DOI: https://doi.org/10.4038/sljom.v5i2.60
Volume 05 Issue 02
September 2024
SLJOM The Sri Lanka Journal of Menopause

Case Report

Rare case of Sertoli-Leydig cell tumour of the ovary

Raguraman S[1], Yogasingham R[2], Thillainathan V[2]

[1]Consultant in Obstetrics and Gynaecology, Department of Obstetrics and Gynaecology, Teaching Hospital Jaffna.
[2]Teaching Hospital Jaffna.
***Corresponding author -** Dr. S. Raguraman*
Email: raguraman@univ.jfn.ac.lk, sivalingarajahraguraman@gmail.com

Introduction

Sertoli-Leydig cell tumours (SLCTs) are a rare and heterogeneous group of ovarian neoplasms which originate from the ovarian sex cord (Sertoli cell tumour, Granulosa cell tumour) or stomal cells (Fibroma, Thecoma, Leydig cell tumour) account less than 0.5% of primary ovarian neoplasms[1]. It commonly occurs in reproductive age groups in their third and fourth decade. It is usually presented as a unilateral mass early, with clinical manifestations varying from asymptomatic to extreme virilization with metastasis. Diagnosis of SLCT is made with histological analysis with a supportive clinical assessment. Management remains challenging due to a lack of standardized management protocols and guidelines. Since it is common in the reproductive age group, fertility-sparing surgery plays a vital role in the management[2].

In this paper, we report a case of a postmenopausal woman with SLCT presented with hirsutism and successfully managed with surgery.

Case presentation

A 60-year-old woman presented with dull-aching recurrent lower abdominal pain for one year. She did not have urinary symptoms, vaginal discharge, or vaginal bleeding. She complained of excessive hair growth on the face, like a male hair pattern. But there are no other features of hyperandrogenism or virilizations. She underwent a lower segment cesarian section, laparoscopic cholecystectomy, and total abdominal hysterectomy for heavy menstrual bleeding due to fibroids. She has hypothyroidism and hypertension, both well controlled with medications, and no family history of malignancy. On examination, she has facial hair growth with a Ferriman-Gallwey classification of 2 in the upper lip and 3 in the chin. Her abdomen examination was normal, and no adnexal masses were found in the bimanual vaginal examination. Ultrasonography of

the abdomen and pelvis revealed a well-defined lobulated solid cystic mass of 6.1 cm × 4.3 cm × 3.9 cm in the left adnexa, which could not be distinguished from the left ovary (Figure 1).

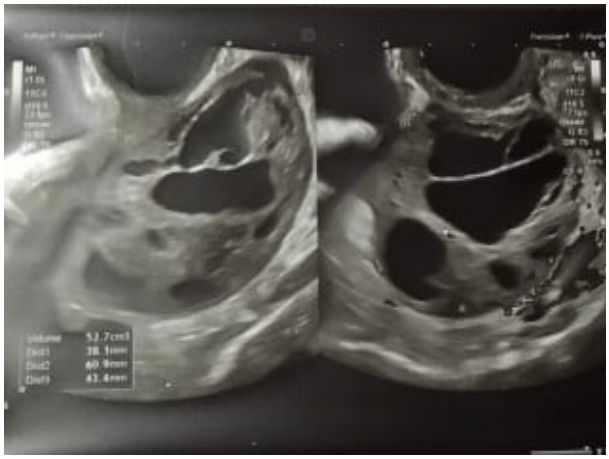

Figure 1 - Ultrasonic feature of well-defined lobulated solid cystic left ovarian mass

When we consider the tumour markers, Cancer Antigen- 125 (CA-125) level was 10.3 U/mL, beta human chorionic gonadotropin (β-hCG) level was 2.8 IU/L, alpha-fetoprotein (AFP) level was 4.2 ng/mL, carcinoembryonic antigen (CEA) level was 2.1 ng/mL and inhibin level was 2.2 pg/mL. Her testosterone level was 4.15 ng/d. All the tumour markers and total testosterone were in the normal range.

Since her Risk Malignant Index (RMI) is low, she underwent laparotomy with bilateral salpingo-oophorectomy with complete excision of the left ovarian mass. Intra-operatively, a well-defined and smooth solid and cystic ovarian mass of 6 cm × 5 cm (Figure 2) was identified and completely excised. Peritoneal wash was taken for cytological assessment. Cytopathology did not show malignant cells, and histopathological examination revealed a normal right-side ovary and tubes and left-side mass with features of moderately differentiated Sertoli - Leydig cell tumour with International Federation of Gynecology and Obstetrics (FIGO) staging was IA. The postoperative period was uneventful. Since the patient had stage IA ovarian tumour and post-operative medical oncological team did not recommend adjuvant chemotherapy. She is on regular follow-up and is doing well six months after surgery.

