




CASE REPORT

Pure Sertoli cell tumor of the ovary: A case report

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Abstract

Pure Sertoli cell tumors are an uncommon variant of rare ovarian Sertoli-Leydig cell tumors. Due to nonspecific clinical and imaging features, diagnosis is often made after histopathological examination. The prognosis is excellent as most are detected in the early stages and surgical resection is often curative in most cases.

KEYWORDS

ovarian tumor, Sertoli cell tumor, Sertoli-Leydig cell tumor

1 | INTRODUCTION

Sertoli-Leydig cell tumors (SLCTs) are a rare and heterogeneous group of ovarian neoplasms which belong to the sex cord-stromal category of tumors that account for <0.5% of primary ovarian neoplasms.¹ Sertoli-Leydig cell tumors are predominantly (90%) reported in the reproductive age group and often present with virilization.² Pure Sertoli cell tumors (SCTs) are even less common than SLCTs and account for only approximately 4% of the tumors in the category of Sertoli-stromal cell tumors of the ovary, which themselves as mentioned are overall uncommon.³

Here, we report a case of a 46-year-old woman with a pure SCT of the ovary who presented with recurrent abdominal pain.

2 | CASE REPORT

A 46-year-old P₄L₄ regularly menstruating woman presented with complaints of dull-aching recurrent pain in

the suprapubic region for one and half years and swelling of lower limbs for 1 month. The abdominal pain was severe during menses and associated with the passage of clots. There was no history of fever, per vaginal discharge, weight loss, abdominal distension, or loss of appetite. She underwent a mini-laparotomy 16 years back. There was no family/personal history of malignancy. Past medical history was unremarkable.

On examination, the abdomen was soft with mild tenderness present over hypogastrium and bilateral pitting edema in the lower limbs. A solid to cystic mass of approximately 10 cm × 8 cm with irregular surface and restricted mobility was found in the left adnexa during the per vaginal examination. Examination of all other systems was unremarkable.

Ultrasonography of the abdomen and pelvis revealed a well-defined lobulated solid cystic mass of 9.4 cm × 6.9 cm × 5.9 cm with multiple septations and mild vascularity in the left adnexa, which could not be distinguished from the left ovary. All the tumor markers including alpha-fetoprotein (AFP), carcinoembryonic

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antigen (CEA), Cancer Antigen- 125 (CA-125), beta-human chorionic gonadotropin (β -hCG), and inhibin were in a normal range. A contrast-enhanced computed tomography (CECT) scan of the abdomen/pelvis showed a large well-defined soft tissue density measuring 52 mm \times 93 mm \times 56 mm with cystic components and mild post-contrast enhancement in the left adnexa without any obviously enlarged lymph nodes in the pelvic/para-aortic regions and indistinguishable from the left ovary; suggestive of an ovarian neoplasm as shown in Figure 1A,B.

The patient underwent staging laparotomy with a total abdominal hysterectomy and bilateral salpingo-oophorectomy with a preoperative diagnosis of left ovarian neoplasm likely ovarian fibroma. Per-operatively, there was a smooth solid cystic ovarian mass of 8 cm \times 10 cm without any surface deposits and papillary projections along with a symmetrically enlarged uterus as shown in Figure 2. The specimen was sent for histopathological examination (HPE) along with an infracolic omental biopsy. Cytopathology of peritoneal washing showed no malignant cells. Histopathological examination of the excised ovary as shown in Figure 3, showed the tumor cells arranged in diffuse sheets and islands of elongated to hollow tubules separated by thin collagenous septa. These tubules were lined by cuboidal cells having eosinophilic cytoplasm, round to oval nuclei, fine chromatin, and inconspicuous nucleoli. Leydig cell components were not seen even on extensive sampling of the mass. Mitotic figures and cellular atypia were not seen. Lymphovascular and perineural invasions and capsular involvement were not seen. Omentum showed no tumor deposits. American Joint Committee on Cancer (AJCC) staging was pT1aNx

and the International Federation of Gynecology and Obstetrics (FIGO) staging was IA. Adenomyosis, chronic cervicitis with a nabothian cyst, and right paratubal cyst were additional pathological findings. The right ovary was normal.

The postoperative period was uneventful. As the patient had stage Ia ovarian tumor, she did not receive any adjuvant chemotherapeutic agents. She is on regular follow-up and is doing well 8 months after surgery.

