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Laparoscopic assisted removal of large ovarian mass causing precocious puberty in an infant—a case report

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Abstract

Background Sex cord and stromal tumors are a heterogeneous group of tumors that arise from gonadal sex cord cells, gonadal stromal cells, or both. They are divided into pure stromal tumors, pure sex cord tumors, and mixed tumors. Some of these tumors are hormonally active, producing androgens and estrogens, and may therefore exhibit virilization or excess estrogen. Sertoli-Leydig cell tumors are rare tumors belonging to mixed tumors representing less than 0.5% of ovarian tumors. Few cases have been reported in the medical literature.

Case presentation We report the case of a 1-year-old girl who was admitted for breast lumps and pubic hair with intermittent painless vaginal bleeding in the past 2 weeks. The abdominal examination objectified the presence of an intraperitoneal mass to which an abdominal ultrasound and computerized tomography (CT) scan had confirmed the presence of a right ovarian mass. A laparoscopy was performed which showed an encapsulated mass arriving from the right ovary replacing the normal ovarian tissue. Laparoscopic-assisted removal of the entire mass was done. The postoperative course was uneventful. The histopathological study demonstrated an intermediately differentiated Sertoli cell tumor with an intact capsule, no lymphovascular invasion, or heterologous elements (TNM stage 1).

Conclusions Although Sertoli-Leydig tumors are commonly seen after the second decade of life in young women, their discovery before puberty is not uncommon. Mass syndrome and signs of virilization are two frequently reported signs that should prompt the clinician to mention this entity among the differential diagnoses. The first-line radiological examination is pelvic ultrasound. The treatment is mainly based on surgery. The prognosis is excellent especially if the tumor is well differentiated.

Keywords Ovarian mass, Laparoscopy, Precocious puberty

Introduction

Sex cord tumors account for 7% of all primary malignant ovarian tumors [1]. They are a heterogeneous group of benign and malignant tumors derived from stromal cells or primitive sex cord cells [2, 3]. They develop in the first two to three decades of life and have a variable spectrum of clinical presentation such as abdominal distention and abdominal mass [4]. Some sex cord-stromal tumors have clinical features related to hormone production including precocious puberty, hirsutism, virilization, and menstrual changes [5]. We report an interesting case of a 1-year-old girl with a large ovarian mass causing precocious puberty which was operated laparoscopically.

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Clinical presentation

A 1-year-old girl was referred to our hospital with a 2-month history of developing breast nodules and pubic hair. The child also had a history of intermittent painless vaginal bleeding for the last 2 weeks. On general examination, the child weighed 12.5 kg and the height was 50 cm. She had bilateral breast nodules and pubic hair. There was no history of any similar complaints in the past and there was no history of any major family illness. Abdominal examination showed her to have a 5×5 cm firm, non-tender intraperitoneal mass with restricted mobility. An ultrasound scan confirmed this to be a right ovarian mass. CT scan confirmed a solid right ovarian tumor with small irregular areas of necrosis and displacing the intestinal loops. MRI Brain was normal. 17-OH Progesterone, Thyroid function tests were within normal limits. Serum testosterone was 234 ng/dl (2–10 ng/dl), Prolactin 10.5 ng/ml (5.18–26.53 ng/ml), alpha-fetoprotein 3.64 ng/ml (0.1–0.2 ng/ml), estradiol (E2) 234 pg/ml (20–53 pg/ml), and β-HCG 10.08 mui/ml (0–1 mui/ml).

In view of the history and findings, laparoscopy was planned. The child was placed supine with the monitor at the foot end. Bladder was catheterized. Three 5-mm ports were used. The camera was placed at the umbilical port and two working ports were placed on either side with a CO2 pressure of 10 mm Hg. At laparoscopy, a large well-capsulated mass (5×5 cm) was seen arising from the right ovary which had entirely replaced the normal ovarian tissue. The left ovary, uterus, and both the fallopian tubes were normal. The entire mass was excised laparoscopically using a harmonic scalpel. The right fallopian tube was preserved. The mass was delivered out of the abdomen by placing a small hypogastric incision (Fig. 1). Post-operative course was uneventful and the child was commenced on feeds 6 h after surgery. The urethral catheter was removed 24 h after surgery and the child was discharged 72 h after surgery. Histology showed an intermediately differentiated Sertoli cell tumor with an intact capsule, no lymphovascular invasion or heterologous elements (TNM stage 1). The child is doing well in follow-up and the size of the breast nodules has regressed after 6 weeks of surgery. There has not been any further episode of vaginal bleeding after surgery. The pubic hair disappeared 6 months after surgery.

