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Case Report

Ovarian Sertoli cell tumor: A rare case of sex cord stromal tumor in a three-year female

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Abstract

Sertoli–Leydig cell tumors (SLCTs), also known as androblastoma, are rare, accounting for less than 1% of ovarian neoplasms. The clinical manifestations of SLCT range from asymptomatic to severe virilization. The majority of Sertoli cell tumors are highly differentiated, with homogeneous nuclei and few mitotic signs among the neoplastic cells. This study is a case report of a 3-year-old girl who was admitted to Kashani hospital with abdominal pain and a protruding stomach. On the right side of the abdomen, there was a large, lump palpable. Inhibin, calretinin, actine, and WT1 were all detected as positive by *immunohistochemistry (IHC)* testing. However, CD30, NSE, synaptophysin, vimentin, chromogranin, S100, and AFP were all detected to be negative. Additional information about ovarian sex cord tumors is provided in this case. Sertoli cell tumor should be considered a differential diagnosis in a young girl who has hirsutism, irregular bleeding, and breast atrophy with a pelvic mass.

Keywords: Sertoli–Leydig cell tumors, Ovarian neoplasms, Young girl

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Introduction

Sertoli–Leydig cell tumors (SLCTs), also referred to as androblastoma, are uncommon, accounting for less than 0.1% of ovarian neoplasms. These tumors belong to the sex cord-stromal tumors, which have a testicular pattern of differentiation (1). Sertoli cellular tumors are tumors that have complicated annular tubules. They are most commonly found in the testis, but they can also appear in the ovaries in females (2). They commonly affect women of childbearing age, although they can also influence children and postmenopausal women. The patients are, on average, roughly 30 years old (3). According to the average age of affected people, the occurrence of this tumor in children is highly rare, thus it is worth reporting. Tumors are functional in 40%-60% of cases, often estrogenic but occasionally androgenic or both, and progestin is only produced by the tumor in rare cases (4).

SLCT has a wide range of clinical manifestations, from asymptomatic to severe virilization. Patients with hormonally inactive neoplasms present with nonspecific symptoms such as pain or abdominal swelling, or their tumors are discovered incidentally. The imaging results of these tumors have yet to be determined due to the rarity of SLCT (5). The degree of tumor grading and staging has a substantial impact on ovarian SLCT prognosis. Considering that there are no established management protocol recommendations for SLCT, it is difficult to handle it. Young women favor fertility-paring surgery.

Microscopic Pathology

Sertoli cells proliferate in mature fibrous or hyalinized stroma and form these malignancies. A tubular pattern is typical. Sertoli cell tumors are divided into lipid-rich tumors with abundant clear, foamy cytoplasm and oxyphilic tumors with extensive granular eosinophilic cytoplasm (2). The majority of Sertoli cell tumors are highly differentiated, with homogeneous nuclei and few mitotic signs among the neoplastic cells. These tumors that are restricted to the ovary at the time of diagnosis and have minimal or no nuclear atypia and mitotic activity are considered benign (6).

Case Presentation

A 3-year-old girl was admitted to Kashani Hospital with abdominal pain and a protruding stomach. As part of the physical examination, a large mass was felt on the right side of the abdomen. Laboratory studies revealed that the tumour markers beta-human chorionic gonadotropin (HCG) and alpha-fetoprotein were both normal. On sonography, a large mass was found in the right ovary, measuring 8*5*4 cm. After surgery, the patient's ovarian tumour was removed (Figure 1) and transferred to the pathology department. Unfortunately, there was no photo of the patient before the operation and tumor removal. The patient had no history of surgery and no underlying disease.

Microscopic and Immunohistochemical Features

Under the microscope, the mass appeared red-brown and

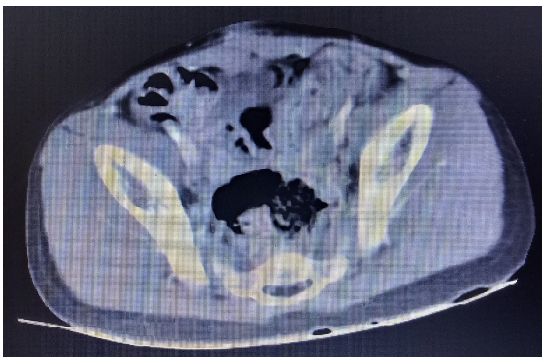

Figure 1. The patient's Ovary After Surgery and Removal of the Ovarian Tumor

solid, devoid of any hair tangles or dental components. On the other hand, an *immunohistochemistry (*IHC) analysis demonstrated that it was positive for actine, inhibin, calretinin, and WT1 while being negative for CD30, NSE, synaptophysin, vimentin, chromogranin, S100 and AFP, and Ki67. The final diagnosis, Sertoli cell tumor as a subset of sex cord-stromal tumor, was confirmed by microscopic and IHC findings. There was no evidence of metastasis in the abdomen or chest during computed tomography scan examinations. The patient is currently doing well and exhibiting no signs of recurrence 10 months after completing a chemotherapy course that included bleomycin, etoposide, and cisplatin.