3 | DISCUSSION

Ovarian SLCT can occur in any age group, however, is more often encountered in young women aged 25–28 years and <10% of ovarian SLCT occurs before menarche or after menopause.^{1,4}

Tumors arising from Leydig and Sertoli cells produce testosterone, are rarely estrogenic, and may present with hirsutism, acne, deepening of the voice, clitoral hypertrophy, amenorrhea, or irregular menstrual cycle, and sometimes along with pelvic masses.^{4,6} Although virilization is the most common manifestation, it is not obligatory, and in many cases, secondary amenorrhea is the only symptom, leading to an intensive search for the source of the disorder.⁷ Moreover, ovarian SLCTs can coexist with other types of ovarian tumors, making them more difficult to diagnose due to more complicated clinical symptoms, and the diagnosis of SLCTs before surgery is thus often very difficult. Our patient presented with abdominal pain, the most common presenting symptom of a Sertoli-Leydig cell ovarian tumor. There was not any androgenic feature however she had a heavy menstrual bleed in the form of

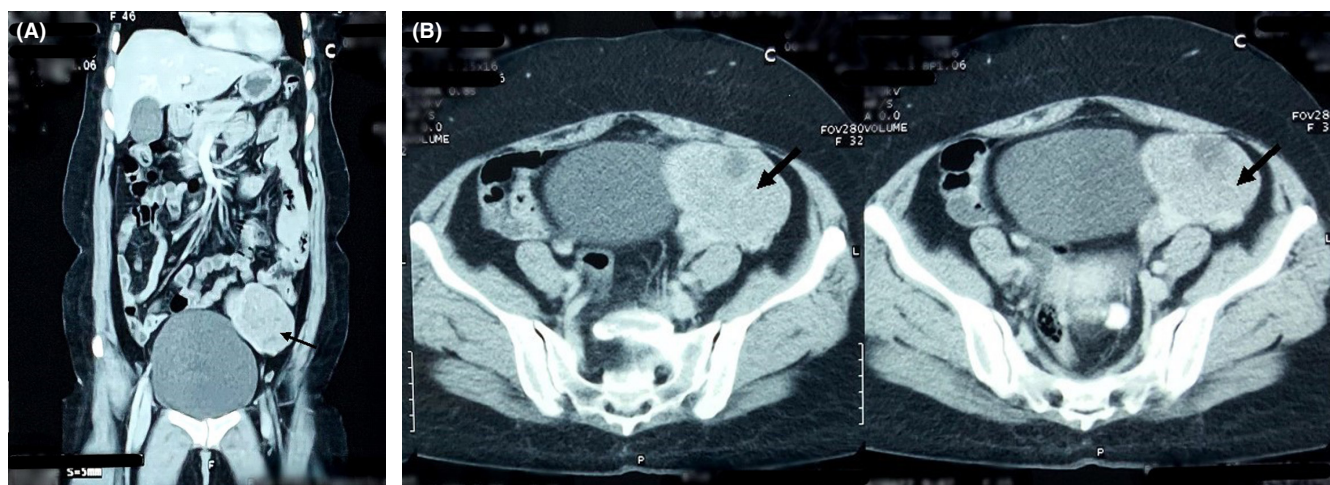


FIGURE 1 (A) Left adnexal mass in 46-year-old. Coronal image of contrast-enhanced computed tomography (CECT) of abdomen shows a heterogeneously enhancing left solid-cystic adnexal lesion with multiple areas of low attenuation (black arrow) along with symmetrically enlarged uterus. (B) Left adnexal mass in 46-year-old. Axial image of contrast-enhanced computed tomography (CECT) of abdomen shows enhancing solid-cystic left adnexal lesion with few areas of low attenuation (black arrows)

FIGURE 2 (A and B) Gross morphological feature of Total Abdominal Hysterectomy and Bilateral Salpingo-oophorectomy (TAH-BSO) specimen showing an irregular mass in the left ovary without areas of hemorrhage and papillary projections