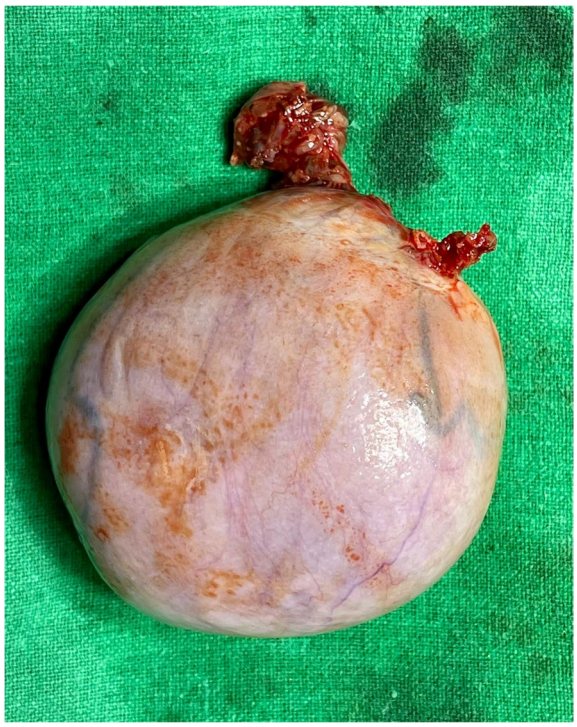

Fig. 1 Photograph of the ovarian mass after removal

Discussion

Sertoli-Leydig cell tumors (SLCT) are rare female reproductive cancers that account for less than 0.5 percent of all ovarian tumors [4]. SLCT can occur at any age, but 75% is present in women under the age of 30 [6]. The pathogenesis of the disease is still unknown. Certain genes such as DICER1, STK11, FOXL2 as well as somatic changes have been implicated in ovarian sex cord-stromal tumors [7]. Germline DICER1 mutations are observed in approximately 60% of SLCTs. Patients with the DICER1 gene mutation develop these tumors at a younger age [8].

Ovarian Sertoli-Leydig cell tumors (OSLCT) are composed of Sertoli cells which produce mainly androgens, some progestin and estrogen and Leydig cells producing mainly estrogen, some progestin, androgen and fibroblasts. Clinical manifestations vary by the composition of the OSLCT. The typical clinical features indicative of an androgenic effect are virilization and hirsutism while the clinical features of an estrogen-secreting tumor are breast swelling, pubic hair, and vaginal bleeding [9]. In clinical practice 40–60% of individuals show signs of virilization and estrogenic symptoms are rare [8]. Acute abdominal symptoms may occur in some patients as a result of torsion of the adnexa/ovaries, capsular rupture, or bleeding [4].

OSLCT is classified as high, intermediate or low differentiation based on tissue morphologies, as well as a reticular growth pattern and heterogeneous component [8]. They may contain heterologous elements, retiform components, or both [8]. More than 95% of these tumors are unilateral FIGO stage 1, and moderate to poorly differentiated [10]. The malignant potential is 0% for well-differentiated tumors, 11% for intermediate tumors, 59% for poorly differentiated tumors, and 19% for tumors

Table 1 Patients' presentation, imaging findings, laboratory findings, management and prognosis of cases published in the last 10 years

Author, year	Patient	Clinical manifestations	Laboratory work up	Radiological features	Treatment	Follow-up
Abu-Zaid et al. (2013) [13]	16-year-old	3-month history of a pelviabdominal mass, acne, hirsutism, and menstrual irregularities	Increased total serum levels of testosterone (T), dehydroepiandrosterone (DHEA), and CA-125	US: intraperitoneal ascites and extremely huge well-vascularized mass with cystic and solid components, mostly arising from left ovary CT: huge, intraperitoneal, complex, cystic, and multilocular lesion, extending from pelvis up to mid-abdomen just above umbilicus, collectively measuring 13.5 × 23.3 × 21.5 cm	Left unilateral salpingo-oophorectomy, omentectomy, and appendectomy	A postoperative 3-month follow-up failed to show any evidence of recurrence
Cabrera-Cantu et al. (2014) [15]	12-year-old girl	Acute abdominal pain with no other clinical manifestations	US: semisolid mass 8.0 × 6.0 × 6.4 cm with cystic areas CT: well-defined 7.4 × 9.5 cm cystic-solid tumor with small irregular areas of necrosis	Lactic dehydrogenase, alpha-fetoprotein, chorionic gonadotropin and carcinoembryonic antigen were normal	A unilateral salpingo-oophorectomy was done by laparoscopy	Follow-up of 2 months: no complications Follow-up of 1 year: no recurrence
Sarkar et al. (2021) [8]	Case 1: 2-year-old	Abdominal pain and distension	Ultrasonography followed by CT: large cystic lesion with septa measuring 17.0 cm across, compressing the adjacent gut loops suggestive of a large mesenteric cyst	Increased serum level of CA-125	Exploratory laparotomy revealed right ovarian mass. Excision of the right ovarian mass with an intact capsule	NA
	Case 2: 2-year-old	3-month history of abdominal pain	US: pelvis mass measuring 8.0 cm across lying just above the uterus	Beta-hCG, alpha-fetoprotein, and CA-125 levels were within normal limits	Exploratory laparotomy with resection of left ovarian mass with intact capsule	The patient is doing well on a follow-up of 3 years
Turkyilmaz et al. (2016) [16]	8-year-old	Huge mass and pain in the lower abdomen	US and CT: solid cystic mass about 24 × 17 × 12 cm in size arising from the subhepatic region to the pelvic area, with multiple thick septations in the cystic lesion	The level of serum CA125 was increased	Exploratory laparotomy and removal of the mass by right salpingo-oophorectomy	Recurrence of the tumor after 10 months of the first operation