Discussion

Sertoli cell tumors are among the most uncommon types of sex cord-stromal tumors (2). The patients' ages range from 2 to 79 years. The two largest series reported mean ages of 21 and 38 years and median ages of 33 and 50 years (4). The mentioned case was three years old, which is extremely rare and outside the average range. Bilateral SLCTs are extremely uncommon, accounting for less than 1%-2% of all SLCT cases (7, 8). According to a case series study, the vast majority of SLCTs are restricted to a unilateral ovary, particularly the right side (4/7, 57.1%) (9) (as in our case). Bilateral SLCTs are extremely rare, accounting for only about 1.5-2.0% of all SLCT cases (10).

At least one-third of SLCTs are associated with androgen-excess symptoms such as virilization, including oligomenorrhea, amenorrhea, breast atrophy, hirsutism, acne, voice deepening, clitoris enlargement, or baldness, as well as increased levels of other androgens. Additionally, SLCTs may exhibit estrogen-excess symptoms such as menorrhagia/metrorrhagia, postmenopausal bleeding, or diagnostic curettage, demonstrating an endometrium that is proliferating irregularly, hyperplasia, or endometrial cancer (8). The symptoms of the abdominal mass effect, including heaviness, abdominal discomfort or pain, ascites, or tumor rupture, may be attributed to SLCTs that have no endocrine manifestation (11).

Precocious pseudopuberty and vaginal bleeding are common in girls with hormonally active tumors. Depending on the type and amount of hormone secreted, older women experience irregular bleeding, postmenopausal hemorrhage, or, in rare cases, virilization. Patients with hormonally inactive neoplasms may experience nonspecific symptoms such as discomfort or abdominal swelling, or their tumors may be discovered by chance (9). In our study, a 3-year-old girl was admitted to the hospital, and a large mass was palpated on the right side of her abdomen. Although tumor markers (alpha photoprotein and beta-HCG) were normal in laboratory tests, a large mass measuring 8*5*4 cm was found in the right ovary on ultrasound. To our knowledge, this is the first known instance of a sex cord-stromal tumor in a child in Iran. Details regarding regular menstruation, past pregnancies, and births could not be accessed because our case was a young girl. This child had no prior history of taking any particular medication.

Most ovarian Sertoli cell tumors with solid architecture have lipid-rich tumor cells or have oxyphilic cytoplasm (6), although the features of our case did not closely reflect those previously reported in oxyphilic Sertoli cell tumors. Keratins, vimentin, and alpha-inhibin positivity vary among Sertoli cell cancers. Steroidogenic factor 1 and Wilm's tumor 1 stain the nuclei of tumor cells in almost all Sertoli cell cancers, making them useful, if not specific, immunohistochemical markers for these tumors. In nearly half of the cases, CD99 and calretinin are positive. Epithelial membrane antigen is absent in the malignancies (12,13). The tumor, in this case, was red-brown and entirely solid, with no hair tangles or dental components. Another diagnostic consideration was immunohistochemical features that showed positive results for actine, inhibin, calretinin, and WT1 in an IHC investigation, but were negative for CD30, NSE, synaptophysin, vimentin, chromogranin, S100, AFP, and Ki67.

Conclusion

Sertoli cell tumor should be considered a differential diagnosis in a young girl who has hirsutism, irregular bleeding, and breast atrophy with a pelvic mass. It is important to distinguish Sertoli cell tumors from stromal ovary, carcinoid, and endometroid carcinoma. If the androgen insensitivity syndrome has not been discovered preoperatively, phenotypic girls with the syndrome may be diagnosed accurately as having a Sertoli cell tumor of the ovary.

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Authors' Contribution

Conceptualization: Kiavash Fekri.
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Funding acquisition: Kiavash Fekri.
Methodology: Kiavash Fekri.
Project administration: Kiavash Fekri.
Supervision: Kiavash Fekri.

Validation: Kiavash Fekri, Shima Rahmati.
Writing–original draft: Shima Rahmati.
Writing–review & editing: Kiavash Fekri.

Competing Interests

The authors declared no potential conflict of interests in connection with the research, authorship, and/or publication of this article.

Ethical Approval

This study protocol was approved by Shahrekord University of Medical Sciences (IR.SKUMS.REC.1401.058). Additionally, informed consent was obtained from the parent of the patient for publication of this report.

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