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Sertoli Leydig Cell tumor of Ovary: A Rare Entity

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Abstract

Sertoli-Leydig cell tumors (SLCTs) of ovary represents <0.2% of ovarian cancer. Seen at early stage confined to one ovary in reproductive age group with good prognosis fertility-sparing surgery is an accepted option. Need for adjuvant chemotherapy and relapse depends on risk factors like advanced stage, poor differentiation, retiform patterns and presence of heterologous elements. In this study, clinical pathological characteristics, management and recurrence of ovarian SLCTs has been noted which might aid to improvise on management of SLCTs. Materials And Methods: A retrospective analysis of 8 patients with SLCTs were done. Clinical features, tumor markers, imaging findings, pathological features, treatment modalities, recurrence and survival were noted. A total of 8 cases were noted. Median age was 24 years, 37.5% patients were postmenopausal. 5 underwent unilateral salpingo-oophorectomy with infra colic omentectomy. 2 patients had pelvic peritoneal disease underwent debulking of the same. 2 patients had Stage Ia disease, 4 had Stage 1c3 and 2 had Stage 2b. 5 patients received adjuvant chemotherapy with BEP or Paclitaxel and Carboplatin. 2 patients were kept under surveillance. 1 patient had recurrence. With 36 months follow up, 7/8 patients are alive and disease free. SLCTs are a very rare entity of tumors were fertility sparing surgery followed by adjuvant chemotherapy is essential in high risk and advanced cases. A registry which documents these kind of rare tumors would be extremely useful for management and surveillance of these patients.

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Key Words
Sertoli, cell tumor, ovary

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INTRODUCTION

Of the Ovarian Cancers, Sertoli-Leydig cell tumors (SLCTs) belonging to Sex cord stromal tumors, form one of the rarest tumors representing less than 0.2%[1]. Though SLCTs can occur over a very wide age range they are most commonly seen in second and third decade of life[2]. SLCTs commonly present with symptoms and signs of androgen excess like hirsutism, acne, and oligomenorrhea-amenorrhea in 50% of cases and the rest can be non-functional tumours or oestrogen-secreting tumours occasionally resulting in endometrial Carcinoma[3].

Most SLCT are usually seen at early stage confined to one ovary in reproductive age group with good prognosis fertility-sparing surgery is an accepted option[4]. Need for adjuvant chemotherapy and chances of relapse increases with high risk factors like advanced stage at presentation with poor differentiation with retiform patterns and the presence of heterologous elements[1]. SLCTs recur in 20% of all SLCT [5]. Because of the rarity of SLCTs management protocol guidelines remain uncertain.

In this study, clinical pathological characteristics, management and recurrence of ovarian SLCTs has been noted which might aid to improvise on management of SLCTs.

MATERIALS AND METHODS

A retrospective analysis of 8 patients with histologically proven diagnosis of SLCTs were done through reviewing of the medical records from Jan 2019-Dec 2022 was done at a Regional Cancer Centre. Clinical features, tumor markers, imaging findings, pathological features, treatment modalities, recurrence and survival were noted.

RESULTS AND DISCUSSION

A total of 8 cases with histological diagnosis of SLCTs were documented from 2019-2021. With median age being 24 years, 3/8 (37.5%) patients were postmenopausal and rest 5/8 (62.5%) were <25 years of age. Commonest complaint was abdominal pain in 5 patients followed by irregular cycles in 2 and oligomenorrhoea in one patient. Only 1/8 patient had hoarseness of voice. All patients had contrast enhanced computerised tomography. All 8 patients had unilateral solid cystic tumor with size ranging from 924cms. Tumor markers Ca 125 ranged from 18- 101, one patient had Ca 19.9 of 1721 and another patient had AFP of 25899. Of 8 patients, 5 underwent unilateral salpingo-oophorectomy with infra colic omentectomy and 3/8 post menopausal women has total abdominal hysterectomy with infra colic omentectomy. Two patients had pelvic peritoneal disease underwent debulking of the same. Histopathologically 2 patients had heterologous elements and 1 patient had retiform pattern. Immunohistochemistry showed Calret and Inhibin positivity in 5/8 patients. Two patients had Stage Ia disease, 4 patients had Stage 1c3 and 2 patients had Stage 2b disease. 5 patients received adjuvant chemotherapy of which 2/5 received BEP regimen and 3/5 received Paclitaxel and Carboplatin. One patient who had Stage 2b disease defaulted for adjuvant chemotherapy reported after 4 months with disease in pelvis who later received BEP regimen. 2 patients with 1a disease were kept under surveillance. Only 1/8 patient who had Stage 1a well differentiated disease who was kept on surveillance in initial treatment had recurrence thrice for which secondary and tertiary cyto reduction was done along with chemotherapy BEP, Paclitaxel carboplatin and VAC regimens. With an average of 36 months follow up, 7/8 patients are alive and disease free and one patient lost for follow up.

