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CASE REPORT

Ovarian Sertoli-Leydig Cell Tumor with Estrogenic Manifestations in a Postmenopausal Lady: a Case Report

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Introduction

Sertoli-Leydig cell tumors (SLCT) are rare tumors, constituting less than 0.5% of ovarian malignancies [1]. They are predominantly solid but cystic areas may be seen [2]. The usual age of presentation is during the second and third decades of life while < 10% present in premenarchal and postmenopausal age group [1]. In the last WHO classification (2014) [3], it has been classified as mixed sex cord-stromal tumors (SCST) and is pathologically staged according to FIGO staging system [4].

We present an unusual case of SLCT in a postmenopausal woman with estrogenic manifestations presenting with large pelvico-abdominal mass.

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Case Report

A 52-year-old, multiparous, postmenopausal woman presented with complaints of lower abdominal pain, loss of appetite, weight loss, on and off burning micturition of 1-month duration, also complained of constipation and on and off bleeding per vaginam for last 1 week. There was no history of any androgenic features. She was a known hypertensive on irregular medication and had history of cerebrovascular accident 2 months back. Her family history was unremarkable.

Her general physical examination was normal. On per abdominal examination, large, firm, fixed central mass arising from the pelvis up to the level of umbilicus was felt. Vaginal examination revealed fullness in all the fornices, no nodularity in the pouch of Douglas while on per rectal examination, an irregular mass was felt high up, rectal mucosa was smooth and free.

USG Imaging

During the initial work up with tran-vaginal ultrasound scan, which was done at some other hospital, a 15.3 × 9.6 cm multilocular, pelvico-abdominal heterogenous mass lesion with solid and cystic components was seen in the left adnexa. Uterus was bulky with endometrial thickness of 15 mm. Risk of malignancy index was 563; hence, she comes under high risk category for malignancy.

PET CT

PET CT showed a 13.5 × 18.4 × 16.8 cm well encapsulated hypodense central mass lesion in the pelvis (SUV-19). No significant pelvic or para aortic lymphadenopathy, omentum was thickened and nodularity seen (SUV-1.2).

Tumor markers were as follows: CA 125–62.6 U/ml, Ca-19-9:29.8 U/ml, CEA 1.03 ng/ml.

Endometrial biopsy was planned, but the patient refused for the procedure. She lost to follow-up for a month, then she came to OPD with frequent episodes of bleeding per vaginam after which she was directly taken up for surgery. She underwent laparotomy with infra umbilical incision (total abdominal hysterectomy with bilateral salpingo-oophorectomy with infra colic omentectomy). Pelvic and para aortic lymph node dissection was not performed.

Operative findings (Fig. 1) were 20 × 25 cm large complex, encapsulated tumor arising from the left ovary. There were no palpable lymph nodes, rest of the intraperitoneal survey was normal. Approximately, 200 ml of ascitic fluid was removed, cytological examination of which was negative for malignant cells.

Frozen section showed features of malignant stromal tumor of ovary.

Grossly, the left ovarian mass measured 24 × 18 × 10 cm. The capsule was intact with smooth external surface. On cut section, 40% solid and 60% cystic areas with hemorrhagic and serous content was seen.

On histopathological examination (Fig. 2), microscopy showed features of malignant sex cord stromal tumor with low mitotic activity. Right ovary, bilateral fallopian tubes, omentum were normal. The endometrium thickness was 0.4 cm, showing features of disordered proliferative phase endometrium, while cervix showed features of chronic cervicitis.

Immunohistochemistry:

IHC marker	
Inhibin	Positive
Calretin	Positive
CD10	Negative
CD56	Negative
CD99	Negative
WT-1.	Negative

Based on the above mentioned findings, diagnosis of Stage IA, moderately differentiated Sertoli Leydig cell tumor without heterologous element was established.

Her postoperative period was uneventful and she was discharged on postoperative day 7. After discussing the case in our multidisciplinary clinic, a consensus was made for adjuvant treatment with chemotherapy, but she refused any further treatment.

Discussion

Sertoli-Leydig cell tumors are extremely rare neoplasm, with unilaterality in 98% of cases while < 2% are bilateral. The usual presentation of these tumors are related to androgen excess (manifesting as amenorrhea, breast atrophy, loss of subcutaneous fat, masculinizing features like deepening of voice, hirsutism) or rarely due to estrogenic manifestations like postmenopausal bleeding, endometrial hyperplasia, myoma, endometrial, or breast cancers [5].

