IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 21, Issue 4 Ser.6 (April. 2022), PP 58-61
www.iosrjournals.org

Sertoli-Leydig Cell Tumor of Ovary: An Incidental Diagnosis

Dr Rahul Mahawar[1*], Dr Silchang K. Marak[1], Dr Y. Indibor Singh[2]
[1]*PGT, Department of Radiation Oncology, Regional Institute of Medical Sciences (RIMS), Imphal, Manipur, India*
[2]*Professor, Department of Radiation Oncology, Regional Institute of Medical Sciences (RIMS), Imphal, Manipur,India*
[*]*Corresponding author*

Abstract
Sertoli-Leydig cell tumors are rare sex-cord stromal tumors of the ovary. Its incidence is less than 0.5% of all primary ovarian malignancies. They are generally benign and seen mostly in reproductive age group women. It can present with a wide variety of histological elements, which can complicate diagnosis and treatment. The paper reports a case of a 24-year-old primigravida women, with an incidental finding of ovarian mass during pregnancy. The patient underwent elective cesarean section with excision of the ovarian mass. The excised specimen was sent for histopathological analysis, which showed features consistent with Sertoli-Leydig cell tumor. The patient received three cycles of adjuvant chemotherapy and is currently under follow up.
Keywords: *Ovarian, Sertoli-Leydig cell, sex-cord stromal tumor*

Date of Submission: 02-04-2022 Date of Acceptance: 15-04-2022

I. Introduction

Sertoli-Leydig cell tumors (SLCTs) also known as androblastoma, are rare sex-cord stromal tumors of the ovary, with incidence of less than 0.5% of all primary ovarian malignancies.[1] The vast majority of SLCTs (90%) are diagnosed in the reproductive age group, with 51% occurring in the second and third decade of life.[2] They are generally unilateral, mostly confined to the ovary and approximately 90% are classified as stage 1 at the time of clinical diagnosis.[3] Approximately 80% of SLCTs are hormonally active with increased level of serum testosterone and androstenedione. Clinical sign and symptoms due to excess androgen production ranges from virilization, voice deepening, hirsutism, acne, clitoromegaly, breast atrophy, amenorrhea to male pattern baldness, and due to excess estrogen includes breast hypertrophy, precocious puberty, endometrial polyps or hyperplasia, postmenopausal bleeding and menometorrhagia.[4,5] Most common clinical feature seen in 50% of patients are abdominal mass, distention and pain, along with ascites.[6] Proper diagnosis and development of standard treatment regimen is quite challenging owing to varying histopathology and tumor differentiation.

II. Case Presentation

A 24-year-old primigravida woman in her third trimester attended the Department of Obstetrics & Gynecology, Regional Institute of Medical Sciences (RIMS) for routine antenatal ultrasound screening. During routine ultrasound screening, a space occupying lesion was incidentally found on the left ovary, along with small amount of ascites in the pouch of Douglas. The lesion was 1.0 cm × 0.8 cm × 2.0 cm in size. There is no history of abdominal pain, acne, hirsutism, voice deepening, decreased appetite or breathing difficulty. There is no family history of malignancy or similar illness in the past or any other co-morbidity.

The patient underwent elective low segment cesarean section (LSCS) along with left ovariectomy on 15/12/21 at RIMS, and the excised left ovariectomy specimen was sent for histopathological analysis (HPE).

On gross examination, the tumor was 1.7 cm × 1.0 cm × 3.0 cm in size, with a smooth and glistening external surface. Cut section shows multilocular cystic cavities containing serosanguinous fluid, with no solid component or signs of necrosis or hemorrhage. Microscopically, the tumor showed dual population cells intermixed with each other in different architecture (polygonal and tall columnar cells). No cellular atypia, heterogenous elements or increased in mitotic activity was seen. The HPE findings were consistent with Sertoli-Leydig cell tumor – well differentiated type (**Fig. A**). Immunohistochemistry (IHC) was positive for Inhibin, Calretinin and CD99 (**Fig. B & C**).

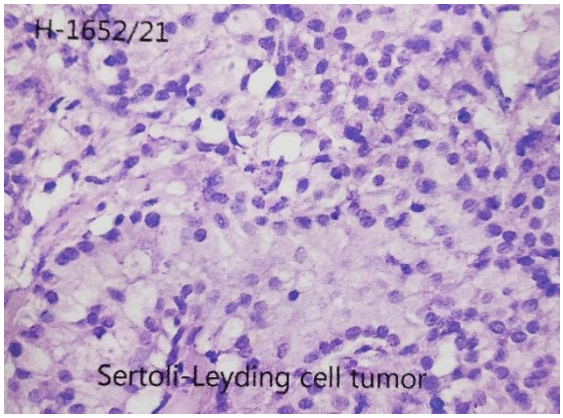

Fig A: HPE of the excised left ovariectomy specimen showing features consistent with Sertoli-Leydig cell tumor of the ovary

