

Case Report

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Histopathology of Sertoli Cell Tumor of the Ovary: A Rare Case Report

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Abstract

Ovarian Sertoli cell tumors are rare and have a wide variety of morphological patterns that can be mimicked by other ovarian neoplasms. They are usually benign, unilateral, and most commonly occurring in the reproductive age group. Due to the low incidence of OSCTs, information on the clinical behavior, morphological spectrum, optimal management, and prognosis is limited. Here we report a very rare case report of 45 year old female presented with a pelvic mass and clinically diagnosed with an ovarian mass on the right side.

Keywords: Sertoli Cell Tumor, Ovary, Sex Cord-Stromal Tumor

1. Introduction

Ovarian Sertoli cell tumors (OSCT) are a subgroup of ovarian pure sex-cord stromal tumors which are very rare entities comprising of Sertoli cells [1]. The characteristics of these tumors are they lack Leydig cell component in contrast to Ovarian Sertoli-Leydig cell tumors (SLCT) which are a subgroup of mixed-sex cord stromal tumors of the ovary [1,2]. They are typically benign occurring most commonly in the reproductive age group and having a variety of histo-morphological patterns. They are detected as a pelvic mass or just symptoms of hormonal manifestation most commonly due to estrogen. Some of these tumors are associated with Peutz-Jeghers syndrome or DICER1 mutation [1]. Due to the low incidence of OSCTs, information on the clinical behavior, morphological spectrum, optimal management, and prognosis is limited [1,2]. Here we report a very rare case report of OSCT with

a variety of histomorphological features.

2. Case Report

A 45 year old female presented with a pelvic mass and was clinically diagnosed as? broad ligament fibroid/?ovarian mass of right side. USG finding shows a mass of 10x9 cm attached to the wall of the uterus. A transabdominal hysterectomy with a bilateral salpingo-oophorectomy specimen sample was submitted to the Department of Pathology.

Grossly the mass was enlarged and measures 11x9x8cm. The outer surface was grey-white with venous prominences. On serially sectioning, the cut surface was entirely solid, tan to yellowish. Necrosis and hemorrhage were not seen. (Figure 1). The attached right fallopian tube was grossly unremarkable.

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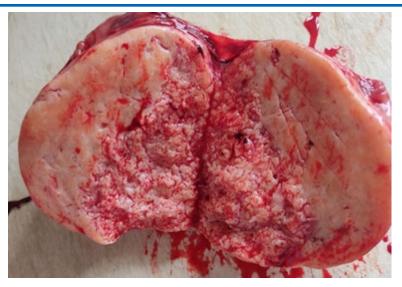


Figure 1: Gross morphology of the mass: entirely solid, tan to yellowish on cut surface.

On histopathological examination, the mass reveals tumor cells which were arranged predominantly in tubules, showing round to oval open glands with central lumen. Also, these tumor cells were arranged in a diffuse solid trabecular and cords which were 2 to 3 cells thick. Individual cells were columnar to cuboidal having small to medium-sized round to oval nuclei with clear to pale eosinophilic cytoplasm. Fibrous stroma ranges from moderate to abundant. However, mitosis, necrosis, nuclear atypia, and Leydig cells were not seen in the entire sections examined. Foci of normal ovarian parenchyma were also seen.

Section examined from the cervix, endometrium, bilateral fallopian tubes, and left ovary was histologically unremarkable.

Hence morphologically, the final histopathological was given as a Sertoli Cell Tumor of the right ovary.

The patient was advised for immunohistochemistry (IHC) to confirm the diagnosis.

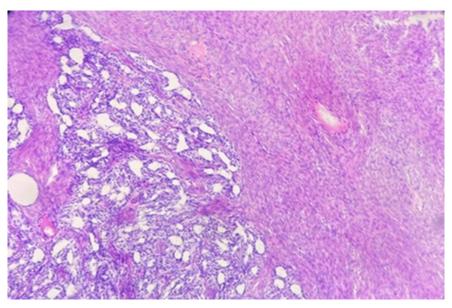


Figure 2: Tumor cell and fibrous stroma (10x H & E)

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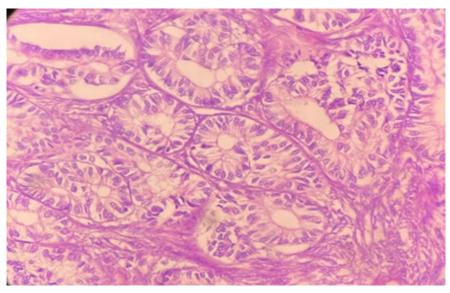


Figure 3: Tumor cells arranged in tubules (40x H & E)

