

Sclerosing Stromal Tumour of Ovary : A Case Study with the Review of Literature

Vijay Kumar Jha^{1*}, Alka Rai², Mohan Kumar¹, Shivi Jain³, Madhu Jain⁴

¹Dept. Of Pathology, Institute Of Medical Sciences, Banaras Hindu University, Varanasi, India

²Deptt. Of Obstetrics and Gynecology, Heritage Institute of Medical Sciences, Varanasi, India

³Deptt. Of Radiodiagnosis and Imaging, Institute Of Medical Sciences, Banaras Hindu University, Varanasi, India

⁴Deptt. Of Obstetrics and Gynecology , Institute Of Medical Sciences, Banaras Hindu University, Varanasi, India

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ABSTRACT

Sclerosing stromal tumour of ovary is a rare and distinct type of sex cord stromal tumours with benign course and excellent prognosis. It affects mostly young females in their second and third decade of life. Nearly all the reported cases of sclerosing stromal tumour of ovary are unilateral with menstrual irregularities, pain abdomen and lower abdominal mass as commonest clinical presentations. Most of the cases are hormonically inactive, however, hormone production has been documented in various studies. Histopathology and IHC confirms the diagnosis. Surgical excision is the treatment of choice. No recurrence has been reported yet. We are describing a histopathologically confirmed case of a young girl who presented with complaints of menorrhagia and pain abdomen. Radiological examination and peroperative findings raised the suspicion of malignancy. It is important to consider sclerosing stromal tumour in young females presenting with complex ovarian mass.

***Corresponding author:**

Dr. Vijay Kumar Jha, Senior resident, Deptt. Of Pathology, Institute Of Medical Sciences, Banaras Hindu University, Varanasi, India, 221005

Phone: +91 8960202107

E-mail: vj.bhu07@gmail.com



Introduction

Sclerosing stromal tumour (SST) is an extremely rare benign ovarian neoplasm, a subtype of sex cord stromal tumours comprising 1.5 - 6% of all ovarian sex cord stromal tumours.^[1] Fewer than 208 cases of SST have been described in literature since its first description in 1973 signifying rarity of this entity.^[2]

SST is the disease of young female occurring in their second and third decade of life.^[2,3,4] About 80% of the cases are seen in patients below 30 years of age.^[1,2,4,5] Patients usually present with menstrual irregularities, pelvic pain and abdominal lump.^[6] Some cases show features like amenorrhoea, anovulation, endometrial hyperplasia and hirsutism related to hormone production.^[2,4,7] Majority of the cases of SST are hormonally inactive, however, there are reports of androgen and estrogen excess in this tumour.^[2,3]

Distinct clinical, histopathological and radiological features differentiate SST from other ovarian sex cord tumours.^[2,4,8] Histopathological examination and IHC confirms the diagnosis.^[2] Sclerosing stromal tumour, a solid cystic mass with rich vascularity is often misdiagnosed as malignancy on ultrasonography and other radiological examinations.^[2,4] Proper diagnosis is essential for preventing morbidity associated with extensive surgical intervention done in cases of ovarian malignancy.

Herein, we describe a case of SST in an 18 year old female who presented with polymenorrhagia and abdominal pain.

Case Report

An eighteen year old unmarried girl presented with complaints of polymenorrhagia for six years and pain abdomen for two years. On examination there was a large firm lower abdominal mass. Past and present medical history were unremarkable.

Patients haematological and biochemical profiles were within normal limit except mild anemia (Hb -12gm/dl). Serum tumour markers were within normal range (AFP -1.0 ng/ml, CA125 - 10.1U/ml, β HCG - 1.0 mIU/l & CEA - 3.63 ng/ml). Hormonal assay was not done in this case.

Radiological findings: USG revealed a solid cystic right adnexal mass of 7.8x6.3 cm with normal left ovary, uterus and endometrium. CT scan showed a well defined complex solid cystic mass measuring 6.7x5.4x5.9 cm with many internal septae. MRI showed a relatively defined heterogenous right adnexal mass measuring 9.8x6.1x8.2cm with solid cystic areas without evidence of haemorrhage and necrosis. Ipsilateral ovary could not be identified separately from the mass (Fig1A). Radiological examination did not reveal any fluid collection in peritoneal

cavity or pouch of Douglas. All other visceral organs were normal and there was no evidence of any mass lesion.

Histopathology: gross specimen (Fig.1B) measured 8x8x4 cm with intact capsule and smooth external surface. Cut surface was pale white in colour, firm to hard in consistency, predominantly solid with scattered cystic areas. Multiple tiny yellowish specks were also noted. Normal ovarian tissue was not identified. Fallopian tube present on the external surface of the mass measured 6x1x0.6 cm and was unremarkable.