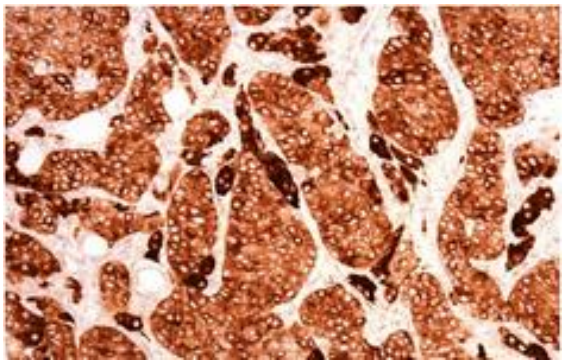

Fig B: IHC report showing Inhibin "positive"

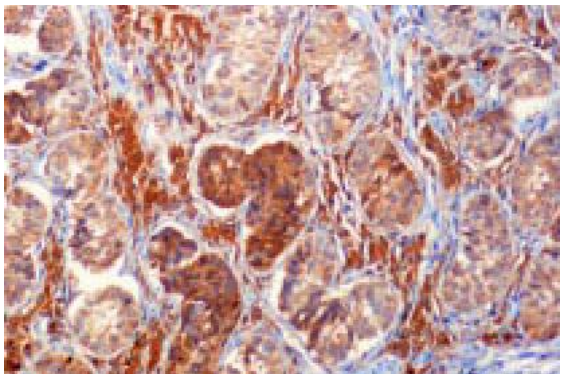

Fig C: IHC report showing Calretinin "positive"

After the confirmation of tissue diagnosis, the patient was transferred to the Department of Radiation Oncology, RIMS were thorough physical examination and metastatic workup was done. No lymph node or any mass were clinically palpable. The LSCS site looked healthy with no signs of inflammation. Routine baseline investigations (CBC, biochemistry & ECG) were done and were found to be uneventful. Chest X-ray showed no signs of metastasis.

Postoperative serum level of tumor markers (CA 125, CA 19-9, CA 15-3, CEA and beta HCG) was within normal range. Serum testosterone levels were not checked as there were no signs of androgen excess. Postoperative CECT of whole abdomen showed features suggestive of intramural fibroid with few areas of hemorrhage within the posterior wall of uterine body, with normal right ovary and residual left ovary appearance, and no cystic lesion. The patient received three cycles of adjuvant chemotherapy with BEP regimen [Injection Bleomycin (30U) on Day 1 & 8, Injection Etoposide (150mg) from Day 1 to 3, and Injection Cisplatin (30mg) from Day 1 to 5], three weekly cycles. The patient showed complete response with no evidence of residual disease and is under regular follow up.

III. Discussion

Ovarian sex-cord stromal tumors (SLCTs) constitute 1.2% of all primary ovarian tumors.[1] It is characterized by uncontrolled proliferation of Sertoli and Leydig cells to varying degree of differentiation in the ovary. The neoplastic Sertoli and Leydig cells exhibits varying degree of differentiation like well differentiated, moderately differentiated, or poorly differentiated, and with heterologous elements.[7]

Approximately 20% of patients have heterologous elements, with the most common being enteric type of mucinous epithelium. Degree of tumor differentiation is age linked, with patients exhibiting poorly differentiated SLCTs appears to be 10 years younger than patients with well differentiated SLCTs.[1,8]

It is commonly seen in young women between second and third decade (75%) of life, with average age of presentation being 25 years. Incidence prior to menarche or after menopause is less than 10%.[1] Clinical signs and symptoms is related either to hormone production (androgen or estrogen excess, or functionally inactive) or presence of mass occupying lesion.[1,8]

About two-third patients presents with androgen excess manifestations. Patients may also present with symptoms related to abdominal mass effect like abdominal/ pelvic mass or pain.[1,8] Acute abdominal pain requires prompt and immediate medical intervention.

Ovarian SCLT are the most common AFP-producing non-germ cell tumor of the ovary. Sonography is the best imaging for initial assessment of the mass, with transvaginal sonography being better than abdominal sonography. CT scan, MRI scan and PET scan cane be used for detection of extraovarian mass, distant metastasis, or for better characterization of ovarian SLCTs.

