# Case Report

## Recurrence of Non-Syndromic Sex Cord Stromal Tumor with Annular Tubules of Ovary: Case Report

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## ABSTRACT:

**One of the unusual variant of ovarian tumor is sex cord stromal tumor with annular tubules (SCTAT). The recurrence in case of malignant ovarian SCTAT ranges from 3mo to 20yr. This report describes the case of recurrence of SCTAT in a 35yr old woman after 4yr of hysterectomy with bilateral salphingo-Oopherectomy. Microscopic examination revealed features of SCTAT. Because of its unusual behavior evidenced by delayed recurrence in spite of bland cellular features, proper long term follow–up is essential.**

***Keywords:*** **Sex Cord Stromal Tumor(SCTAT), Recurrence, India**

## Introduction

Sex cord stromal tumor with annular tubules (SCTAT) is rare and unusual variant of ovarian neoplasm. It was described by Scully in 1970 (1). The classification of SCTAT is controversial. It shares the features of sertoli cell tumor and granular cell tumor. Because of its special features it was considered as a separate entity (1). World Health Organization (WHO) included SCTAT in the category of mixed/ unclassified cell type SCTAT(2). One third of the cases are associated with Peutz-Jeghers syndrome (PJS) and the rest are sporadic. Both sporadic and associated with PJS have been reported in patients ranging in age from childhood to old age. The average age is 36 yr. Symptoms of hyperesternism are seen in 50% of cases. Tumors associated with PJS are bilateral, multifocal, small and calcified.Sporadic (non-syndromic) SCTAT are unilateral, large, and malignant in 22% of the cases(3). Few cases are reported in the literature. Young *et al.* reviewed 74 cases of SCTAT, 47 cases were non-syndromicSCTAT,of which 4 cases presented with recurrence (3). Another study reported one case with late recurrence of non- syndromicSCTAT(4).

This report describes a rare case of recurrence of ovarian non-syndromic SCTAT in a 35yr old woman after 4yr of total hysterectomy with bilateral salphingo-Oopherectomy emphasizing on pathological features and behavior of SCTAT during the diagnosis.

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## Case Report

A 35yr old woman presented to Surgical Outpatient Department, with distension of abdomen since 20 days ago. The distension of abdomen was insidious in onset, gradually progressed to the present size. There was no history of loose motions, vomiting, pain per abdomen and fever. She underwent hysterectomy with bilateral salphingo-oopherectomy for fibroid uterus and malignancy of ovary4yrs back. Histopathological examination revealed SCTATwith annular tubules of ovary with leiomyoma of uterus. Subsequently she did not receive chemotherapy or radiotherapy. Family history was insignificant. She was non-diabetic and her blood pressure was within normal limits. Furthermore, there was no history of TB, asthma, and epilepsy. On physical examination the respiratory, cardiovascularand-central nervous systems were normal. Abdominal examination revealed soft distended abdomen with no organomegaly. The vital signs and laboratory investigations are shown in Table.1 which were within normal limits.

Pelvic ultrasonagraphy revealed a very large echoic mass in the mid-abdomen close to the third part of duodenum compressing right ureter and inferior venacava. Radiologist diagnosed it as gastrointestinal stromal tumor (GIST).

The patient was subjected for laparotomy under epidural anaesthesia. Abdomen was opened in layers. Mesenteric tumor of about 20×10cm in size was noted.Intra operative diagnosis was a metastatic tumor. About 3X3cm of biopsy was taken and sent for histopathological examination. Per-operatively liver and spleen were normal. Abdomen was closed and there were no intra-operative complications. Antibiotics were given prophylactically for surgery.

The specimen was processed routinely in histopathological section and the sections were stained with haematoxylin and eosin (H&E). On microscopic examination a dimorphic population of tumor cells was seen.

**Table1-**Vital signs and laboratory findings

**Vital Signs**:
Pulse Rate: 86/minute
Blood Pressure: 130/80 mm of Hg
Respiratory Rate: 28/minute
Temperature: 38.5$^{o}$c

**Laboratory Investigations:**
Hb: 11g/dl (normal:12.5-14.5g/dl)
RBC Count: 3.8millions/cmm (normal:4.5-5.5millions/cmm)
Haematocrit: 33% (normal: 35% to 45%)
WBC Count: 8000 cells/cmm (normal: 4000 to 11,000/cmm)
Differential Count: Neutrophils: 68%
Lymphocytes: 27%
Eosinophils: 02%
Monocytes: 02%
Basophils: 01%
Random Blood Sugar: 78mg/dl (normal: 80 to 110mg/dl)
Blood Urea: 19mg/dl (normal: 5 to 25mg/dl)
Serum Creatinine: 0.8mg/dl (normal: 0.8 to 1.2mg/dl)

**Serological Investigations:**
HIV-1 & 2:Negative
HBsAg : Negative

**Radiological Findings:**
Chest X-ray : Normal
Abdomen X-ray: Normal

**Tumor Markers:**
CA-125 : 4units/ml (normal: 0 to 35 units/ml)

The cells composed of (a) darkly stained oval to columnar type arranged in palisading pattern around eosinophilic hyaline like material giving an annular tubular picture. These tubules were surrounded by large vacuolated cells with centrally placed hyperchromatic cell differentiation and at places resemble the sertoli cells. (b) The larger vacuolated cells had differentiation towards leydig or theca cells. The surrounding fibrocollagenous tissue showed infiltration of these annular tubular structures with eosinophilic material associated with lymphocytic infiltration at places, the stroma showed papillary invaginations into cavitary spaces and were lined by ser-

toli and granulosa cells. Mitotic activity in this tumor was rare as shown in Fig.1&2. This was diagnosed as sex cord stromal tumor with annular tubules.

