Case Report

Sertoli Leydig cell tumor with rare form of presentation

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INTRODUCTION

Ovarian sex cord-stromal tumors are rare kinds of neoplasm of the upper female genital tract. Sertoli-Leydig cell tumor of ovary belongs to sex cord-stromal tumors of ovary and accounts for less than 0.5% of all primary ovarian neoplasms. It is suggested to be arising either from gonadal mesenchyme of ovary or from remnants of hilum.

CASE REPORT

Mrs. Rukkumani, 30 year old multiparous lady, presented to us with abdominal pain for 2 days and abdominal distension for 4 months, with a diagnosis of fibroid uterus or ovarian mass, referred to our institution for further evaluation.

She had associated loss of weight, loss of appetite, menorrhagia for 10 yrs, for which she did not undergo any treatment. On examination, she was poorly built and nourished. General condition was good. Vitals were stable. Examination revealed a pelvic mass up to the level of umbilicus, not felt separately from the uterus. She was subsequently evaluated with pre-operative investigations; CA 125 was 62.2, in favour of malignancy. Ultrasound showed a 19.5 x 11.1cms well defined, predominantly solid mass lesion noted arising from the right adnexa occupying pelvis and lower abdomen. The lesion showed few small 1 cm diameter areas within it. There were no calcifications, but was associated with increased internal vascularity. Low resistance arterial flow was noted. Similar lesion of 7.2 x 3.27 cms seen arising from the left adnexa. MRI Pelvis was done, reported as bilateral solid neoplastic ovarian lesions with associated moderate ascites, likely of sexcord stromal origin, more of fibrothecoma spectrum. Right ovarian lesion measuring 14.8 x 10 x 14 cms and left ovarian lesion measuring 8.8 x 4.7 x 7 cms. Risk malignancy index was 186, by which she comes into low risk category for malignancy (the cut off being 250).
She underwent staging laparotomy followed by total abdominal hysterectomy with bilateral salpingo-oophorectomy, infracolic omentectomy. Gross findings - There was a right ovarian tumor measuring 15x20 cms and left ovarian tumor measuring 5 x 6 cms. 30 ml ascitic fluid was aspirated. Post-operative period was uneventful.

By the above HPE report, FIGO surgical staging of this patient comes as STAGE 3B. Since she required post-operative chemotherapy; she was referred to higher centre.

**DISCUSSION**

Ovarian sex cord-stromal tumors are rare kinds of neoplasm of the upper female genital tract. Sertoli-Leydig cell tumor of ovary belongs to sex cord-stromal tumors of ovary and accounts for less than 0.5% of all primary ovarian neoplasms.\(^1\)

It is characterized by uncontrolled proliferation of naturally occurring testicular structures (Sertoli and Leydig cells) of varying degrees of differentiation which include well differentiated, moderately differentiated, poorly differentiated, and with heterologous elements.\(^3\) It is suggested to be arising either from gonadal mesenchyme of ovary or from remnants of hilum. Patients with SLCT present most commonly in second and third decades of life. These are usually unilateral but bilateral tumours can be seen in < 2% of cases.\(^2\)

Clinical signs and symptoms of SLCT can be related to either hormonal production or presence of mass-
occupying lesion. While SLCTs can be functionally inactive, abnormal hormonal production (mostly androgen or rarely estrogen excess) can be identified in more than half of patients. Clinical expression of virilization is recognized in more than one-third (33–38%) of patients. Androgen-excess manifestations with varying degrees include virilism, hirsutism, hyperseborrhea, acne, receding hairline, alopecia, and hoarseness of voice, loss of subcutaneous tissue deposits, breast atrophy, clitoromegaly, oligomenorrhea and amenorrhea. Manifestations suggestive of excess estrogen include: precocious puberty, abnormal uterine bleeding, abnormal vaginal bleeding, menstrual irregularities, generalized edema, weight gain, breast hypertrophy, endometrial hyperplasia, endometrial polyps and endometrial carcinoma.  

Macroscopically, the majority of SLCTs range in size between 5 and 15 cm in diameter. In addition, SLCTs can be solid, solid and cystic or cystic. Average SLCT diameter is 13.5 cm and can reach as huge as 50 cm in poorly differentiated histological variants. However, as the size of SLCTs can sometimes be undetectable by ultrasonic, other imaging modalities such as computed tomography (CT), magnetic resonance imaging (MRI), and positron imaging tomography (PET) scans can be used for better recognition of SLCTs. The malignant potential of this tumor is lower than that observed for epithelial ovarian tumors, and the survival rate is 70% to 90% after a 5-year follow-up. Though the MRI calculation showed low risk for malignancy, the HPE report turned out to be malignancy in our case.

On USG, they have solid appearance, richly vascularization, lower vascular resistive index and peripheral necrosis. In general, malignant tumors have neovascularization and distension of pre-existing vessels, which result in a low resistance to blood flow. On the basis of endocrine symptoms, the woman’s age and ultrasound findings; it should be possible to suggest a correct preoperative diagnosis of Sertoli cell, Sertoli-Leydig cell or Leydig cell tumors in many cases. Standard management guideline of ovarian SLCTs is still uncertain. Recommended treatment varies with patient age, tumor stage, and differentiation. Surgery is the initial method to deal with SLCTs. Fertility-sparing surgery (unilateral salpingooophorectomy) can be carried out in all well-differentiated ovarian SLCTs. Patients desiring fertility but exhibiting moderately or poorly differentiated ovarian SLCTs can be considered for unilateral salpingo-oophorectomy plus standard staging surgery (omentumectomy, appendectomy, and pelvic lymphadenectomy). Patients who do not desire fertility or poorly-intermediately differentiated with intraoperative evidence of rupture and mesenchymal heterogenous elements containing subtypes should be considered for total hysterectomy, bilateral salpingooophorectomy in addition to complete standard staging surgery.

Patients should be followed up with serum testosterone levels 3 months once for one year, 4 months once for one year, 6 months once for one year and then yearly once for rest of life. During follow-up, history, examination, serum testosterone and ultrasound abdomen and pelvis are done. If required, CT or MRI may be done. Age of the patient, stage of the disease and degree of tumor differentiation based on morphology are the most important factors considered in the management of the case.  

As most Sertoli-Leydig cell tumors of the ovary are seen in young women and detected in early stages, favourable outcome can be achieved by conservative surgery. SLCTs have a good prognosis although we find recurrence of poorly differentiated tumors. The prognosis of patients with grade 1 SLCT is excellent without adjuvant chemotherapy. Patients with tumors of grade 2 or 3 appear to benefit from postoperative chemotherapy.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES
