



Sex Cord-Stromal Tumor with Annular Tubules of the Ovary

CASE REPORT

Hale Demir¹, Tülin Kılıçaslan Öztürk¹, Zerrin Calay¹, Şennur İlvan¹, Fuat Demirkıran²

ABSTRACT

Sex cord-stromal tumors with annular tubules (SCTAT) of the ovary are rare. They have two clinical presentation forms: the syndromic form, which is associated with Peutz-Jeghers syndrome, and the non-syndromic form, which is frequently seen in the second or third decades. We describe a 13-year-old patient who underwent left oophorectomy. Macroscopically, the mass was 16×13×8 cm in diameter, encapsulated, and lobulated. It showed a yellow-to-orange solid cut surface with small cystic areas and hemorrhagic cleft-like spaces. Microscopically, the tumor had nodular areas that were separated with fibrous stroma. They were composed of tubules, which were lined by columnar shaped cells with eosinophilic cytoplasm, nuclear palisading, and central Periodic Acid Schiff (PAS)-positive eosinophilic hyaline bodies. Immunohistochemically, the tumor cells were strongly positive for inhibin. Finally, the tumor was diagnosed as non-syndromic ovarian SCTAT.

Keywords: Sex cord stromal tumor, SCTAT, ovarian tumor, ovary

INTRODUCTION

Sex cord-stromal tumors of the ovary account for only 5% of all ovarian neoplasms, and sex cord-stromal tumors with annular tubules (SCTAT) are very rare tumors in this group. Many of these tumors are clinically indolent, so long period follow-up is required to precisely clarify outcomes data.

Sex cord-stromal tumors with annular tubules has two clinical presentation forms: one-third of the tumors are associated with Peutz-Jeghers syndrome, and the other form occurs as solitary neoplasm in the general population (1, 2).

In patients with Peutz-Jeghers syndrome, the tumors are usually bilateral, multifocal, and almost always very small tumorlets found incidentally in the ovaries; all of them are clinically benign (1, 2). There is some calcification in the tumoral tissues. The syndromic form can be seen in all age groups, and the mean age is 27 years. In addition, nearly 15% of patients with Peutz-Jeghers syndrome may have cervical adenocarcinoma (3-5).

The second one is non-syndromic form, which is not associated with Peutz-Jeghers syndrome as in this case report. They are usually unilateral, unifocal, and larger than 3 cm, and sometimes behave in a malignant manner (up to 1/5 of patients), with lethal metastases or recurrences. The mean age is 34 years (1, 2).

Serum inhibin, Mullerian inhibitory substance, and progesterone can be used as tumor markers for SCTATs. In addition, nearly half of tumors from the latter group have manifested clinical signs of hyperestrogenism or hyperprogesteronism (1, 2). Therefore, SCTAT can present with precocious puberty, menstrual disorder, or post-menopausal vaginal bleeding, depending on presentation age. The histopathological features of SCTAT cases are identical for both syndromic and non-syndromic forms.

Most authors have suggested that these tumors are related to granulosa cell tumors and, partly, Sertoli cell tumors, but actually these tumors have histological features of both cell types. In brief, SCTATs are accepted as an intermediate category (1, 2).

CASE REPORT

The patient was a 13-year-old girl, and she complained of abdominal pain and swelling and long duration of menstrual hemorrhage. Abdominal ultrasonography described left adnexal solid mass that was 137 mm in diameter. Then, left oophorectomy and regional lymph node dissection were performed in Department of Obstetrics and Gynecology, İstanbul University Cerrahpaşa School of Medicine. The pathologic examination was performed in Department of Pathology, İstanbul University Cerrahpaşa School of Medicine.

This case was presented at the 17th National Congress of Pathology, 1-6 October, 2004, Gaziantep, Turkey.

¹Department of Pathology, İstanbul University Cerrahpaşa School of Medicine, İstanbul, Turkey

²Department of Obstetrics and Gynecology, İstanbul University Cerrahpaşa School of Medicine, İstanbul, Turkey

Received
14.03.2016

Accepted
15.03.2016

Correspondence

Dr. Hale Demir, İstanbul Üniversitesi Cerrahpaşa Tıp Fakültesi, Patoloji Anabilim Dalı, İstanbul, Turkey
Phone: 414 30 00/22505
e.mail: patdhd1@hotmail.com

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Figure 1. Macroscopic appearance of the tumor. Ovarian tumor is predominantly solid and cleft-like spaces are visible