Fig. 1 Intra operative finding. 20 x 25cm large complex , encapsulated tumor arising from the left ovary

We reviewed the literature on SLCT and found 12 out of 33 postmenopausal women with SLCT presented with features of hyperestrogenism [1, 4–7].

Our patient had attained menopause 2 years back and recently presented with on and off bleeding per vaginam, had no features of virilization or defeminization. Her initial assessment with ultrasonography showed endometrial thickness of 15 mm, indicating hyperestrogenism. Monappa V et al. [2] and Rubio et al. [8] have reported similar cases of postmenopausal woman of SLCT with features of hyperestrogenism.

Hormonal assays like serum testosterone, inhibin, androstenedione, dehydroepiandrosterone, estradiol, FSH aid to the diagnosis but since there was no suspicion of SLCT in our case and due to financial constraints, these were not taken into consideration prior surgery to establish the diagnosis.

Complete surgical staging is the mainstay of treatment for SLCTs. Lymph node metastases is rare, hence pelvic and para aortic lymphadenectomy can be omitted [5]. Patients with early stage SLCTs, desiring pregnancy, fertility sparing surgery with unilateral salphingo-oophorectomy is offered followed by complete surgical staging after child-bearing is complete [5, 9]. Those patients with low-risk stage I SLCTs observation is recommended, while those with high risk features like poor differentiation, high mitotic count, presence of heterologous element, rupture of tumor, stage 1C, the recommendation is to offer observation or platinum-based chemotherapy. Recommendation for women with stage II to IV disease is adjuvant platinum-based chemotherapy, while limited

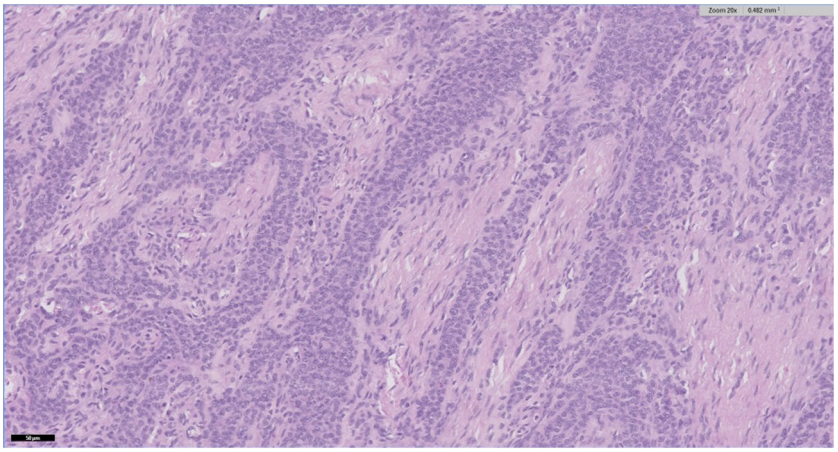

Fig. 2 Histopathologic findings of the ovarian Sertoli-Leydig cell tumor tubules of round to oval sertoli cell are surrounded by Leydig cells with pale granular cytoplasm. Features of malignant sex cord stromal tumor with low mitotic activity

data suggest radiotherapy as an alternative option particularly for patients with disease limited to the pelvis. [4]

Immunohistochemistry studies in the present case showed positivity for inhibin and calretinin which are considered specific markers for SCST [5]. Zhang H Y et al. [4] and Kim Y et al. [6] have emphasized the use of calretinin and vimentin on immunohistochemistry in aiding the diagnosis of SLCTs.

Prognosis is usually good and depends upon the grade and stage of the tumor. The presence of high risk factors is associated with increased risk of metastasis [2]. In our patient, the tumor was moderately differentiated with low mitotic activity without any intraoperative rupture or heterologous element; hence, even though she opted not to take adjuvant treatment, the probable risk of metastasis is less. Patients with SLCTs should be followed up with serum testosterone, ultrasound abdomen and pelvis every 3 months during the first year, every 4 months during second year, six monthly during the third year and thereafter annually for the rest of their life. At each visit history and examination is necessary and if needed CT or MRI of the abdomen and pelvis can be done. [4]

Conclusion

Sertoli-Leydig cell tumor is a rare ovarian neoplasm belonging to sex cord stromal tumors. Presentation of this tumor in a postmenopausal age group with hyper estrogenic features is unusual; moreover, absence of androgenic hormonal symptoms may mislead the diagnosis. Histopathology when combined with immunohistochemistry aids in establishing more accurate and definitive diagnosis.

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