Sertoli-Leydig cell tumor of ovary in a young female: A case report and literature review

Sertoli-Leydig cell tumor (SLCT) of the ovary is an exceedingly unusual neoplasm that belongs to a group of sex cord-stromal tumors of the ovary and accounts for less than 0.5% of all primary ovarian neoplasms. We present a case of primary ovarian SLCT in a 21-year-old female who presented with a 6-month history of pelvic pain, acne, hirsuitism and oligomenorrhea with elevated testosterone and dehydroepiandrosterone levels. Ultrasound and contrast computed tomography revealed a well-defined hypervascular heterogeneous solid lesion in the left adnexa. Magnetic resonance imaging demonstrated an ovarian origin of the tumor, which indicated an SLCT on histopathology. In this case, we discuss the multimodality imaging findings of SLCT and its management aspects with a review of the literature.

**Key words:** Acne, hirsuitism and oligomenorrhea, primary ovarian neoplasms, Sertoli-Leydig cell tumor

**INTRODUCTION**

Sertoli-Leydig cell tumor (SLCT) is a rare ovarian tumor that belongs to the group of sex-cord stromal tumors. These constitute less than 0.5% of ovarian tumors. Most tumors are unilateral and confined to the ovaries, and are seen during the second and third decades of life. These tumors are characterized by the presence of testicular structures that produce androgens. Hence, many patients have symptoms of virilization depending on the quantity of androgen production. We present a case of SLCT with multimodality imaging findings and its management issues.

**CASE REPORT**

A 21-year-old nulliparous female presented with complaints of pelvic pain, acne, hirsuitism, hoarseness of voice and oligomenorrhea for the last 6 months. No history of anorexia, weight loss, increased libido or breast recession was noted. Her medical and family history was unremarkable. Vaginal examination revealed clitoromegaly and a firm and mobile solid mass in the left adnexa. Laboratory tests showed elevated serum testosterone of 27.6 ng/mL (normal range 0.2-1.2 ng/mL) and dehydroepiandrosterone of 16.3 ng/mL (normal range 0.8-3.2 ng/mL) and normal CA-125 of 22 U/mL (normal range <35 U/mL). All other laboratory tests including complete blood count (CBC), renal, bone, hepatic and coagulation profiles, alkaline phosphatase, carcino-embryonic antigen (CEA), luteinizing hormone (LH) and follicle stimulating hormone (FSH) levels were within the normal range.

Further, the patient was referred for imaging workup to our department. Ultrasound showed a large, well-circumscribed, well-margined heterogeneous mixed solid-cystic lesion [Figure 1a] in the left adnexa with internal vascularity on color Doppler [Figure 1b]. The right ovary and the uterus were normal [Figure 1c].

Subsequently, cross-sectional imaging was performed; unenhanced computed tomography (CT) demonstrated a large, well-circumscribed solid lesion on the left side of the pelvic cavity [Figure 2a]. On contrast-enhanced computed tomography (CECT), the lesion showed heterogenous enhancement more in the peripheral part (corresponds to the solid part of the lesion on ultrasound) and less in the enhancing central part (corresponds to the cystic part of the lesion on ultrasound).
Figure 1: A 21-year-old female with left ovarian Sertoli-Leydig cell tumor. (a) Ultrasound shows heterogeneous mixed solid (thin arrow) and cystic (large arrow) lesion in the left adnexa (white arrow), whereas the left ovary is not visualized separately; (b) on color Doppler ultrasound, the left adnexal lesion shows internal vascularity (white arrow) and (c) ultrasound shows normal uterus (white arrow).

Figure 2: A 21-year-old female with left ovarian Sertoli-Leydig cell tumor. (a) NECT abdomen - axial section shows a large, well-defined, well-circumscribed solid soft tissue lesion on the left side of the pelvic cavity (white arrow). (b) On contrast-enhanced computed tomography of the abdomen-axial section, the lesion shows heterogeneous enhancement in the peripheral part [corresponds to the solid part of the lesion on ultrasound (small arrow)] and less-enhancing central part [corresponds to the cystic part of the lesion on ultrasound (large arrow)]. The black arrow shows a vascular pedicle to the lesion on its anterior aspect.
The left ovary is not distinctly visualized from the lesion. Blood supply to the lesion was coming from a vascular pedicle on its anterior aspect [Figure 2b]. The lesion causes displacement of the uterus inferiorly and toward the right side. The right adnexa appeared unremarkable [Figure 3a]. No evidence of paraaortic lymphadenopathy was noted [Figure 3b].

