Case Report

Non-syndromic unilateral sex cord tumor with annular tubules in an adolescent female: A Rare Diagnosis

Vaishali Rohilla[1,*], Shalini Gupta[1], Manju Raghava[1], Vishal[2]

[1]Department of Pathology, Mahatma Gandhi Medical College and Hospital, Sitapura, Jaipur, Rajasthan, India

[2]Department of Pathology, SHKM Medical College and Hospital, Nuh, Haryana, India

***Correspondence:** vvvvaishali@gmail.com

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Abstract

Sex cord tumor with annular tubules (SCTAT) is a rare tumor of ovarian origin which is usually benign in origin but can show extraovarian spread in some cases. Here we present a case of 14-year-old female with abdominal pain which on radiology show unilateral ovarian cyst. Based on histopathological findings, diagnosis of SCTAT was confirmed. Non syndromic unilateral SCTAT usually presents in older women but can also be seen in younger age group and it should always be considered a differential diagnosis in ovarian tumors.

Keywords: sex cord tumor; annular; tubules; peutz-jeghers

Introduction

Sex cord stromal tumor (SCTAT) with annular tubules is a rare neoplasm. It constitutes <1% of all sex cord tumors. [1] It could be associated with Peutz-jeghers Syndrome which usually occurs in younger age and presents with bilateral involvement. Cases not associated with the syndrome usually presents in older patients, predominantly cystic, unilateral and have malignant potential. [2]

We report this rare case on non-syndromic unilateral SCTAT presenting in an adolescent female.

Case Report

14-year-old female presented with chief complaint of pain in lower abdomen only (right iliac fossa). There was no association with nausea, vomiting, weight loss, or loss of appetite. Patient attained menarche at the age of 13 years and had irregular menstrual cycles since then. On per abdominal examination- a 20-week size, tense cystic mass was present in right side lower abdomen region which was tender on palpation.

Routine laboratory investigations were normal. Hormone profile: Estradiol (E2- 151 pg/ml), Progesterone (P- 1.2 ng/ml), Anti-mullerian hormone (AMH- 20.2 ng/ml) were raised. However, CA 125, CEA, CA19-9 and Serum LDH were within normal range.

Radiological investigations: MRI showed a multiseptated cystic mass measuring 88 x 83 x 78 mm in right ovary which was suggestive of serous cystadenoma. Uterus and left ovary were normal. Ultrasound findings showed a well-defined cystic lesion measuring 92 x 62 x 93 mm with multiple internal cysts in right adnexa likely arising from right ovary suggestive of mucinous cystadenoma.

After this patient underwent laparotomy followed by right salpingo-oophorectomy. 10 ml peritoneal fluid was drained which was sent for cytological examination. 10 ml of fluid was received for fluid cytology. Hematoxylin & eosin-stained slide was prepared and examined for any malignant cells. It showed presence of lymphocytes and mesothelial cells against a background of proteinaceous fluid; no malignant cells were seen.

Intra-operatively: Right ovary had an 8 x 9 cm cyst containing clear fluid. Right ovary and fallopian tube were removed and sent for histopathological examination. Based on these findings, a diagnosis of right ovarian mucinous cystadenoma, FIGO stage 1A was made.

Macroscopic examination revealed a 7.5 x 6.5 x 3 cm ovarian cystic soft tissue piece with attached fallopian tube. External surface was smooth and encapsulated. No capsular breach was present. No surface involvement was present. On cut it had multilocular cysts which showed clear fluid and few haemorrhagic areas. Attached fallopian tube was unremarkable.

Microscopic examination revealed presence of multiple cysts lined by stromal cells along with solid areas showing nests composed of tubules which encircle hyaline basement membrane material. Cells were columnar with foamy cytoplasm, mild nuclear atypia, nuclear grooving, and inconspicuous nucleoli. Cells had antipodal nuclear distribution. (Figure 1)

Immunohistochemistry (IHC) was not performed as SCTAT has distinctive histomorphological features which clearly defines its diagnosis. The patient did not show any features of Peutz-jeghers Syndrome on physical examination. Also, there was no family history of such lesions. For follow-up, patient underwent ultrasonography after 3 months of surgery. No recurrence was seen yet.

