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Case Report

Ovarian Sex Cord Stromal Tumor with Annular Tubules - A Case Report & Review of Literature

Archana S. Bembde[1], Irmeen Manzoor[2*], Shweta Somani[3], Smita S Mulay[4]

[1]Lecturer, [2]Senior Resident, [3]Junior Resident, [4]Head of Department,
Department of Pathology, MGM Medical College & Hospital, Aurangabad, Maharashtra, India.

[*]Correspondence Email: drirmeenm@gmail.com

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ABSTRACT

Ovarian sex cord stromal tumor with annular tubules (SCTAT) is a distinctive, rare subtype of sex cord stromal tumor of the ovary, predominant component of which has morphological features intermediate between that of granulosa cell and sertoli cell. The majority of ovarian SCTAT are benign. So far, malignant behavior in SCTAT has been reported only in sporadic cases. We have presented a case of SCTAT in a 45 year old lady (P_3A_1) with no association of Peutz-Jegher (P-J) syndrome. The patients chief complaints were post-menopausal bleeding since 1 year on and off along with menorrhagia. History of amenorrhea five years back followed by heavy bleeding. Dilatation and curettage was done and bleeding was relieved. Thyroid profile showed features of hypothyroidism (TSH=20 uIU/ml). USG findings showed antiverted bulky uterus with cystic endometrial hyperplasia. MRI abdomen was suggestive of intensely enhancing solid tissue mass lesion in the left adnexa, features suggestive of ovarian mass. Panhysterectomy was done. Grossly uterus and right adnexa appeared to be normal. Left ovary enlarged, measuring 8x5x4cms with attached fallopian tube. Cut section yellowish, lobulated, homogenous, solid with no cystic areas. Microscopy revealed sex cord stromal tumor with annular tubules of granulosa cell type. PAS stain supported the diagnosis.

Keywords: SCTAT, sex cord stromal tumors, annular tubules, ovarian tumors.

INTRODUCTION

Robert Scully in 1970 first described 13 cases of this unusual variant of sex cord stromal tumor.[(1)] Young et al., reviewed a large series (74 cases) including those associated with P-J syndrome.[(2)] In one third of the cases, hyperestrinism and P-J syndrome association was seen.[(3)] SCTATs with P-J syndrome are small, <3 cm, benign, bilateral, and multicentric in young women, whereas, in the absence of this syndrome, it is seen as huge, unilateral tumor, and approximately one-fifth (15-20%) tend to be malignant,[(1-5)] comprising only 1.2% of all cases of ovarian cancer.[(6)]

Ovarian sex cord tumor with annular tubules (SCTAT) is an unusual variant of sex cord stromal tumors of the ovary.[(2)] The origin of this tumor remains unclear, with some favoring granulosa cell derivation theory,[(7,8)] and others favoring sertoli cell origin. [(9-11)] Also, there have been some

postulations that the tumor is composed of pluripotent stem sex cord cells of the gonads which have the potential for differentiating into either granulosa or sertoli cells.[12,13]

CASE REPORT

A 45 year old multiparous lady (P_3A_1) was admitted for post-menopausal bleeding since 1 year, on and off along with menorrhagia. History of amenorrhea five years back followed by heavy bleeding. Dilatation and curettage was done and bleeding was relieved. Investigations on present admission showed following features. Thyroid profile showed features of hypothyroidism (TSH=20 µIU/ml). All the hormone levels-estradiol, progesterone and testosterone were within normal range. USG findings showed antiverted bulky uterus with cystic endometrial hyperplasia. MRI abdomen was suggestive of intensely enhancing solid tissue mass lesion in the left adnexa, features suggestive of ovarian mass. Panhysterectomy was done. Grossly uterus and right adnexa appeared to be normal. Left ovary enlarged measuring 8x5x4cms with attached fallopian tube and bosselated external surface. Cut section yellowish, lobulated, homogenous, solid with no cystic areas (Fig1a & 1b). Microscopically, the cervix showed squamous metaplasia and mild dysplastic features. No adenoma malignum or mucinous metaplasia of endometrial or tubal epithelium was seen. Endometrium showed features of simple cystic endometrial hyperplasia. Left ovary was replaced by solid tumor composed of round to oval granulosa cells with vesicular nuclei and occasional nuclear grooving. Cytoplasm was moderate to scant and eosinophilic. Cells were arranged in sheets, nests, micro follicular, macro follicular, and insular pattern (Fig2a, 2b & 3). Simple and complex tubules encircling nodules of hyalinised PAS-positive basement membrane-like material were seen (Fig4a & 4b). Nuclei showed antipodal arrangement with focal sertoliform pattern. Luteinisation was evident in areas. A diagnosis of SCTAT of granulosa cell type was rendered.

Fig 1a: Gross photograph of panhysterectomy specimen with enlarged ovary measuring 8x5x4cms with bosselated external surface.