Nonenhanced magnetic resonance imaging (MRI) of the pelvis - T2-weighted images showed heterogeneous signal intensity lesion with few central cystic areas in the left adnexa. The lesion causes compression and peripheral displacement of the follicles, suggestive of ovarian origin [Figure 4]. The lesion showed multiple signal voids. The uterus and right ovary appeared unremarkable [Figure 5a]. The T1-weighted image demonstrated isointense signal and multiple signal voids [Figure 5b]. On the basis of the above clinical, biochemical and imaging findings, a diagnosis of androgenic neoplastic primary ovarian tumor was made.

The patient underwent exploratory laparotomy. Intraoperatively, the left ovary was replaced by a large capsulated solid grey-white mass [Figure 6a]. The right ovary and uterus were found unremarkable. Left salpingo-oophorectomy was performed. Gross examination of the pathologic specimen [Figure 6b] showed an ovarian mass with a smooth external grayish surface. A cut-section of the specimen [Figure 6c] revealed solid as well as cystic areas filled with clear fluid. The histopathological examination showed a tumor composed of poorly formed cords, nests and tubules of tumor cells. Tumor cells had hyperchromatic nuclei, a moderate amount of cytoplasm (Sertoli cells) and occasional mitosis [Figure 6d]. Interspersed between these were nests of polygonal cells with round nuclei and abundant granular cytoplasm (Leydig cells). Final diagnosis was intermediate differentiated SLCT on histopathology.
Figure 5: A 21-year-old female with left ovarian Sertoli-Leydig cell tumor. Nonenhanced magnetic resonance imaging of the pelvis - (a) T2-weighted axial section, the left ovarian lesion shows multiple signal voids (large arrow) and causes displacement of the uterus toward the right side (curved arrow). The uterus and right ovary appear unremarkable (small arrow). (b) T1-weighted axial section, the left ovarian lesion shows an isointense signal intensity (small arrow) and multiple signal voids (large arrow).

Figure 6: (a) Intraoperative highly vascular ovarian lesion (small arrow) and fallopian tube (large arrow) and uterus (black arrow). (b) Gross specimen shows a well-encapsulated grayish solid mass. (c) The cut-section of the specimen shows a tan-yellow nodular appearance (white arrow). (d) Microscopically, photomicrograph showing hyperchromatic nuclei, moderate cytoplasm [Sertoli cells (black arrow)] and nests of polygonal cells with round nuclei and abundant granular cytoplasm [Leydig cells with heterologous elements showing mucinous epithelium of the gastrointestinal type (white arrow)]. (H and E, x40)
Surgical resection represents the mainstay of management of ovarian SLCTs. Therefore, fertility-sparing surgery (unilateral salpingo-oophorectomy) can be considered in all patients with well-differentiated ovarian SLCTs. Generally, postoperative chemotherapy is considered for patients with poor prognostic factors such as advanced disease staging, moderate-to-poor tumor grading, high mitotic profile, existence of heterologous elements and tumor rupture. Prognosis of ovarian SLCTs is significantly correlated with degree of tumor differentiation (grading) and tumor extent (staging). Long-term follow-up is highly advised in all patients.

CONCLUSION

SLCT is a rare ovarian sex-cord tumor of the ovary with good prognosis postoperatively. Therefore, imaging findings may help in the early detection and preoperative differentiation and staging of SLCT in young females. Its management depends on the degree of differentiation and staging of tumor, which mostly depend on histopathology. Intermediately differentiated tumors require additional management of chemotherapy along with conservative surgery to prevent recurrence.

REFERENCES


How to cite this article: Gupta MM, Bahri NU, Rathod K. Sertoli-Leydig cell tumor of ovary in a young female: A case report and literature review. Onc Gas Hep Rep 2014;3:40-4.

Source of Support: Nil, Conflict of Interest: None declared.