Discussion

SCTAT is a rare tumor type in sex cord stromal tumors. It was first described by Scully et al in 1970. They described the possibility of origin of this tumor from granulosa cells and growth pattern resembling Sertoli cells. There are 2 types of SCTAT: 1) Syndromic SCTATs associated with Peutz-jeghers Syndrome; 2) Non-syndromic SCTAT. [3] Syndromic cases are associated with germline STK11 gene mutation on chromosome 19p13.3 and are usually benign. Non-syndromic cases might show extraovarian spread in some cases. [1]

It is an incidental finding in most of the cases and presentation varies with age groups. In young girls- precocious puberty, reproductive women- menstrual irregularities or amenorrhea, post-menopausal women- bleeding. Symptoms are associated with estrogen and progesterone secretion. Hormonal stimulus could lead to bleeding, endometrial hyperplasia, polyps, and intraepithelial lesions. [2]

Macroscopically SCTATs could vary from small, bilateral lesions in syndromic cases to large, unilateral masses in non-syndromic cases. These masses are usually solid and cystic, yellow to tan with some haemorrhagic and necrotic areas. [1]

Microscopically there is presence of variably sized nests which are comprising of simple or complex tubules encircling hyaline basement membrane like material. Cells are tall having pale cytoplasm and basal nuclei. In syndrome associated cases there could be presence of extensive calcification, solid proliferation of different sex cord cells or Sertoli like tubular pattern seen occasionally. Essential criteria for diagnosis is tubular pattern with antipodal distribution of nuclei and basement membrane like material. Tumor cells show positivity for calretinin, WT1, inhibin, SF1, FOXL2, CD56 and negativity for EMA & CD10. [1]

Differential diagnosis for SCTATs would be Sertoli cell tumor, granulosa cell tumor and gonadoblastoma.

Very few cases of SCTAT are reported in Indian literature:

