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Pathology

SEX CORD-STROMAL TUMOR WITH ANNULAR TUBULES : A RARE ENTITY

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ABSTRACT Sex Cord-Stromal Tumor with Annular Tubules (SCTAT) is an uncommon ovarian neoplasm accounting for only 6% of all sex cord-stromal tumors, mostly seen in the reproductive age group. Here we describe a case of 48 years old female who presented with amenorrhea and galactorrhea of 4 months duration. On histopathological examination, it was diagnosed as SCTAT and confirmed on IHC.

KEYWORDS : SCTAT, ovarian tumor

INTRODUCTION

SCTAT was first described by Scully in 1970[1]. It is a rare entity accounting for only 6% of all sex cord-stromal tumors of the ovary. Sex cord stromal tumors comprise for 8% of all the ovarian neoplasms[2]. It is a distinctive neoplasm with morphological features between Sertoli cell tumor and granulosa cell tumor. It is distinguished by its ring-shaped tubules[3].

Clinically, SCTATs mostly present in reproductive age group with menstrual irregularities and sterility but rarely can present in pediatric age group as precocious puberty. These clinical features are detected earlier than the ovarian neoplasm[4].

Its clinical diagnosis is very difficult. Therefore, diagnosis of SCTAT is mostly made on histopathological examination.

Case Report

A 48 years old female presented with amenorrhea and galactorrhea for 4 months in the gynaecology department. She had one live birth. As per abdominal examination, there was a mass arising from pelvis of around 14 weeks size. MRI brain showed normal pituitary gland. On USG abdomen, she had a right adnexal mass. CA-125 levels (11.2 U/ml) were not raised but she had increased serum prolactin levels (123.22 ng/ml).

Right salpingo-oophorectomy was done and specimen sent to Pathology department. We received a right adnexal mass measuring about 10 cm in diameter alongwith attached fallopian tube for histopathological examination.

Grossly, it had smooth outer surface and homogenous grey brown to yellow cut section.(Fig.1)

Microscopic examination revealed predominant solid sheets and nests of tumor cells separated by thin fibrovascular septa having minimally pleomorphic round vesicular nuclei, fine chromatin, focally present small nucleoli and abundant eosinophilic granular to clear cytoplasm with focally well defined cell boundaries. Microcystic areas filled with eosinophilic secretions and tubular pattern were also present. Distinct annular tubules having nuclei oriented peripherally around a central hyalinized body composed of PAS positive material and intervening anuclear cytoplasmic zone were characteristic. In view of these findings, diagnosis of SCTAT was made and further confirmed by inhibin A positivity on IHC.

Discussion

SCTATs are classified as subgroup of sex cord stromal tumors as they have common features with sertoli and granulosa cell tumors. They arise from sex cord cells of the ovary. The clinical manifestations of SCTAT such as menorrhagia, postmenopausal bleeding, precocious puberty and sterility are mainly due to estrogen-progesterone secretion.

One third of sex cord-stromal tumors with annular tubules are associated with Peutz-Jeghers syndrome and other occurs sporadically[5]. Patients with PJS usually presents with bilateral, multifocal and very small tumors (<3cm) found incidentally in the ovaries. This syndromic form can be seen in all age groups with mean age of 27 years and are benign. Contrary to that, sporadic forms are usually unilateral, unifocal and larger than 3 cm with mean age of presentation being 36 years. These tumors have low malignant potential and can metastasize sometimes. Rarely, these tumors are associated with adenoma malignum of cervix, Turners syndrome, dysgerminoma, gonadoblasoma, endometrial carcinoma endometriosis of fallopian tube and endometriotic cyst[6].

Macroscopically, SCTATs can be solid, cystic, mixture of solid and cystic, yellow to tan, and some with haemorrhage and necrosis. Size usually ranges up to 30 cm[6]. Microscopically, SCTATs are typically characterized by circumscribed columnar epithelial nests composed of ring-shaped tubules, which are encircled by acidophilic hyalinized basement membrane-like material and some with calcification[7].(Fig. 2,3 &4).

These tumors should be distinguished from Sertoli cell tumor, gonadoblastoma and microfollicular granulosa cell tumor. In sertoli cell tumors, complex tubules are not seen. In microfollicular granulosa cell tumor, hyaline bodies in tubules are not dominant. In gonadoblastoma, there is presence of germ cell component which is absent in SCTAT[6,8,9].

The exact behavior and long term prognosis in these patients are almost unknown. The present management guidelines suggest that surgery alone with preservation of fertility (if possible) should be attempted in these patients.

CONCLUSION

SCTAT is a rare and distinctive neoplasm whose clinical diagnosis is very difficult. Histopathological examination of this ovarian neoplasm is a must for the diagnosis.

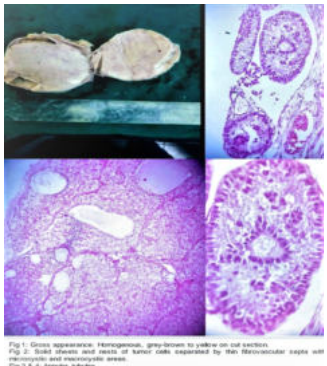

Fig 1: Gross appearance: Homogenous, grey-brown to yellow on cut section.
Fig 2: Solid sheets and nests of tumor cells separated by thin fibrovascular septa with microcystic and macrocystic areas.
Fig 3 & 4: Annular tubules.

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