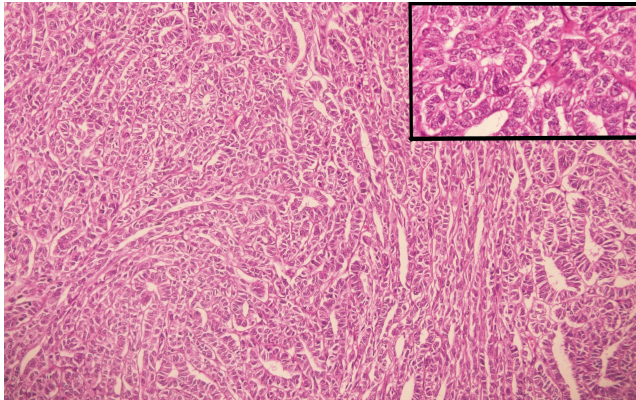
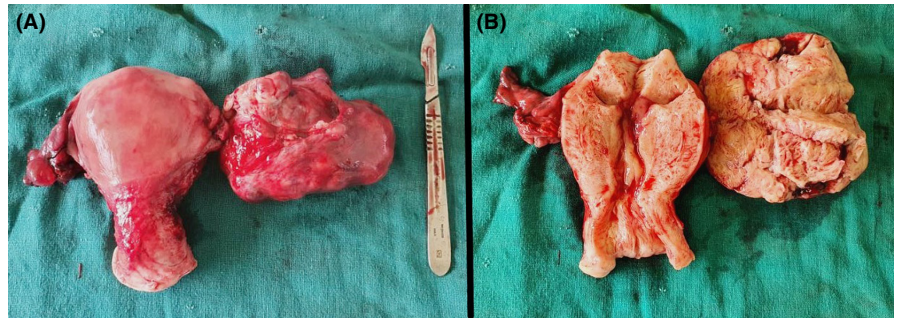


FIGURE 3 Hematoxylin and Eosin (H&E) stain from a cut section of the left ovary shows tumor cells arranged in solid to hollow tubules lined by cuboidal to columnar cells with eosinophilic cytoplasm with mild nuclear pleomorphism (inset, 400 \times)

passage of clots which could be an estrogenic feature of pure SCT. In addition, the dysmenorrhoea could be explained by the presence of adenomyosis as well as by venous congestion due to obstruction of blood flow which also explains the edematous legs.

Serum CA-125 level can be elevated in sex cord ovarian tumors including pure SCT and studies have shown that preoperative serum CA-125 determination has a clinical value in predicting the curability and stage of the disease.⁹ Our patient had normal CA-125 levels as well as all other tumor markers.

Sertoli-Leydig cell tumors are typically unilateral (only 1.5% occur bilaterally) solid tumors, mostly confined to the ovary at the time of diagnosis.⁹ Histologically, SLCTs are characterized by the proliferation of Sertoli and Leydig cells in varying proportions and are classified as well-differentiated, intermediate differentiation, and undifferentiated.¹⁰ Pure Sertoli cell tumors as in this case are rare. By definition, pure SCTs lack Leydig cells in the stroma. Microscopic examination shows cuboidal/columnar cell lined round/elongated tubules and usually without atypical features or mitotic activity. The microscopic

pattern of SCT can however be variable, ranging from well-differentiated to poorly differentiated form.⁸ The distinction of SCT from well-differentiated SLCT is based simply on the finding of more than rare cells consistent with Leydig cells in SLCTs. Two morphologic features of SLCTs, a retiform pattern and heterologous elements, strongly favor SLCT rather than SCT.¹¹ Inhibin is considered a sensitive and relatively specific marker for sex cord-stromal tumors and has been reported to be positive in almost all granulosa cell tumors, SCTs, and SLCTs of the ovary.¹² As SCTs usually have a distinctive tubular pattern that facilitates the diagnosis, immunohistochemical analysis is used for the differential diagnosis. As the distinct histological pattern was also present in our case, immunohistochemistry was not performed.

In the ultrasound, they are echogenic solid masses and on Computed Tomography (CT) and Magnetic Resonance Imaging (MRI), they appear as encapsulated solid enhancing masses.¹³ SLCTs should be differentiated from such ovarian neoplasms as granulosa cell tumors, fibrothecoma, and sclerosing stromal tumors. Granulosa cell tumors are usually estrogen tumors and rarely androgenic tumors. The majority of patients with SLCTs (81.8%–100%) have stage I cancers according to the FIGO. So did our patient (stage IA).^{7,10,14}

At present, there is no standard treatment for ovarian SLCT, surgery is still the first treatment option. The choice of operation method is based on age, fertility requirement, clinical stage, tumor size, and differentiation degree. Patients with well-differentiated stage Ia/Ib tumors could undertake unilateral salpingo-oophorectomy with or without exploration of the contralateral ovary. Patients with intermediate or poorly differentiated stage Ic (rupture of capsule) tumors could undergo unilateral salpingo-oophorectomy plus standard staging surgery (omentectomy, appendectomy, and pelvic lymphadenectomy).⁷ As our patient had completed family and was in perimenopausal period total abdominal hysterectomy and bilateral salpingo-oophorectomy (TAH-BSO) was performed for the post-operative diagnosis of unilateral solid ovarian tumor, likely ovarian fibroma, as most of the

tumor markers sent were normal. As lymph node metastasis in ovarian sex cord-stromal tumors is rare, lymphadenectomy may be omitted when staging surgery is performed. Avoidance of tumor spillage is critical since it has been associated with an increased risk of relapse.¹⁵ As radiological findings showed no enlarged lymph nodes in our case, lymph node dissection was not done.