Fig 1b: Photograph of cut section of ovary yellowish, lobulated & solid.

DISCUSSION

SCTAT was documented to be oestrogen- progesterone-secreting tumor based on the observation of glandular atrophy, decidual change of endometrial stroma, and assays of steroid hormone.[3] Age at presentation was 20-30 years with a mean age of 20.6 years.[3] Symptoms suggestive of hyperestrinism such as meno-metrrhagoia, followed by persistent amenorrhea, postmenopausal bleed, sexual precocity, and/or pelvic mass were

presenting features.[1-6,12] Association with P-J syndrome and adenoma malignum of cervix is known.[1-6,12] When patients with ovarian tumor present with amenorrhea preceded by meno-metorrhagia, endometrial sampling should be done to look for glandular atrophy and decidual change to rule out the possibility of SCTAT. Our case presented at the age of 45 years with post-menopausal bleeding. She had no associated P-J syndrome or adenoma malignum of cervix. Setoli-Leydig cell tumor presents with similar symptoms and may morphologically simulate SCTAT. Steroid assay including estradiol, progesterone, and testosterone are helpful in differentiating the two. In Setoli-Leydig cell tumor, all three-estradiol, progesterone, and testosterone levels are elevated. If both estradiol and progesterone are elevated with normal testosterone levels, the diagnosis of SCTAT is ascertained.

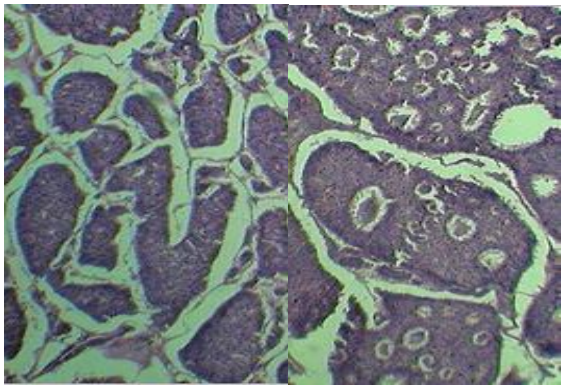

Fig 2a: Microscopic examination showing sheets & nests of cells arranged in microfollicular, macrofollicular & insular pattern. (H&E x100).

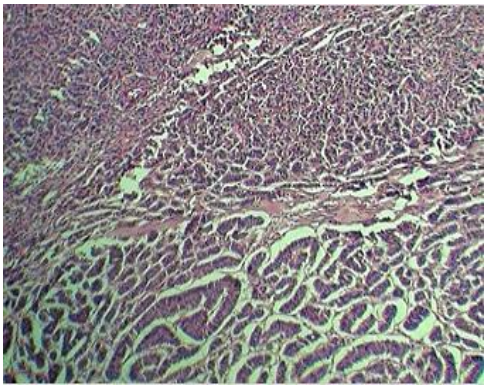

Fig 2b: Microscopic examination show sheets & nests of cells arranged in insular pattern. (H&E x100)

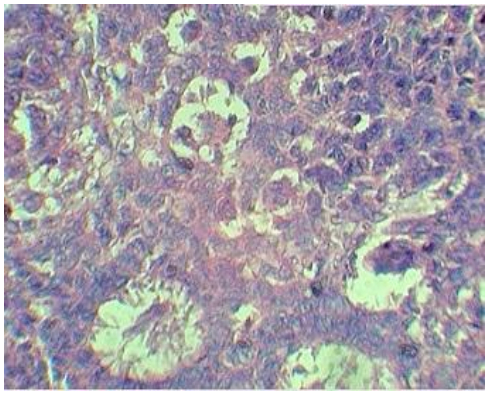

Fig 3: High power view showing follicular pattern & occasional grooving of nuclei. (H&E x400)

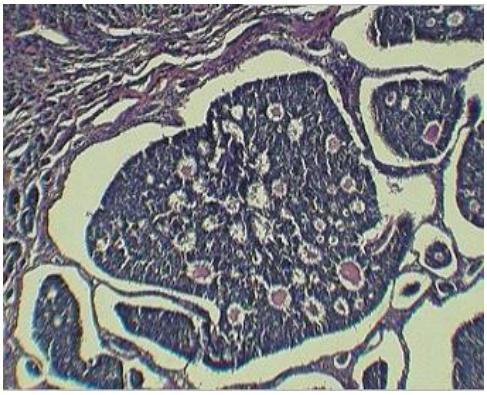

Fig 4a: Special stain PAS positive for peripheral basement membrane & hyaline globules. (H&E x100)

