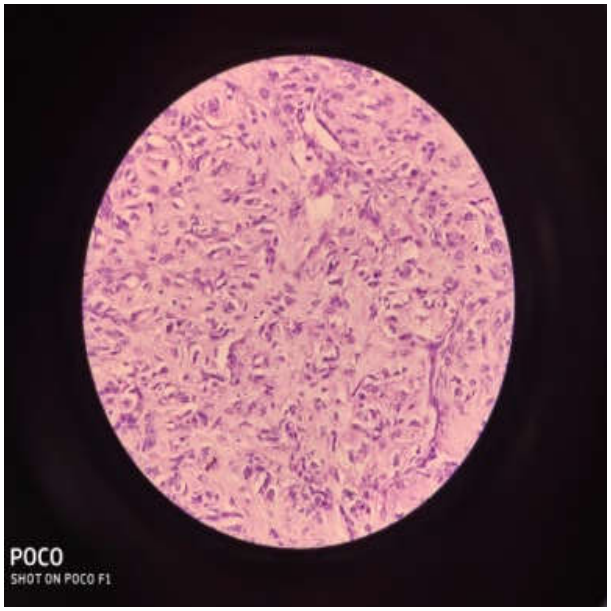


**Figure showing Per-operative findings**

IMMUNO HISTO CHEMISTRY work up was done for further diagnosis. Masson trichrome showed focal positivity for collagen. Scattered focal positivity for desmin immune stain was also found in the stroma. In view of Histo pathological changes and Immunohistochemistry finding a final diagnosis of SCLEROSING STROMAL TUMOR was made. During her follow up period she had conceived with in7 months of postoperative period.

## DISCUSSION

Ovarian Sex cord stromal tumors are relatively rare tumor of ovary account for about 5-8% of all ovarian malignancies (Atram *et al.,* 2014). Sclerosing stromal tumors very uncommon subcategory of sexcord stromal tumors comparising of 2-6% of allsexcord stromal tumors.[1]Sclerosing stromal tumors are benign, ovarian neoplasm first described by Chalvardjian and Scully in 1973 (Chalvardjian and Scully, 1973). SST usually are unilateral but rarely can be bilateral. The etiology of the SST is unknown, based on ultrastructure features SST are supposed to arise from pluripotent immature stromal cells of ovarian cortex. This group is distinct from the thecoma-fibroma group and steroid (lipid) cell tumors of sexchord stromal tumor clinically, pathologically, and radiographically.



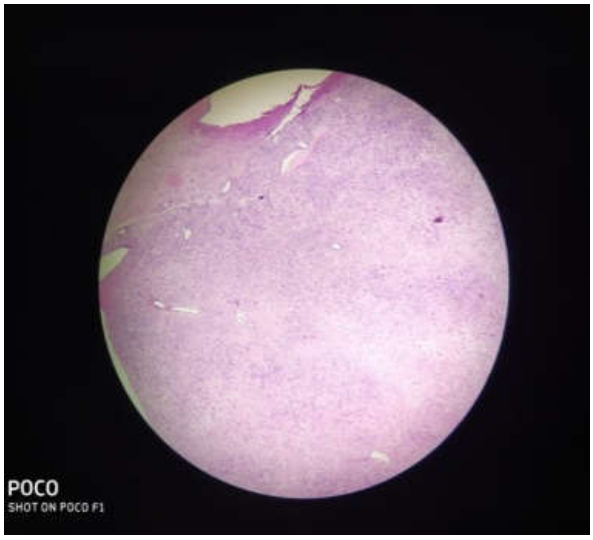




**Figure showing cells in diffuse pattern and as aggregates in abundant stromaand Cellular areas composed of spindle cells withvesicular nuclei and pale eosinophilic to clear cytoplasm**

Unlike the thecoma-fibroma group of ovarian stromal tumors, which tend to occur in the fifth and sixth decades, the SST predominantly affects young women in their second and third decade of life (Ozdemir *et al.,* 2014; Kaygusuz *et al.,* 2013).

Most common clinical presentation of SST are menstrual irregularities, pain in abdomen and lump in abdomen (Blake *et al.,* 2014). Some cases may present with anovulatory infertility and features of hirsutismthat resolves after removal of the tumor and is probably due to the hormone production by the tumor cells. Menstrual irregularities return to normal cycles after excision of tumor. Earlier SST were considered to be inactive tumors but now several reports state that these are active tumor that produce the hormone (estrogen, androgen). These tumors synthesis dehydroepiandrosterone and that when steroidogenesis occurred which causes irregular menstrual cycles, amenorrhea, infertility, precocious puberty, hirsutism (Kaygusuz *et al.,* 2013).

The hormonal levels and sign and symptoms become normal and get resolved respectively following excision. Certain parameters are required for accurate diagnosis of these ovarian tumors include history and examination such as patients age, sign and symptoms, histo pathological examination and immune histo chemistry (Rao *et al.,* 2018). In refrence to present case in which young female of 23 years presented to jnmch with chief complaints of menorrhagia and primary infertility with a palpable lump in right iliac region extending upto supra pubic region of abdomen. The final diagnosis was that of sclerosing stromal tumour of the ovary which was made in her post operative period through histopathological examination and immunohistochemistry. Post-operative period was uneventful. Characteristic histological finding of the SST of ovary is the pseudolobular pattern that is formed by the cellular nodules that are separated from each other by hypocellular, oedematous and collagenous stroma.

The hemangiopericytomatous pattern-like dilated vascular structures are the characteristics of cellular areas, and the luteinised theca-like cells with vacuolised cytoplasm and fusiform fibroblast-like are the characteristics of hypercellular areas (Clement *et al.,* 1994). Inhibin has been shown to be useful marker for ovarian sex cord stromal tumours. Inhibin is a specific, but less sensitive marker than calretinin in the diagnosis of ovarian sex cord-stromal tumours. In addition, a correlation was observed between the calretinin and α-inhibin expressions and the luteinisation level of tumour cells (Zekioglu *et al.,* 2010). Also, inhibin and calretinin have been shown to be more sensitive and specific marker than CD99, A103 (melan-A), CD10 and WT-1for ovarian sex cord stromal tumours (Zekioglu *et al.,* 2010; Ismail and Walker, 1990).

H&E staining is enough to diagnose SST and fibromas. Immunohistrochemistery may not be required as both the tumors have distinctive features. It is only required when tumor has mixed histomorphological pattern. Sst are positive for smooth muscle actin and desmin and its immunohistochemistry shows negativity for epithelial markers like S100 protein and CD99 (Lam and Geittmann, 1988). SST cannot be diagnosed precisely in preoperative period on the basis of clinical and ultrasonography finding but their distinct histopathology appearance and IHC confirms the diagnosis. Although these tumors were initially believed to be non-functional (Young *et al.,* 2005; Marelli *et al.,* 1988), there have been more recent reports suggesting the presence of hormone production both from a biochemical point of view and associated clinicalManifestations of infertility and irregular menses (Suit *et al.,* 1988; Joja *et al.,* 2001).

## Conclusion

Patient age, unilaterality of tumor and distinct histopathological, IHC findings are essentially characteristic for diagnosis of ovarian SST. Although rare SSTs are rare tumors but possibility should be kept in mind in young female with ovarian mass as they can be treated successfully by enucleation or U/L ovariotomy.

**Conflict of Interest statement**: None

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