Specimen was fixed in 10% buffered neutral formalin and processed according to standard laboratory protocols. Haematoxylin and eosin (H&E) and PAS stained slides were examined. Immunohistochemistry for Inhibin, Vimentin, SMA, VEGF, CK, EMA, CD34, estrogen receptor (ER) and progesterone receptor (PR) was done.

Microscopic examination revealed an encapsulated tumour having cellular and hypocellular areas imparting a pseudolobular appearance (Fig.1C). Cellular areas showed dual population of cells comprising of spindled fibroblastic cells and round, oval to polygonal cells with clear to eosinophilic cytoplasm and round regular nuclei (Fig.1D). Cellular lobules were separated by dense collagenised and oedematous stroma (Fig.1E). Many thin walled blood vessels, some with branching simulating hemangiopericytomatous pattern (Fig.1F) were also seen in the cellular areas. Normal ovarian stroma was identified in few sections. No mitotic activity was noted. PAS stain was negative.

IHC findings: tumour cells showed positive staining for Inhibin (fig.2A), PR (Fig.2B) vimentin (Fig.2C), SMA (Fig.2D), VEGF (Fig.2E) and CD34 (Fig.2F) while CK, EMA and ER were negative.

Based on above clinical, radiological, histopathological and IHC findings a diagnosis of sclerosing stromal tumour of ovary was established.

Discussion

Ovarian tumours are very uncommon in adolescent girls and majority of these tumours are of germ cell origin.^[3] SST of the ovary is an extremely rare and distinct subtype of sex cord stromal tumours.^[3] Most common clinical presentations are menstrual irregularities, pain abdomen and palpable abdomino-pelvic mass.^[2,3,4,6,7] Some of the patients present with anovulation, infertility and features of hirsutism that resolves after removal of the tumour^[2,7] and is probably due to hormone production. Both androgen and estrogen production has been described.^[2] Return of normal menstruation in patients with menstrual irregularities

