International Journal of Research in Medical Sciences
Patel K et al. Int J Res Med Sci. 2019 Sep;7(9):3591-3594
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: http://dx.doi.org/10.18203/2320-6012.ijrms20193955

Case Report

Sporadic ovarian sex cord-stromal tumor with annular tubules: a rare case report

Krutika Patel*, Devang Painter, Vashudha M. Bhagat, Pinkal Shah

Department of Pathology, Government Medical College, Surat, Gujarat, India

Received: 30 June 2019
Revised: 07 August 2019
Accepted: 14 August 2019

***Correspondence:**
Dr. Krutika Patel,
E-mail: krux.hnh@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 40 year old lady with no association of Peutz-Jegher (P-J) syndrome. The patient's chief complaints were post-menopausal bleeding for 1 year on and off along with menorrhagia. MRI abdomen was suggestive of intensely enhancing solid tissue mass lesion in the right adnexa, features suggestive of ovarian mass. Panhysterectomy was done. Grossly uterus and left adnexa appeared to be normal. Right ovary showed mass measuring 17x11x9cm^3 in size, on cut section, solid, homogenous lobulated, yellowish areas identified. Microscopic and Immunohistochemistry findings confirmed the diagnosis of sex cord stromal tumor with annular tubules of granulosa cell type. PAS stain supported the diagnosis.

Keywords: Annular tubules, Inhibin, Sex cord stromal tumors, Sex cord tumors with annular tubules

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.[1,2] Sex cord tumors with annular tubules (SCTAT) are first described by Scully.[1] 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.[2,3]

In patients with Peutz-Jeghers syndrome, the tumors are usually bilateral, multifocal, and almost always very small <3cm tumor lets found incidentally in the ovaries.[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.

The second one is non-syndromic form, which is not associated with Peutz-Jeghers syndrome as presented in this case report. They are usually unilateral, unifocal, and larger than 3 cm, and sometimes behave in a malignant manner (upto 1/5 of patients), with lethal metastases or recurrences.[1,2] 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 have manifested clinical signs of hyperestrogenic or hyperprogesteronism.[1-4] Therefore, SCTAT can present with precocious puberty, menstrual disorder, or postmenopausal vaginal bleeding, depending on presentation age. The histopathological features of

SCTAT cases are identical for both syndromic and non-syndromic forms.

CASE REPORT

A 40 year old lady presented with complain of irregular menstrual bleeding and pelvis mass for 1 year. USG and CT (abdomen and pelvis) was suggestive of enhancing solid mass lesion arising in the pelvis cavity extending from L3 to S1vertebral level, features were suggestive of malignant neoplastic etiology from ovary. Pan hysterectomy was done, and specimen sent for Histopathological examination.

Gross findings

Grossly uterus and left adnexa appeared to be normal, right ovarian mass measuring 17x11x9 cm^2. On cut section reveled solid, homogenous, yellowish white, lobulated areas. No cystic areas identified.

Microscopic findings

The sections from ovarian mass showed rounded/nodular aggregates, nests of simple and complex closed tubules with areas of eosinophilic hyaline material (PAS positive) with focally interspersed fibrous stroma. Tumour cells were columnar with round/oval hyperchromatic nuclei with small nucleoli with clear/formy cytoplasm. Occasional nuclei were showing nuclear grooving.

Focal areas of infarction were also seen. Mitosis, calcification or germ cell component were not seen. Immunohistochemical study showed the tumor cells were strongly positive for inhibin, calretinin and negative for CK and EMA and, the tumor was diagnosed as Sporadic Sex Cord Tumor with Annular Tubules of ovary.

Figure 1 shows Gross ovarian mass; cut section showing solid, homogenous, firm, yellow white, and lobulated areas.

Figure 1: Gross appearance; cut section; solid and lobular.

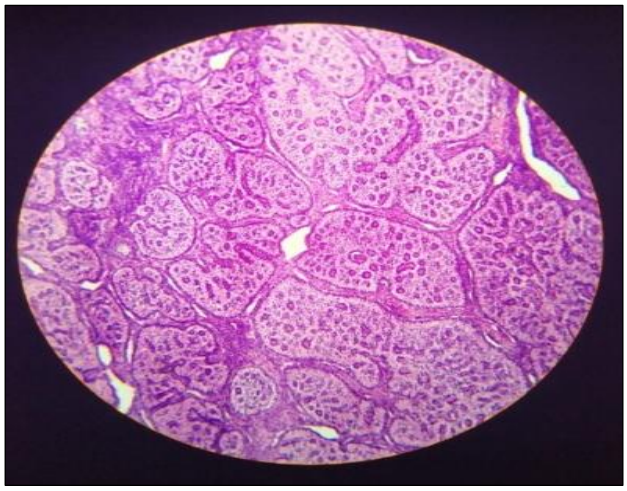

Figure 2: 10x view; tumor cells arranged in nodular aggregates, having simple and complex tubules with interspersed fibrous stroma.