The age of the patient ranged between 19-65 years with a median of 24 years which was similar in past studies[6]. Most of our patients presented with abdominal pain as presenting complaint and only in one patient hirsutism and oligomenorrhoea was noted which was different from other studies which had many androgen/ estrogen excessive symptoms nearing upto 50% of cases[1]. All 8 cases were multiloculated solid cystic on CECT which was similar to other studies[7]. Surgery still remains the main stay of management in SLCTs as most of them present in early stage. Also, it is noted that most of the cases are unilateral and occur in younger and reproductive age group, fertility sparing surgery is the most accepted optional surgical management[8]. 5/8 patients in reproductive age group underwent fertility sparing surgery in the form of unilateral salpingo-oophorectomy with infra colic omentectomy 3/8 post menopausal patient underwent total abdominal hysterectomy with infra colic omentectomy and pelvic peritonectomy. Recommendations suggest adjuvant chemotherapy from Stage Ic onwards with retiform pattern or heterologous component[9]. BEP and TC are the most common regimens used[6]. 5/8 patients received adjuvant chemotherapy including 4 patients with Stage Ic3 with retiform pattern, heterologous element and poorly differentiated as these patients with high risk factors have found to have higher recurrence as well as higher mortality rate[10] and 2 patients with Stage 1a disease was kept for follow up. 1 patient with stage IIb disease defaulted for 4 months and came back with progressive disease after 4 months, was managed with Paclitaxel and carboplatin. SLCTs with advanced stage or with poor differentiation are more likely to recur[11].Whereas in our series patient with Stage 1a disease with well

differentiation with no high risk factors who was on surveillance only developed recurrence in pelvis after 26 months and she was managed with paclitaxel and carboplatin initially after which she underwent secondary and tertiary cytoreductive surgeries followed by BEP and VAC regimen of chemotherapy. 7/8 patients are alive disease free till date and 1 patient is lost for follow up. Most of our patients has got married but no conception has been documented.

CONCLUSION

SLCTs are a very rare entity of tumors were fertility sparing surgery followed by adjuvant chemotherapy is essential in high risk and advanced cases. Very few case series are available which tell us about SLCTs, hence a registry which documents these kind of rare tumors would be extremely useful for management and surveillance of these patients.

REFERENCES

1. Young, R.H. and R.E. Scully, 1985. Ovarian sertoli - leydig cell tumors. . A clinicopathological analysis of 207 cases. Am. J. Surg. Pathol., 9: 543-569.
2. Colombo, N., G. Parma, V. Zanagnolo and A. Insinga, 2007. Management of ovarian stromal cell tumors. J. Clin. Oncol., 25: 2944-2951.
3. Seidler, S.J., A. Huber, J. Nef and D.E. Huber, 2020. Sertoli-leydig cell ovarian tumors: Is fertility or endocrine-sparing surgery an option upon relapse? Case Rep. Oncol., 13: 935-940.
4. Morice, P., D. Denschlag, A. Rodolakis, N. Reed, A. Schneider, V. Kesic and N. Colombo, 2011. Recommendations of the fertility task force of the European society of gynecologic oncology about the conservative management of ovarian malignant tumors. Int. J. Gyne Cancer, 21: 951-963.
5. Litta, P., C. Saccardi, L. Conte, A. Codroma, S. Angioni and R. Mioni, 2013. Sertoli–leydig cell tumors: Current status of surgical management: Literature review and proposal of treatment. Gyne End., 29: 412-417.
6. Wang, G., R. Zhang, C. Li and A. Chen, 2021. Characteristics and outcomes analysis of ovarian sertoli–leydig cell tumors (slcts): Analysis of 15 patients. J. Ovarian Res., Vol. 14, No. 1 .10.1186/s13048-021-00909-7.
7. Demidov, V.N., J. Lipatenkova, O. Vikhareva, C.V. Holsbeke, D. Timmerman and L. Valentin, 2007. Imaging of gynecological disease (2): Clinical and ultrasound characteristics of sertoli cell tumors, sertoli–leydig cell tumors and leydig cell tumors. Ultr Ob. amp Gyn., 31: 85-91.
8. Gui, T., D. Cao, K. Shen, J. Yang and Y. Zhang et al., 2012. A clinicopathological analysis of 40 cases of ovarian sertoli–leydig cell tumors. Gyne Oncol., 127: 384-389.
9. Ettinger, D.S., D.E. Wood, D.L. Aisner, W. Akerley and J.R. Bauman et al., 2023. Nccn guidelines® insights: Non–small cell lung cancer, version 2.2023. J. Nat. Compr. Cancer Network, 21: 340-350.
10. Gouy, S., A. Arfi, A. Maulard, P. Pautier and E. Bentivegna et al., 2018. Results from a monocentric long-term analysis of 23 patients with ovarian sertoli-leydig cell tumors. Oncologist, 24: 702-709.
11. Nef, J. and D.E. Huber, 2021. Ovarian sertoli–leydig cell tumours: A systematic review of relapsed cases. Eur. J. Obstet. amp Gyn. Reprod. Biol., 263: 261-274.