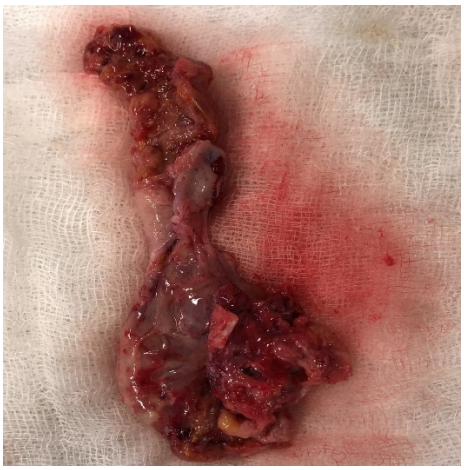

Figure 2 - Completely excised ovarian mass

Discussion

A Sertoli-Leydig cell tumour is a rare type of sex cord-stromal tumour of the ovary, accounting for approximately <0.5% of all primary ovarian neoplasm[3]. It is common in the reproductive age group, and less than

10% can be found prior to menarche and after menopause[2]. Most SLCTs have an excellent prognosis due to early diagnosis with well-differentiated stages[4]. Our case illustrated a 60-year-old postmenopausal woman with SLCT.

Clinical manifestations of SLCT include oligomenorrhea, hirsutism, breast atrophy, acne, lower tone of voice, laryngeal protuberance, and clitoromegaly[1]. Endocrine manifestations are pathognomonic symptoms, and virilization and defeminization have been described in up to 77% of the cases due to testosterone secretion[5]. About half of SLCTs have no endocrine manifestation, and patients presenting symptoms may be related to abdominal mass effects such as a feeling of heaviness, abdominal discomfort or pain, ascites, or tumour rupture[6]. In this case, she presented with recurrent abdominal pain and hirsutism. Furthermore, her testosterone level was normal.

Hirsutism may be the result of either androgen excess or increased sensitivity of the hair follicle to androgens. Most often, hirsutism is an expression of elevated levels of testosterone. Hyperandrogenism may be caused by excessive production of androgens by the ovaries (polycystic ovary syndrome [PCOS], tumors) or adrenal glands (non-classical adrenal hyperplasia [NCAH], tumors), insulin resistance, idiopathic hyperandrogenism, acanthosis nigricans (HAIRAN syndrome), Cushing syndrome, the menopause, and certain medications. Not all patients with hirsutism, however, have a detectable androgen excess; approximately 10 to 15% of women with hirsutism suffer from idiopathic hirsutism. The term idiopathic hirsutism should be applied only to hirsute women with a normal ovulatory function and normal detectable androgen levels. The cause of this indifferent hair growth in these women is thought to be associated with abnormalities in peripheral androgen activity or increased sensitivity of the hair follicle[7]. In the case of our patient, hirsutism may be attributed to idiopathic hirsutism.

Another rare presentation of SLCT is estrogenic manifestations, such as postmenopausal bleeding, endometrial hyperplasia, and endometrial carcinoma due to estrogen secretion by Sertoli cells or conversion of testosterone to estrogen at peripheral cells by the aromatase cytochrome P450[1]. However, in our case, she did not have estrogenic manifestations.

Radiological studies are helpful in the diagnosis of SLCTs. However, it has no well-known characteristics[1]. Ultrasound scan features of the tumour with good colour Doppler and clinical manifestations support the diagnosis. In this case, her

ultrasonography revealed a well-defined lobulated solid cystic pelvic mass without blood flow. The Doppler findings in Sertoli-Leydig cell tumors can exhibit significant variability. These tumors may present a spectrum of blood flow patterns, ranging from limited vascularity to marked abundance within the tumour[8].

Therefore, we planned to manage as postmenopausal women with ovarian mass[9]. Serum Cancer Antigen- 125 (CA-125) level can be elevated in sex cord ovarian tumours, and preoperative serum CA-125 determination has a clinical value in predicting the curability of the disease[10]. Our patient had a normal level of (CA-125) with a low-Risk Malignant Index.

Surgery is the gold standard treatment for SLCTs, and it is based on age, fertility requirement, clinical stage, and tumour differentiation degree[10]. Total hysterectomy and bilateral salpingo-oophorectomy or cytoreductive surgery are recommended in women without fertility wishes. The necessity of pelvic lymphadenectomy is controversial. Lymph node involvement in SLCTs of the ovary is extremely rare. Therefore, routine pelvic and para-aortic lymphadenectomy could be safely omitted for SLCTs. In this case, since she already underwent a total Abdominal Hysterectomy in the past, bilateral salpingo-oophorectomy with complete excision of the left ovarian mass was performed.

The prognosis of SLCT depends on several factors, such as tumour stage, differentiation, and postoperative chemotherapy[1]. The good prognostic factors are low grade, absence of heterologous elements, and poor prognostic factors are postoperative chemotherapy, radiotherapy, or a combination of both. The overall 5-year survival rate for moderately differentiated SLCT is 80%[11]. A fifth of all SLCT relapses and most recurrences appear within three years. The reported relapse rates of 5–8% for stage IA tumours and approximately 30% for stage IC, with a mortality rate of 70% in relapse cases[11]. Risk factors for recurrence include advanced FIGO stage, poor differentiation, heterologous elements, and a retiform pattern.