Table 1 (continued)

Author, year	Patient	Clinical manifestations	Laboratory work up	Radiological features	Treatment	Follow-up
Gómez-Peñaloza et al. (2018) [17]	11-year-old	Palpable mass in the left iliac fossa with clinical characteristics of virilization	US: Well-defined solid–cystic lesion of 102 × 88 × 62 mm with limited vascularity CT: right ovarian lesion with characteristics similar to those found on ultrasound, displacing intestinal loops and partially compressing the right ureter	Slightly increased level of testosterone	A unilateral salpingo-oophorectomy was performed	Twenty-nine months later, a new abdominal mass was discovered; a salpingo-oophorectomy was performed and a second SLCT, poorly differentiated was diagnosed
Present case	1-year-old	Breast enlargement, pubic hair with intermittent painless vaginal bleeding with retroperitoneal mass	US: ovarian mass, CT: solid right ovarian tumor with small irregular areas of necrosis and displacing the intestinal loops.	Increased level of testosterone, alpha-fetoprotein, estradiol and beta HCG	Laparoscopic assisted removal of right ovarian mass	Uneventful

US, ultrasonography; *CT*, computed tomography; *NA*, not available

with heterogeneous components [11]. The final diagnosis is histopathological with the best reliability coming from a combination of hematoxylin and eosin (H&E) staining and immunohistochemical testing [12].

Ultrasound remains the imaging modality of choice for the initial assessment of adnexal tumors [13]. Alternative imaging modalities such as computed tomography (CT), magnetic resonance imaging (MRI), and positron tomography (PET) may be used for improved characterization of ovarian SLCTs. SLCTs must be differentiated from papillary serous tumors, yolk sac tumors, endometrioid carcinomas, carcinoid tumors and carcinosarcomas. The use of immunohistochemistry helps in accurate diagnosis in difficult cases [8]. Immunohistochemistry was not carried out in our case as the diagnosis was clear on H&E staining.

Due to the rarity of the tumor, evidence on the optimal treatment of SLCT is limited. It is based on a small number of case reports and retrospective series [13, 14]. Table 1 summarizes the patient presentation, imaging findings, laboratory findings, management and prognosis of some cases published in the last 10 years. Surgery remains the cornerstone of treatment and appears to be the most successful method when possible [13]. Because the majority of tumors present as limited-stage FIGO Ia tumors, fertility-preserving surgery such as oophorectomy or adnexectomy, may be the only treatment option for these tumors [13, 14]. Prior ovarian biopsy is discouraged [14]. Poor prognostic parameters such as advanced disease stage, moderate to poor tumor grading, high mitotic profile, presence of heterologous components and tumor rupture are considered for postoperative chemotherapy [13, 14].

In general, SLCTs have a favorable prognosis. However, the presence of poor differentiation, retiform developmental patterns and heterologous components can contribute to a poor outcome [8].

Conclusion

Sertoli-Leydig cell tumor (SLCT) of the ovary is a rare ovarian neoplasm that belongs to the sex cord stromal malignancies category and few cases have been documented in the literature. It must be evoked in the differential diagnoses of androgen-excess manifestations which are frequent or estrogen-excess manifestations which remain rare. The final diagnosis relies on a histopathological study with immunohistochemistry when indicated. Surgical treatment remains the cornerstone, particularly in the case of a localized tumor (stage Ia). Although the prognosis depends on the degree of tumor differentiation and tumor extent, the overall 5-year survival rate for stage I is favorable.

Abbreviations

SLCT Sertoli-Leydig cell tumors
OSLCT Ovarian Sertoli-Leydig Cell tumor

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Authors' contributions

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Declarations

Ethics approval and consent to participate

Not necessary as this is a case report from a private hospital owned by the author. Consent for surgery has been obtained from the family.

Consent for publication

Consent for publication has been obtained from the family.

Competing interests

The authors declare that they have no competing interests.

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