Morphologically, these tumors are solid, tan to yellow colored with few tiny cysts and focal calcification. Microscopically, circumscribed epithelial nests composed of ring-shaped tubules are seen with antipodal nuclear arrangement encircling hyaline globules, which is continuous with the basement membrane. The rings have two patterns: single tubule with a central rounded hyaline mass and complex communicating tubules encircling multiple hyaline masses. Our case showed similar morphologic and microscopic features. Previous reports of both benign and malignant SCTAT, including both sporadic and P-J syndrome-associated tumors, have

described additional histological patterns such as sertoliform tubules, endometrioid areas, and foci of granulosa cell-like differentiation. On the basis of such findings, some authors have suggested that SCTAT represent variants of either Sertoli cell or granulosa cell tumors, whereas others regard SCTAT as a distinctive neoplasm with features intermediate between those of Sertoli cell and granulosa cell tumors.[12] Mullerian-inhibiting substance is a glycoprotein hormone produced by fetal sertoli cells. Sex cord stromal tumors secrete a large amount of this hormone. The degree of the elevation correlates well with the tumor burden.[1-4] Gustafson et al., have reviewed over 5,60,000 ovarian biopsies obtained over a period of 40 years and stated that the measurement of this substance may help detect persistent or recurrent disease.[4] Malignant SCTAT seems to spread mainly via the lymphatics with typical sites of tumor metastasis being the pelvic, para-aortic, and supraclavicular lymph nodes. Other sites of tumor recurrence and metastasis include the retroperitoneum, peritoneum, liver, kidney, and lung. Unilateral salpingo-oopherectomy together with ipsilateral pelvic and para-aortic lymphadenectomy is suggested as an effective treatment for SCTAT. Radiotherapy is reserved for local recurrence and distant metastasis.[3-6,12]

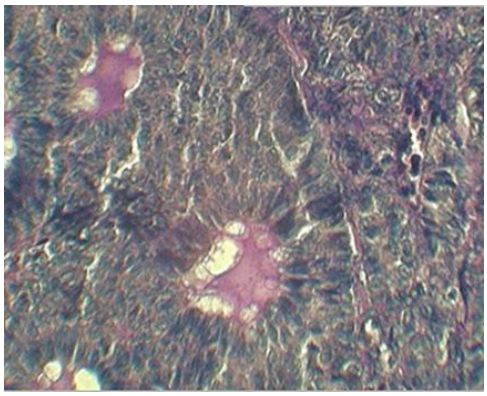

Fig 4b: Special stain PAS positive for peripheral basement membrane & hyaline globules. (H&E x400).

Although there is considerable disagreement as to whether the cells in a SCTAT are of granulosa[7,8] or sertoli type,[9-11] there is some consensus that the tumor is composed of primitive cells of sex cord origin which have a potential for differentiating into either granulosa or Sertoli cells.[13,14] Some authors, based on light microscopic finding that is, the nuclear grooves, the central hyaline bodies resembling Call Exner bodies, the macrofollicular pattern, the formation of solid nests and the absence of tubular structures which had true lumen, have suggested that SCTAT differentiated into granulosa cell line.[7,8,15] Those who believed that the SCTAT cells derived from sertoli cells based their hypothesis on the morphological resemblance of SCTAT to sertoli cells demonstrated by the pattern of palisading of the nuclei, the tubular arrangement with cells of pale cytoplasm and the presence of cytoplasmic lipid.[9-11]

Immunohistochemistry staining of SCTAT show positivity for vimentin and cytokeratin.[16,17] The authors favouring granulosa cell differentiation of SCTAT have presented some untrastructural evidence: the deeply indented nuclei, numerous randomly distributed fibrils, abundant desmosomes attaching the interdigitating processes or adjacent cells, and complexes of fibrils with desmosome, the resemblance of the concentric layers of basal lamina of the central hyaline bodies which were characteristic and consistent features of both SCTAT and granulosa cell tumors.[7,8,18] Those who believed that SCTAT cells derived from sertoli cells based their opinion on the findings of Charcot-Bottcher filaments, the presence of intra-cytoplasmic lipid, the tubular arrangement with occasional findings of lumen and microvilli.[9-11]

SUMMARY

A 45 year old multiparous lady (P_3A_1) was admitted for post-menopausal bleeding since 1 year, on and off along with menorrhagia. MRI abdomen was suggestive of intensely enhancing solid tissue mass lesion in the left adnexa, features suggestive of ovarian mass. Panhysterectomy was done. A diagnosis of SCTAT of granulosa cell type was rendered. Patient responded well to our treatment.

CONCLUSION

Ovarian sex cord stromal tumor with annular tubules (SCTAT) is a distinctive, rare subtype of sex cord stromal tumor of the ovary. SCTATs with P-J syndrome are small, <3 cm, benign, bilateral, and multicentric in young women, whereas, in the absence of this syndrome, it is seen as huge, unilateral tumor, and approximately one-fifth (15-20%) tend to be malignant, comprising only 1.2% of all cases of ovarian cancer. In our case it was not associated with P-J syndrome and was sporadic benign ovarian SCTAT of granulosa cell type.

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Conflicts of interest: Nil

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