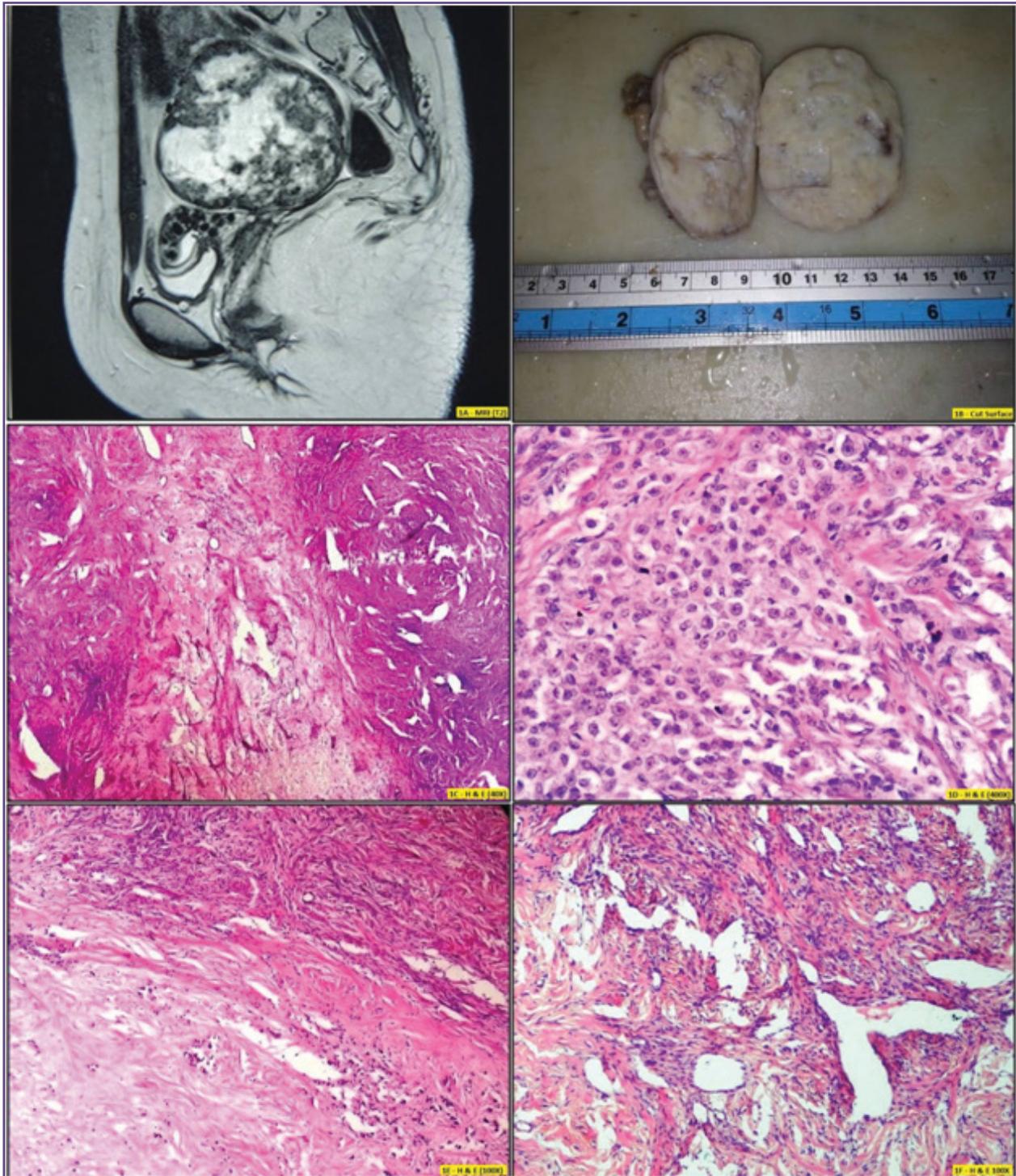


Fig. 1: Panel of photographs displays MRI image (T2 weighted) showing heterogeneously enhancing solid cystic ovarian mass without evidence of haemorrhage and necrosis (Fig.1A), cut surface of the mass showing predominantly solid pale white areas with interspersed cystic spaces (Fig.1B), micrograph shows pseudolobular arrangement with cellular nodules and hypocellular interlobular areas (Fig.1C, H&E stain, 40X), cellular area having clear theca like cells with round regular nuclei (Fig.1D, H&E stain, 400X), Interlobular hypocellular collagenized oedematous stroma (Fig.1E, H&E stain, 100X), Cellular nodule showing marked vascular proliferation, some with branching (Fig.1F, H&E stain, 100X)