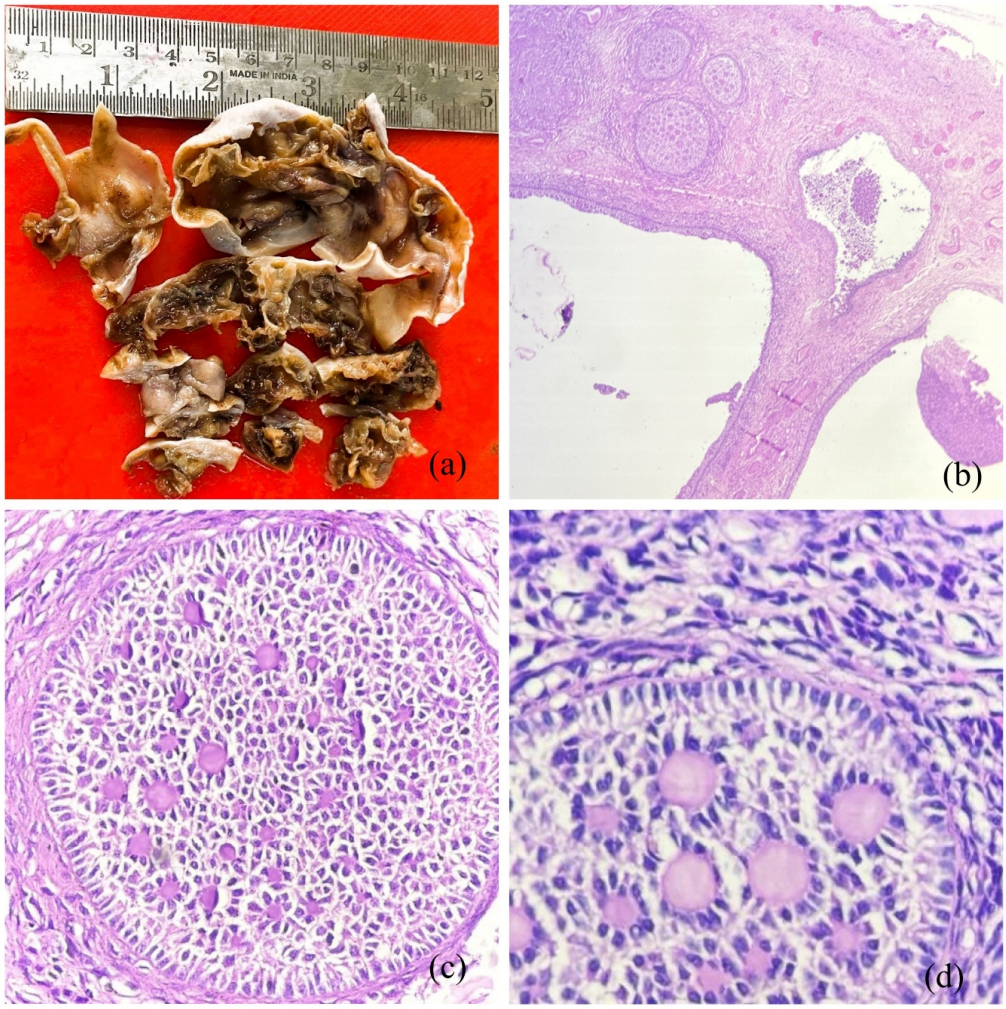

Figure 1: Gross specimen: multicystic ovarian tissue. (b) Microscopy: cysts lined by stromal cells along with solid areas showing tumor nests (h&e stain; 40x); (c) tubules encircling hyaline basement membrane material (h&e stain; 200x); (d) Cells showing antipodal nuclear distribution (h&e stain; 400x).

Table 1: Differential diagnosis for SCTATs.

Tumor	Microscopy	IHC
Sertoli cell tumor [5]	Sertoli cells arranged in tubules having cuboidal to columnar cells, round to oval nuclei and pale cytoplasm. No complex architecture or hyaline basement membrane like material seen.	Positive: Inhibin, SF1, Calretinin Negative: CK7, PAX8, GATA3, Chromogranin, EMA
Granulosa cell tumor [6]	Tumor cells arranged in diffuse, trabecular, insular, micro and macrofollicular pattern. Cells are cuboidal to polygonal having scant pale cytoplasm, uniform angulated nuclei having grooving (coffee bean appearance). Call-exner bodies (small follicle like structure filled with eosinophilic material) are a characteristic feature.	Positive: Inhibin-A, Calretinin, FOXL2, SF1, Vimentin, WT1, CD56, SMA Negative: CK7, EMA
Gonadoblastoma [7]	Immature sertoli cells and germ cells are seen in clusters surrounding basement membrane like material.	Positive: Oct3/4, PLAP, CD117, Inhibin, CK, FOXL2, SF1 Negative: EMA

1 Shah S N (2007) [9] Bilateral sex cord tumor with annular tubules of ovary without Peutz-jeghers syndrome a case report 2 Bembde A S et al (2013) [10] Ovarian sex cord stromal tumor with annular tubules- a case report and review of literature 3 Momin Y A et al (2013) [11] Non Peutz-Jegher syndrome associated malignant sex cord stromal tumor with annular tubules 4 Singh M et al (2014) [12] Sex cord tumor with annular tubules: an incidental finding in an endometriotic cyst-the first known occurrence 5 Patel K et al (2019) [13] Sporadic ovarian sex cord stromal tumor with annular tubules: a

rare case report 6 Gaikwad L et al (2020) [14] Sex cord stromal tumor with annular tubules of the ovary- a case report 7 Choudhary F et al (2020) [8] Non syndromic sex cord tumor with annular tubules: a rare diagnosis 8 Kumar RN et al (2020) [15] Cytomorphology of sex cord stromal tumor with annular tubules with histopathological correlation: a rare case report 9 Maity P et al (2020) [16] Sex cord tumor with annular tubules with unusual morphology in an infant with Peutz-jeghers syndrome 10 Rani P et al (2020) [17] Occult expression of non-syndromic sex cord tumor with annular tubules in ovary with coexistent cervical fibroid 11 Jawalkar S et al (2021) [18] Sex cord stromal tumor of ovary- an unusual case report 12 Sharma D et al (2022) [19] Sex cord stromal tumor with annular tubules: a rare entity 13 Khandeparkar S G S et al (2023) [20] A rare case of unilateral ovarian sex cord tumor with annular tubules 14 Malapaka S et al (2025) [3] Sex cord stromal tumors with annular tubules: exploring a rare ovarian tumor in a teenage patient

The case which we are describing is of a young adolescent female with no features of Peutz-jeghers syndrome presenting with unilateral lesion showing the characteristic antipodal nuclear distribution in tubules and hyaline basement like membrane material. This case presents as a rare presentation of an already rare neoplasm as non-syndromic unilateral SCTAT in an adolescent female.

Mainstay of treatment is surgical resection of tumor. Recently, taxane plus bevacizumab has also been tried in cases where complete resection cannot be done. [8]

Conclusion

SCTAT is a rare neoplasm with characteristic microscopic features. It should be considered in differential diagnosis of ovarian lesions even in absence of Peutz-jeghers Syndrome. It is usually benign in nature but non syndromic cases should be kept in mind for its aggressive nature and should be followed up regularly.

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