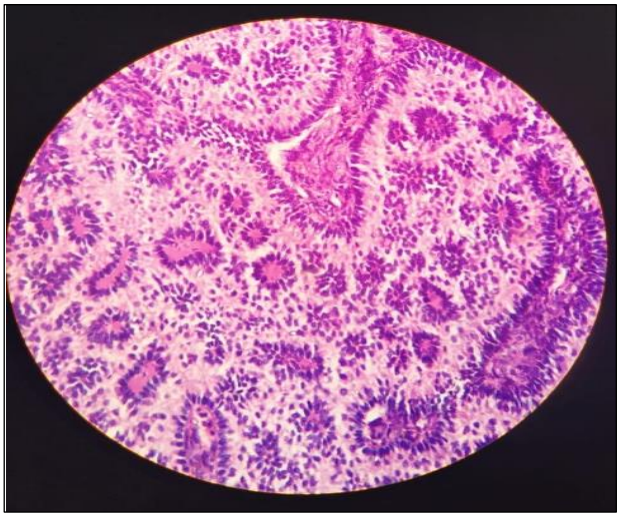

Figure 3: 40 x view: tubules eosinophilic hyaline material.

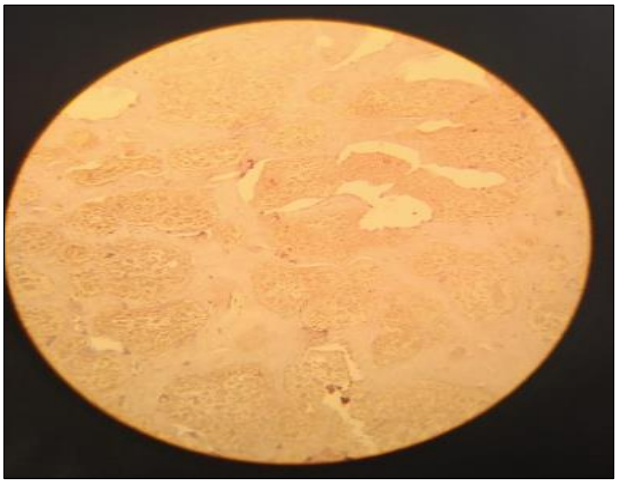

Figure 4: Tumor cells are immunopositive for inhibin.

Figure 2 shows 10x magnification view by using Hematoxylin and eosin stain showing tumor is composed of nodular aggregates of complex and simple tubules.

With the help of 40x magnification view by using hematoxylin and eosin stain tumor which showing antipodal arrangement of nuclei within tubules with eosinophilic hyaline material (Figure 3). 10x magnification view which shows inhibin immunopositivity of tumor cell (Figure 4).

DISCUSSION

In the past, SCTAT have been placed in the category of unclassified sex cord-stromal neoplasms. In the recently published classification of tumors of female reproductive organs by WHO (4th edition), SCTAT was classified as pure sex cord neoplasms. The most frequent clinical manifestations of SCTAT relate to estrogen production, including iso sexual precocity, menstrual irregularity and post-menopausal bleeding.[2,3] SCTAT is divided into two different variants by whether it is associated with Peutz-Jeghers syndrome (PJS) or not.[4-6]

Roughly one-third of SCTAT is associated with PJS. PJS is an autosomal dominant hereditary disorder with variable penetrance characterized by mucocutaneous melanin pigmentation, hamartomatous polyps of the gastrointestinal tract, and an increased risk for cancer of gastrointestinal and non-gastrointestinal sites.[7] Both sporadic and PJS-associated SCTAT have been reported in patients ranging in age from 4 to 76 years old. In these cases, it was 40 year.

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.[3] This case showed similar histomorphologic features, however calcification was not seen. The tumour cells are usually positive for inhibin, calretinin, FOXL 2, SF-1, WT1 and CD56.[3,4]

In this Case tumor cells were immunoreactive for inhibin, calretinin and negative for EMA, other IHC markers were not available. By morphological features it should be diffentiated from Granulosa and Sertoli cell tumours. Presence of germ cell component is distinguished these tumors from Gonadoblastoma.[3]

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 retro peritoneum, peritoneum, liver, kidney, and lung.[8-9] Unilateral salpingo-oophorectomy 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.[6] The histologic picture of SCTAT is distinctive and is generally adequate to exclude other tumors in the differential diagnosis such as granulosa cell tumor and gonadoblastoma.[7]

CONCLUSION

Ovarian sex cord stromal tumor with annular tubules (SCTAT) is a distinctive, rare subtype of sex cord stromal tumor of the ovary. In these cases, it was large, >3 cm, unilateral and not associated with P-J syndrome, so diagnosed as sporadic ovarian SCTAT of granulos cell type and follow up was advisable as the tumour size was large.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Patel K, Painter P, Bhagat VM, Shah P. Sporadic ovarian sex cord-stromal tumor with annular tubules: a rare case report. Int J Res Med Sci 2019;7:3591-4.