The good prognostic factors are low grade, low stage, and absence of heterologous elements.¹ The prognosis of SLCTs is generally favorable with a much better prognosis than ovarian epithelial carcinomas as most SLCTs are detected in the early stages and have a favorable outcome with conservative surgeries, given the young age of the patients. In a study of ovarian SLCT, all 40 cases were at stage I, with an average follow-up period of 70 months, and no patients died from widespread tumors or metastasis.⁷ The overall 5-year survival rate was reported as 100% for well-differentiated and a collective 80% for moderately and poorly differentiated, 95% for stage I, and nearly none for stages III and IV SCLTs.¹

Chemotherapy is usually used in the ovarian SLCT that has advanced clinical stage, middle or low-differentiation, active karyokinesis or/and heterogenic component; and the chemotherapy protocol (bleomycin, etoposide, and cisplatin) is often applied in ovarian SLCT.^{17,18} The optimal chemotherapeutic regimen remains unclear and moreover, a study found that there was no significant difference in the rate of disease-free survival (DFS) between the group receiving chemotherapy and the group without chemotherapy ($p > 0.05$).⁸ As SLCT harbors hormone receptors, and consequently, hormonal replacement therapy is contraindicated.¹⁹

4 | CONCLUSION

Pure SLT is rare and thus SLCT poses a diagnostic challenge owing to its rarity. Early stage SLCTs have a favorable prognosis. An accurate diagnosis and staging based on clinical and pathological features have important therapeutic implications.

AUTHOR CONTRIBUTIONS

Pooja Paudyal (PP), Neeta Katwal (NK), Suniti Rawal (SR) = Study concept, Data collection, and surgical therapy for the patient. Suraj Shrestha (SS), Sushan Homagain (SH), Suraj Bhatta (SB), Roshan Aryal (RA), and Rishikesh Rijal (RR) = Writing—original draft preparation. Suraj Shrestha (SS), Sushan Homagain (SH), Suraj Bhatta (SB), Roshan Aryal (RA), Sansar Babu Tiwari (SBT), and Rishikesh Rijal (RR) = Editing and writing. PP, NK, and SR = senior author and manuscript reviewer. All the authors read and approved the manuscript.

CONFLICT OF INTEREST

None to declare.

DATA AVAILABILITY STATEMENT


All the necessary data and materials are within the manuscript.

CONSENT

Written informed consent was obtained from the patients' father for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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REFERENCES

1. Young RH, Scully RE. Ovarian Sertoli-Leydig cell tumors. A clinicopathological analysis of 207 cases. *Am J Surg Pathol*. 1985;9(8):543-569. doi:10.1097/00000478-198508000-00001
2. Zaloudek C, Norris HJ. Sertoli-Leydig tumors of the ovary. A clinicopathologic study of 64 intermediate and poorly differentiated neoplasms. *Am J Surg Pathol*. 1984;8(6):405-418. doi:10.1097/00000478-198406000-00001
3. Scully RE, Young RH, Clement PB. Tumors of the ovary, maldeveloped gonads, fallopian tube, and broad ligament. *Int J Gynecol Pathol*. 1999;18(3):288. doi:10.1097/00004347-199907000-00019
4. Caringella A, Loizzi V, Resta L, Ferreri R, Loverro G. A case of Sertoli-Leydig cell tumor in a postmenopausal woman. *Int J Gynecol Cancer*. 2006;16(1):435-438. doi:10.1111/j.1525-1438.2006.00203.x
5. Levin G, Zigron R, Haj-Yahya R, Matan LS, Rottenstreich A. Granulosa cell tumor of ovary: a systematic review of recent evidence. *Eur J Obstet Gynecol Reprod Biol*. 2018;225:57-61. doi:10.1016/j.ejogrb.2018.04.002
6. Chen J, Liu Y, Zhang Y, Wang Y, Chen X, Wang Z. Imaging, clinical, and pathologic findings of Sertoli-leydig cell tumors. *Sci Prog*. 2021;104(2):368504211009668. doi:10.1177/00368504211009668
7. Sigismondi C, Gadducci A, Lorusso D, et al. Ovarian Sertoli-Leydig cell tumors. A retrospective MITO study. *Gynecol Oncol*. 2012;125(3):673-676. doi:10.1016/j.ygyno.2012.03.024
8. Gui T, Cao D, Shen K, et al. A clinicopathological analysis of 40 cases of ovarian Sertoli-Leydig cell tumors. *Gynecol Oncol*. 2012;127(2):384-389. doi:10.1016/j.ygyno.2012.07.114
9. Al-Agha OM, Tahmasebi FC, Nicastrì AD. A 67-year-old woman with abdominal distention, vaginal bleeding, and elevated CA 125 level. Pure Sertoli cell tumor of the ovary with differentiation varying from well-differentiated tubules, to intermediate foci, to sarcomatoid spindle cell areas. *Arch Pathol Lab Med*. 2006;130(5):e70-e73. doi:10.5858/2006-130-e70-AYWWAD
10. Hanby AM, Walker C, Tavassoli FA, Devilee P. Pathology and genetics: Tumours of the breast and female genital organs. WHO