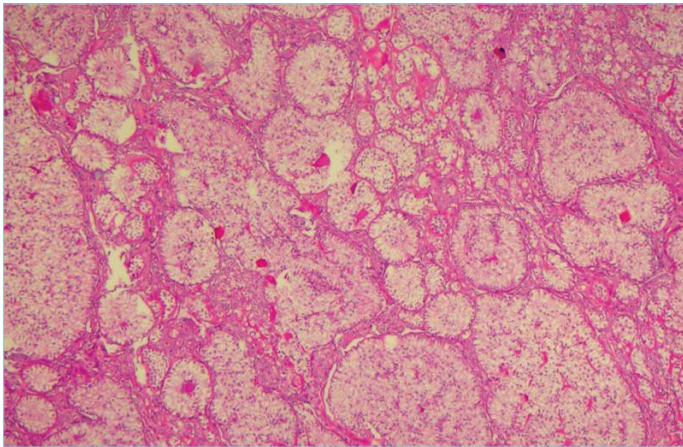


Figure 2. Sex cord-stromal tumor with annular tubules shows complex annular tubules containing hyaline bodies set in fibrous stroma (HE $\times 100$)

Macroscopically, the removed ovarian mass was $16 \times 13 \times 8$ cm, encapsulated by a thick fibrous capsule, and lobulated. On sectioning, the specimen showed a yellow-to-orange solid cut surface with small cystic areas and hemorrhagic cleft-like spaces (Figure 1).

Microscopically, the ovarian mass had nodular areas that were separated with fibrous stroma. The tumor was composed of tubules containing central eosinophilic hyaline bodies. The cells consisting of the tubules were columnar and had abundant eosinophilic cytoplasm and nuclear palisading (Figure 2). The nuclei of these cells were round or oval and had a single small nucleolus. The central hyaline bodies stained strongly positive with PAS. Immunohistochemical staining for inhibin was strongly positive in tumor cells' cytoplasm (Figure 3).

In the light of all these findings, the tumor was diagnosed as SCTAT of the ovary. Informed consent has been taken from patient for publication.

DISCUSSION

Sex cord-stromal tumors with annular tubules are rare ovarian neoplasms that can occur in two different clinical settings. One of them

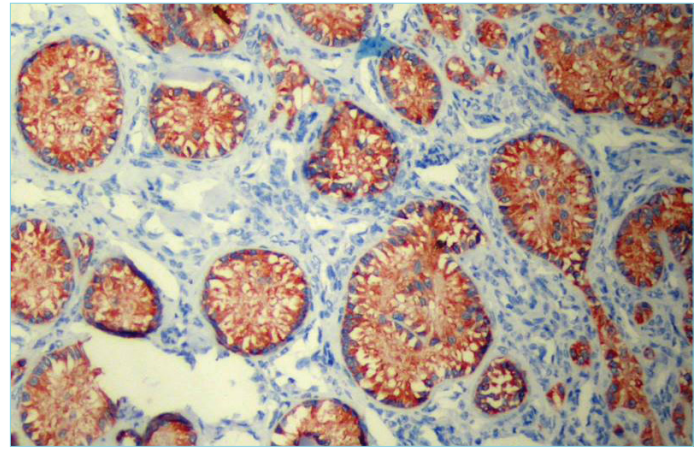


Figure 3. Small typical cell nest displaying strong cytoplasmic immunostaining for inhibin ($\times 100$)

is described in patients with Peutz-Jeghers syndrome. The other is named as non-syndromic or sporadic form (1, 2).

Our case was of a non-syndromic SCTAT. Non-syndromic SCTATs were not seen frequently in the teenage group. Most of the tumors were diagnosed when the patients were in their second or third decades. The mean age is 34 years. The presented case was 13 years old.

The non-syndromic SCTATs are usually unilateral, unifocal, and larger than 3 cm, and sometimes behave in a malignant manner (in up to 1/5 of patients) with lethal metastases or recurrences. The tumor size in our patient was also considerably big. The tumor sizes of previously reported cases have ranged from 5 to 30 cm in diameter (6-9).

Most authors have suggested that the SCTATs are related to granulosa cell tumors and, partly, Sertoli cell tumors. SCTATs have histological features of both cell types (1, 2). Therefore, SCTATs must be distinguished from microfollicular granulosa cell tumors, well-differentiated Sertoli cell tumors, as well as gonadoblastomas.

CONCLUSION

Sex cord-stromal tumors with annular tubule is a rarely determined entity of the ovary. In order to add a case to medical literature, we wanted to present our case who was an adolescent unlike that previously presented in the literature.

Informed Consent: Informed consent has been taken from patient for publication.

Peer-review: Externally peer-reviewed.

Authors' Contributions: Conceived and designed the experiments or case: HD, TKO. Performed the experiments or case: SI, ZC, FD. Analyzed the data: HD, TKO, SI, ZC. Wrote the paper: HD, TKO. All authors have read and approved the final manuscript.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

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