Ovarian SLCTs are usually unilateral, firm, lobulated, well-encapsulated solid mass. Microscopically, they are made up of uncontrolled proliferation of varying degrees of tubular differentiation, lined by Sertoli cells and intervening nests of Leydig cells. Well and moderately differentiated histological variant being most common, with poorly differentiated offering a diagnostic challenge. Immunohistochemically (IHC), all SLCTs stain positive for Inhibin, Calretinin and CD56, and negative for Epithelial membrane antigen (EMA).[9]

Differential diagnosis of ovarian SLCTs includes granulosa cell tumors, endometroid carcinoma, serous carcinoma, hepatoid carcinoma of the ovary and endodermal sinus tumor.[10] Thereby, making IHC markers important in the diagnosis of ovarian SLCTs.

The prognosis of ovarian SLCTs is usually favorable and depends on the tumor stage, grade, patient's age and postoperative chemotherapy, with well differentiated having an excellent prognosis. Metastatic disease is rare in well-differentiated type of SLCTs. As ovarian SLCTs are rare, its management is difficult as there are no standardized treatment guidelines.[11] Surgery is the treatment of choice of ovarian SLCTs.[10] Fertility sparing surgery with preservation of the uterus and contralateral ovary is recommended for stage 1 patients, who desire future childbearing.[1] For patients with advanced stage progressive disease, total hysterectomy with bilateral salpingo-oopherectomy with complete standard staging surgery is the treatment of choice.

Postoperative chemotherapy, radiotherapy or their combination is considered for patients with poor prognostic factors and high recurrence rate like moderate to poor tumor grading, advanced disease stage, presence of heterologous elements and tumor rupture.[1,12] The most commonly used regimen is BEP (Bleomycin, Etoposide & Cisplatin) or EP regimen. Other chemotherapy regimens that are used are CAP (Cisplatin, Adriamycin & Cyclophosphamide) and PVB (Cisplatin, Vinblastine & Bleomycin) regimen.[13]

IV. Conclusion

Sertoli-Leydig Cell Tumors of the ovary are rare sex-cord stromal tumors, seen mainly in reproductive age group women, during the first three decade of life. Early detection, appropriate initial surgery, along with adjuvant use of chemotherapy and radiotherapy offers the best prognosis and treatment option. Our patient was diagnosed as a case of Sertoli-Leydig cell tumor (SLCT) of the ovary after histopathological and IHC confirmation of the excised left ovariectomy specimen. The patient is currently under regular follow up.

References

[1]. Young RH, Scully RE. Ovarian Sertoli-Leydig cell tumors. A clinicopathological analysis of 207 cases. Am J Surg Pathol 1985; 9:543-69.
[2]. 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; 12:290-4.
[3]. Young RH, Scully RE. Sex cord-stromal, Sertoli cell, and other ovarian tumors. Blaustein's Pathology of Female Genital Tract, R. J. Kurman, ed., p. 929, Springer, New York, NY, USA, 5th edition, 2002.
[4]. Osborn RH, Yannone ME. Plasma androgens in the normal and androgenic female: A review. Obstet Gynecol Surv. 1971; 26:195-228.
[5]. Zhang HY, Zhu JE, Huang W, et al. Clinicopathologic features of ovarian Sertoli-Leydig cell tumors. Int J Clin Exp Pathol 2014; 7:6956-64.
[6]. Zanotti KM. The clinical manifestations and diagnosis of Sertoli-Leydig cell tumors of the ovary. CME J Gynecol Oncol 2002; 7:129-33.
[7]. Chen VW, Ruiz B, Killeen JL, et al. Pathology and classification of ovarian tumors. Cancer 2003; 97(10):2631-42.

[8]. Roth LM, Anderson MC, Govan ADT. Sertoli-Leydig cell tumors: a clinicopathologic study of 34 cases. Cancer 1981; 48(1):187-97.
[9]. McCluggage WG, Young RH. Immunohistochemistry as a diagnostic aid in the evaluation of ovarian tumors. Seminars in Diagnostic Pathology 2005; 22(1):3-32.
[10]. Weng CS, Chen MY, Wang TY, et al. Sertoli–Leydig cell tumors of the ovary: a Taiwanese gynecologic oncology group study. Taiwan J Obstet Gynecol 2013; 52: 66-70.
[11]. Bhat RA, Lim YK, Chia YN, et al. Sertoli-Leydig cell tumor of the ovary: analysis of a single institution database. J Obstet Gynaecol Res 2013; 39(1):305-10.
[12]. Edge SB, Compton CC. The American Joint Committee on Cancer: the 7th edition of the AJCC cancer staging manual and the future of TNM. Ann Surg Oncol 2010; 17:1471-4.
[13]. Roth BJ, Greist A, Kubilis PS, et al. Cisplatin-based combination chemotherapy for disseminated germ cell tumors: long-term follow-up. J Clin Oncol 1988; 6(8):1239-47.

Dr Rahul Mahawar, et. al. "Sertoli-Leydig Cell Tumor of Ovary: An Incidental Diagnosis." *IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)*, 21(04), 2022, pp. 58-61.