- classification of tumours series – volume IV. Lyon, France: IARC press. *Breast Cancer Res.* 2004;6(3):250. doi:[10.1186/bcr788](https://doi.org/10.1186/bcr788)
11. Gouy S, Arfi A, Maulard A, et al. Results from a monocentric long-term analysis of 23 patients with Ovarian Sertoli-Leydig cell tumors. *Oncologist.* 2019;24(5):702-709. doi:[10.1634/theoncologist.2017-0632](https://doi.org/10.1634/theoncologist.2017-0632)
 12. Young RH, Scully RE. Ovarian Sertoli-Leydig cell tumors with a retiform pattern: a problem in histopathologic diagnosis. A report of 25 cases. *Am J Surg Pathol.* 1983;7(8):755-771.
 13. Deavers MT, Malpica A, Liu J, Broaddus R, Silva EG. Ovarian sex cord-stromal tumors: an immunohistochemical study including a comparison of calretinin and inhibin. *Mod Pathol.* 2003;16(6):584-590. doi:[10.1097/01.MP.0000073133.79591.A1](https://doi.org/10.1097/01.MP.0000073133.79591.A1)
 14. Zung A, Shoham Z, Open M, Altman Y, Dgani R, Zadik Z. Sertoli cell tumor causing precocious puberty in a girl with Peutz-Jeghers syndrome. *Gynecol Oncol.* 1998;70(3):421-424. doi:[10.1006/gyno.1998.5063](https://doi.org/10.1006/gyno.1998.5063)
 15. Akman L, Ertas IE, Gokcu M, et al. Ovarian sertoli-leydig cell tumors: a multicenter long-term clinicopathological analysis of 27 patients. *J Cancer Res Ther.* 2016;12(1):290-294. doi:[10.4103/0973-1482.158037](https://doi.org/10.4103/0973-1482.158037)
 16. Melero Cortés LM, Martínez Maestre MÁ, Vieites Pérez-Quintela MB, Gambadauro P. Ovarian Sertoli-Leydig cell tumours: how typical is their typical presentation? *J Obstet Gynaecol.* 2017;37(5):655-659. doi:[10.1080/01443615.2017.1291590](https://doi.org/10.1080/01443615.2017.1291590)
 17. Sachdeva P, Arora R, Dubey C, Sukhija A, Daga M, Singh DK. Sertoli-Leydig cell tumor: a rare ovarian neoplasm. Case report and review of literature. *Gynecol Endocrinol.* 2008;24(4):230-234. doi:[10.1080/09513590801953465](https://doi.org/10.1080/09513590801953465)
 18. Huang LJ, Shi LY, Duan J. Clinicopathological analysis of ovarian sertoli-leydig cell tumor with postmenopausal vaginal bleeding as the first symptom: a case report. *Medicine.* 2021;100(13):e24922. doi:[10.1097/MD.00000000000024922](https://doi.org/10.1097/MD.00000000000024922)
 19. Rousset-Jablonski C, Selle F, Adda-Herzog E, et al. Fertility preservation, contraception and menopause hormone therapy in women treated for rare ovarian tumours: guidelines from the French national network dedicated to rare gynaecological cancers. *Eur J Cancer.* 2019;116:35-44. doi:[10.1016/j.ejca.2019.04.018](https://doi.org/10.1016/j.ejca.2019.04.018)

How to cite this article: Shrestha S, Homagain S, Bhatta S, et al. Pure Sertoli cell tumor of the ovary: A case report. *Clin Case Rep.* 2022;10:e05892. doi:[10.1002/ccr3.5892](https://doi.org/10.1002/ccr3.5892)