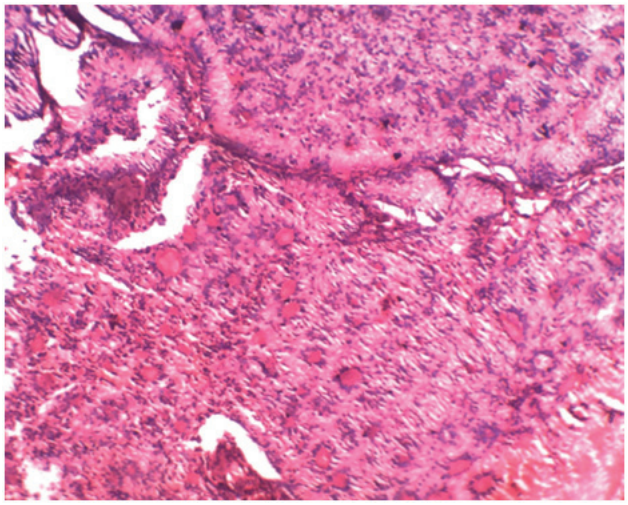

**Fig. 1:** Sex cord stromal tumor with annular tubules of ovary (H&E×10)

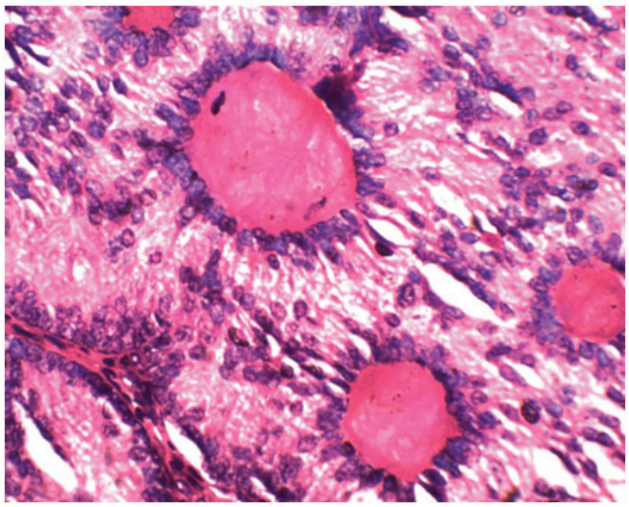

**Fig. 2:** Sex cord stromal tumor with annular tubules of ovary (H&E ×40)

## Discussion

SCTAT arerare ovarian tumor that approximates 7% of all ovarian malignancies(5). SCTAT is an unusual form of ovarian tumor where the histogenesis and differentiating potentiality is not known (7). Majority of the ovarian SCTAT are benign neoplasms that arise sporadically. Malignant behaviour in SCTAT has been reported only in sporadic cases(6). Lele SM reported malignant ovarian SCTAT in a patient with PJS(7).

This survey describes an exceptional tumor, namely recurrence of ovarian SCTAT after 4yr of hystrecetomy with bilateral salphingo-oopherectomy.Malignant SCTAT seems to spread mainly via the lymphatics with typical sites of tumor metastasis being the pelvic, para-arotic and supraclavicular lymph nodes. Other sites of tumor recurrence and metastasis include the retroperitoneum, parietal and visceral peritoneum, liver, kidney and lung. Unique to our case was the presence of tumor in the visceral peritoneum.Recurrence is reported after 9yr of primary surgery(4). In literature, the reported time of first recurrence in cases of malignant SCTAT ranges from 3mo to 20 yr(2,3,8). Although SCTAT is capable of producing hyperesternism symptoms,in this case none of the symptoms of hyperesternism were present. There was no elevation of CA-125 or CEA as in the present case, but inhibin and mullerian inhibiting substance proved to be effective markers in this rare malignancy(9). Thirteen previously reported cases of malignant SCTAT (sporadic) were relatively large with a diameter ranging from 11 to 22cm, and all were unilateral(5). In this regard the findings in our case were similar.

Sertoliform tubules, endometroid and foci of granulose cell differentiation are some of the histiologic pattern seen in both benign and malignant SCTAT including sporadic and associated with PJS as per the literature (3). In the present case sertoliform tubules and granulose cell differentiation were seen. The malignant potential of SCTAT cannot be reliably predicted by microscopic examination of the primary tumor. Mitotic activity in the 13 reported malignant cases ranged from rare to 7-10 mitosis/10HPF(10). In agreement with that, the reported case imparted bland cellular features and mitosis was rare. The differential diagnosis in the present case was gonadoblastoma. But gonadoblastoma is differentiated by the presence

tumor nests surrounded by fibrous stroma. The nests contain basement membrane like material, large germ cells and small dark sex- cord stromal cells.

Considering its unusual behavior evidenced by delayed recurrence,in spite of bland cellular features, proper long term follow–up is essential.

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