In this case, we are approached as a postmenopausal woman with an ovarian cyst. She did not have significant hyperandrogenic features except hirsutism or hyperestrogenic features. Since her RMI is low and suspicion of SLCT is minimal due to her age, clinical and biochemical features. Laparotomy was performed, and bilateral salpingo-oophorectomy with complete excision of the left ovarian mass. Since her prognosis factors are good

postoperative follow-up was planned and avoid adjuvant chemo or radiotherapy.

Conclusion

The malignant prevalence of ovarian mass in a postmenopausal woman is high. Therefore, attentively investigative an ovarian mass in a postmenopausal woman in the view of malignancy is of paramount importance. Systematic clinical evaluation and management guided by the Risk Malignancy Index (RMI), are suitable and give optimal outcome for this age group, particularly in the absence of overt clinical manifestations. However, individualized clinical evaluation and pertinent biochemical assays remain imperative for the diagnosis and formulation of a management plan, especially in the case of a rare ovarian tumor. This is because RMI in isolation, does not suffice as the sole determinant for postmenopausal ovarian mass management.

SLCTs are uncommon tumours in postmenopausal women with various clinical presentations. Hence, the patient specific clinical assessment will facilitate early and possible diagnosis, and enabling timely interventions, collectively contributing significantly to the improvement of the prognosis.

Key words

Sertoli-Leydig cell tumours, primary ovarian neoplasm, tumours of postmenopausal women, Low Risk Malignant Index.

Conflict of interest statement

The authors declare that there is no conflict of interest.

Funding

This is a self-funded study supported by the corresponding author.

References

1) Xu Q, Zou Y, Zhang XF. Sertoli–Leydig cell tumors of ovary A case series. *Medicine (United States)* 2018; *97*(42). https://doi.org/10.1097/MD.0000000000012865
2) Berek JS, Renz M, Kehoe S, Kumar L, Friedlander M. Cancer of the ovary, fallopian tube, and peritoneum: 2021 update. *International Journal of Gynaecology and Obstetrics: The Official Organ of the International Federation of Gynaecology and Obstetrics* 2021; *155 Suppl 1*: 61–85. doi.org/10.1002/IJGO.13878
3) Gautam P, Rao M, Gothwal M, Garg P, Bhattacharya S. Sertoli-leydig cell tumor of ovary: A rare

case report with heterologous elements and focal marked anaplasia. *International Journal of Applied and Basic Medical Research* 2019; *9*(1): 62. https://doi.org/10.4103/IJABMR.IJABMR_84_18

4) Hillman RT, Gershenson D. Sex cord-stromal tumors of the ovary: road map for progress. *International Journal of Gynecological Cancer: Official Journal of the International Gynecological Cancer Society* 2021; *31*(2):169–170. https://doi.org/10.1136/IJGC-2020-002329
5) Tsuzuki Y, Kikuchi I, Nojima M, Yoshida K, Hashizume A, Tomita S. A Case Report: Ovarian Sertoli-Leydig Cell Tumor with Hyper-estrogenism and Endometrial Hyperplasia in a Postmenopausal Woman. 2017; *8*: 117906601769523. https://doi.org/10.1177/1179066017695239
6) Melero Cortés LM, Martínez Maestre MÁ, Vieites Pérez-Quintela MB, Gambadauro P. Ovarian Sertoli-Leydig cell tumours: How typical is their typical presentation? *Journal of Obstetrics and Gynaecology : The Journal of the Institute of Obstetrics and Gynaecology* 2017; *37*(5): 655–659. https://doi.org/10.1080/01443615.2017.1291590
7) de Kroon RWPM, den Heijer M, Heijboer AC. Is idiopathic hirsutism idiopathic? *Clinica Chimica Acta*, 2022; *531*: 17–24. https://doi.org/10.1016/j.cca.2022.03.011
8) Demidov VN, Lipatenkova J, Vikhareva O, van Holsbeke C, Timmerman D, Valentin, L. Imaging of gynecological disease (2): Clinical and ultrasound characteristics of Sertoli cell tumors, Sertoli-Leydig cell tumors and Leydig cell tumors. *Ultrasound in Obstetrics and Gynecology* 2008; *31*(1): 85–91. https://doi.org/10.1002/UOG.5227
9) *Ovarian Cysts in Postmenopausal Women (Green-top Guideline No. 34) | RCOG*. (n.d.). Retrieved August 17, 2023, from https://www.rcog.org.uk/guidance/browse-all-guidance/green-top-guidelines/ovarian-cysts-in-postmenopausal-women-green-top-guideline-no-34/
10) Shrestha S, Homagain S, Bhatta S, Tiwari SB, Rijal R, Aryal R, Sharma N, Paudyal P, Katuwal N,

Rawal SJ. Pure Sertoli cell tumor of the ovary: A case report. *Clinical Case Reports* 2022; *10*(5): e05892. https://doi.org/10.1002/CCR3.5892

11) Seidler SJ, Huber A, Nef J, Huber DE. *Sertoli-Leydig Cell Ovarian Tumors: Is Fertility or Endocrine-Sparing Surgery an Option upon Relapse?* 2020. https://doi.org/10.1159/000508532