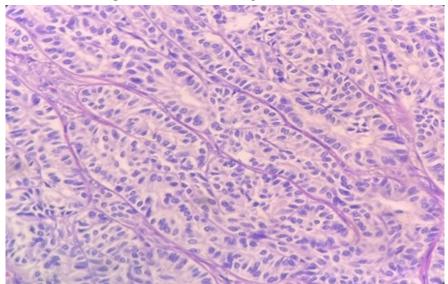


Figure 3: Tumor cells arranged in diffuse cords and trabecular pattern

3. Discussion

Ovarian Sertoli cell tumor (OSCT) is a distinctive pure sex cord neoplasm of Sertoli cells unlike Sertoli-leydig cell tumors which are mixed sex cord stromal tumors of the ovary according to WHO classification of female genital tumor, 5th edition [1]. Previously this tumor has been regarded as a variant of granulosa cell tumor as well as in the category of Sertoli-leydig cell tumor (SLCT) [3]. The majority of the patients reported in the various series with OSCT have been in the reproductive age group though it has a wide range of age groups. Clinically they present as pelvic mass as the incidental findings or sometimes just the symptoms of hormonal manifestation. The hormonal manifestations are mainly due to estrogen and rarely due to the production of androgens, renin, progesterone, or aldosterone. Strikingly some of these tumors are associated with Peutz-Jeghers syndrome and pathogenesis related to DICER1 mutation [1,3,4].

Grossly SCTs are usually unilateral with a mean size of 8cm with a cut surface typically solid with a tan to yellow lobulated appearance as in our case. However, they may be entirely cystic or solid-cystic with sometimes the presence of hemorrhage and necrosis. Though in our case hemorrhage and necrosis were not seen [1,3].

Microscopically, OSCTs shows the variety of pattern but the tubular pattern is the most common pattern seen in various literature. Other pattern includes trabecular, diffuse, alveolar, and pseudopapillary. The hollow or solid tubules present in these tumors are the major clue to their nature as they are practically always present at least focally. Tumor cells are cuboidal or columnar and usually have round to oval nuclei with pale cytoplasm, sometimes lipidrich to eosinophilic cytoplasm. Rarely bizarre nuclei or marked cytological atypia and brisk mitotic activity can be seen. Stroma

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ranges from scant to abundant and hyalinized. However, in our case, cytological atypia, bizarre nuclei, mitosis, necrosis, and Leydig cells were not present. The presence of these features are predictive of malignant behavior [1,3-6].

Occasional Leydig cells within the tumor can be present, however, it does not exclude the diagnosis of OSCTs and is most commonly misdiagnosed as SCLTs. The presence of heterologous components strongly supports the diagnosis of SLCT, as no OSCT with heterologous elements and retiform pattern has been reported[1,3]. Another pure sex cord tumor like adult granulosa cell tumors, poses challenges in differential diagnosis. OSCTs exhibit diverse growth patterns such as cords, trabeculae, solid tubules, or islands, which can resemble the growth patterns seen in adult granulosa cell tumors. Additionally, the nuclei of OSCTs may exhibit grooving, a characteristic noted in existing literature. The identification of well-defined solid or hollow tubules becomes crucial for pathologists to distinguish OSCTs from other ovarian tumors. Also one of the most important differential diagnoses that can be included is carcinoid as it comprises the pattern of cords and trabeculae as well as acini which can be confused with Sertoli tubules. However, acini of tubules contain neuroendocrine granules and have nuclei with "salt and pepper" chromatin. Also, immunohistochemistry may help in difficult cases. The positive markers are Inhibin, SF1, Calretinin, CD 99, and cytokeratins [1,3,5-7].

Most OSCTs are usually benign found at stage I, and have a non-aggressive clinical course. Because these tumors are mostly found in the reproductive age group, for whom the preservation of fertility is particularly important, it is necessary to consider the future prognosis and choose the optimal treatment like fertility-sparing surgery [1,2,5].

4. Conclusion

Due to its rarity OSCTs pose difficulty in diagnosis. Also so many ovarian tumors in the same broad category exhibit patterns of hollow and solid tubules making it problematic in evaluating the OSCTs. Because OSCTs are rare, they often evade suspicion. Hence, a comprehensive understanding of their clinicopathological and immunohistochemistry is vital for accurate diagnosis and effective treatment planning, as well as for ensuring proper follow-up care.

Conflict of interest

None to declare.

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Consent to participate

Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

Ethical approval

Not required.

Registration of research studies

Not applicable.

Guarantor

Ratan Shah

Declaration of competing interest

The authors declare no competing interests.

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