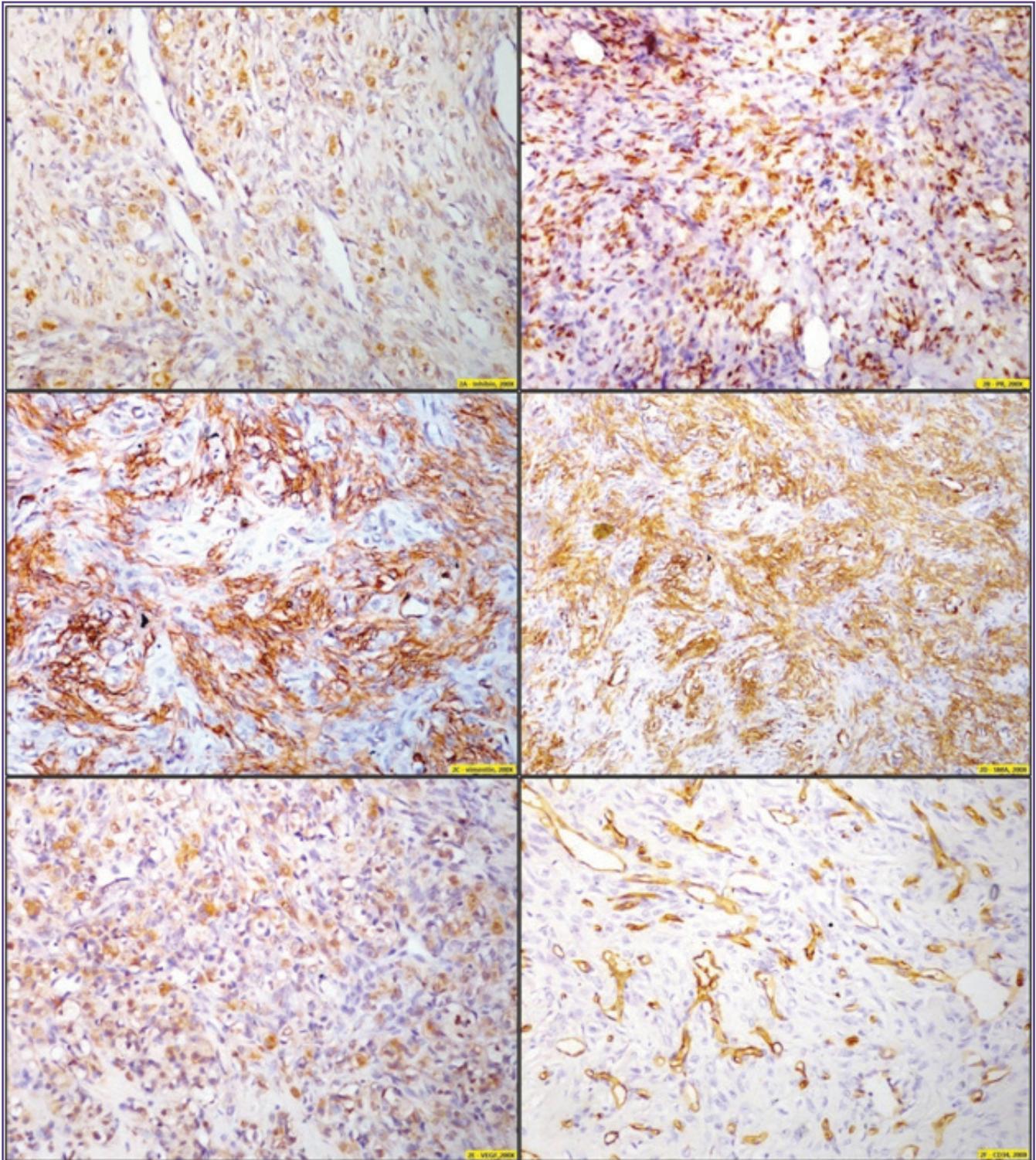


Fig.2 Panel of photographs shows immunohistochemical expression of inhibin (Fig.2A, 200X) progesterone receptors (Fig.2B,200X), vimentin (Fig.2C,200X) SMA (Fig.2D, 200X), VEGF (Fig.2E, 200X) and CD34 (Fig.2F, 200X)

following surgical excision of the mass has been described.

^[2] Sclerosing stromal tumour is unilateral mostly.^[2,4]

Etiology of this tumour is not very well defined, however, ultrastructural features suggest origin from pluripotent immature myoid stromal cells of ovarian cortex.^[3,9]

Histopathology reveals characteristic pseudolobular arrangement with cellular areas and hypocellular interlobular areas.^[2,6] Cellular lobules consists of dual population of cells comprising of spindle fibroblastic cells and round, oval to polygonal lipid containing cells.^[2,4] In some cases clear cells show signet ring cell morphology with eccentric nuclei. Such cases need differentiation from Krukenbergs tumour.^[1,4] Cellular areas also show many thin walled blood vessels mimicking vascular tumours.

^[3,4] Interlobular areas show fibrosis, collagen deposition and oedema.^[2] Oedema in SST is focal or confined within interlobular areas in contrast to massive ovarian oedema.

Immunohistochemical analysis of SST shows positive staining for alpha inhibin, calretinin, SMA, vimentin, ER, PR and VEGF.^[2,3,4,10-12] Some authors have reported VEGF positivity in round, clear theca like cells.^[6] Epithelial markers CK and epithelial membrane antigen (EMA) are negative.^[2,4,5] In various studies inhibin, calretinin, alpha glutathione S-transferase (α -GST) and CD34 were found useful to differentiate SST from thecoma, fibroma and other sex cord stromal tumours.^[2,5,10] CD34 highlights endothelium of proliferated vascular channels that differentiates SST from thecoma and fibroma.^[2] In our case SMA, vimentin, inhibin, PR, VEGF and CD34 were positive while CK, ER, and EMA were negative. Immunohistochemical studies in SST show variable results, however, predominant positivity for inhibin, calretinin, SMA and vimentin suggests sclerosing stromal tumour of ovary.^[2]

Cytogenetic studies using FISH technique show trisomy of chromosome 12 in about 13-21% of the cases while in one study trisomy 16 was found.^[6,13]

This tumour needs differentiation from other sex cord stromal tumours like fibroma, thecoma, Juvenile granulosa cell tumour, metastatic carcinoma, Krukenbergs tumour, massive ovarian oedema and vascular tumours.^[2,4] Characteristic clinical presentation, radiological, histopathological and immunohistochemical profile differentiates SST from other benign and malignant mimickers.^[8]

It is difficult to diagnose this tumour based on USG and other radiological investigations due to complex solid cystic appearance and rich vascularity simulating malignant ovarian mass.^[2,4] However, MRI findings are more specific

in differentiating SST from malignancy and other sex cord stromal tumours.^[4] MRI show pseudolobulation having low intensity cellular nodules against high intensity stroma on T2 weighted images.

Conclusion

Despite of its rarity, SST needs consideration in Young females presenting with unilateral solid cystic complex ovarian mass and related symptoms as it has benign course and very good prognosis with less aggressive surgical treatment. Characteristic histopathology and immunohistochemistry establishes the diagnosis.

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Competing Interests

None declared

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