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Case Report

The unusual presentation of sertoli - leydig cell tumor: a case report

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ABSTRACT

Sertoli cell tumor is very rare variety of ovarian Sertoli-Leydig cell tumors. Due to nonspecific clinical and imaging features, diagnosis is often made after histopathological examination. Sertoli cell tumors usually presents with the features of virilization. The prognosis is excellent as most are detected in the early stages and surgical resection is often curative in most cases. Here we present a case of Sertoli Cell tumor, where no feature of virilization was present.

Keywords: Ovarian tumor, Sertoli cell tumor, Sertoli-leydig cell tumor

INTRODUCTION

Sertoli-Leydig cell tumors (SLCTs) are very rare and heterogeneous ovarian neoplasms which belong to the sex cord-stromal category that account for <0.5% of primary ovarian neoplasms.[1] These tumors are predominantly (90%) reported in the reproductive age group and often present with virilization.

Here, we report a case of a 30 years old married P3L1 female presented to the outdoor gynaecology department with complaints of abdominal heaviness with abdominal lump from 3 months, without any sign and symptoms of virilisation.

CASE REPORT

A 30-year-old married P3L1 female presented to the outdoor gynaecology department with chief complaints of abdominal heaviness with abdominal lump for last 3 months, which was gradually increasing in size, as well as associated with loss of appetite. Her bowel and bladder habits were normal. There was no history of sudden weight loss, menstrual irregularities, acne, excessive facial hair growth, voice changes. Her medical, surgical and family history were insignificant. On general examination, all parameters were found to be normal. On per abdominal examination, a well-defined, firm mass of approximately 20 weeks' size gravid uterus felt with smooth surface, regular margins, with side-to-side mobility, and non-tender. The lower limit of mass could not be reached confirming its origin from pelvis. On local examination there were no signs of virilisation seen. On per speculum examination cervix and vagina appears healthy. On bimanual examination uterus was anteverted, firm, mobile, normal size, non-tender, with B/L forniceal mass. Both masses were separately felt, freely mobile, firm in consistency, and non-tender (size- right sided approximately 13×10 cm, left sided approximately 6×6 cm).

Considering bilateral solid ovarian tumor her tumor markers along with basic preanesthetic investigation were sent. Tumor markers were found to be within normal range. Other investigations were also found to be normal. Further patient was referred for imaging. CT showed a large well-defined mass of approximately 15×9 cm with solid cystic areas noted in pelvis seems to be arising from right adnexa. On contrast enhancement, lesion showed more enhancement at periphery in comparison to central

part. Left ovary could not be visualized separately. No evidence of ascites and Para aortic lymphadenopathy was noted. Patient refused for further imaging and insisted for surgery and removal of whole tumor.

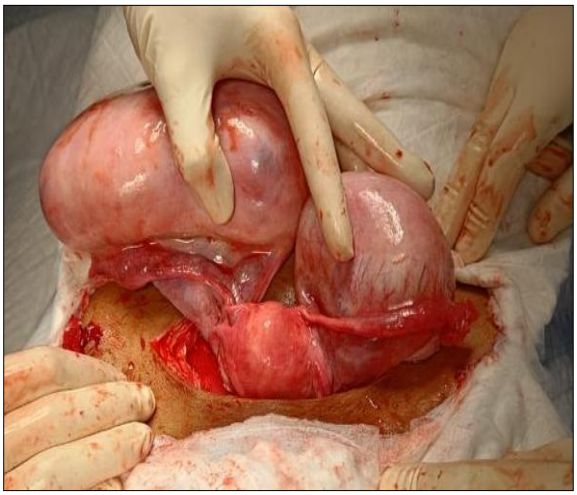

Figure 1: Per-operative picture of bilateral ovarian tumor with normal uterus and both tubes.

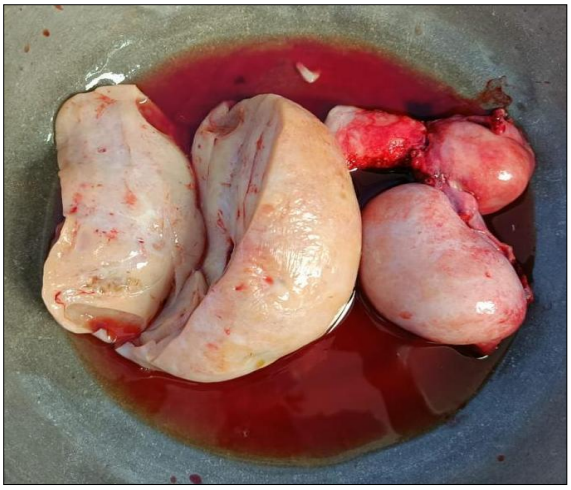

Figure 2: Gross specimen of Total Hysterectomy with bilateral salpingoophorectomy showing solid ovarian tumor.

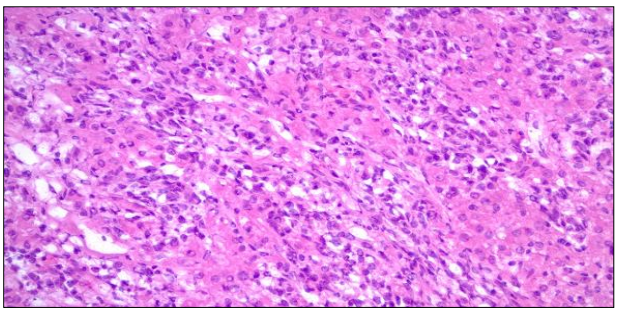

Figure 3: Microscopic picture suggestive of sertoli-leydig cell tumor.

Her exploratory laparotomy was done. Preoperatively approximately 50 CC peritoneal fluid drained out, which was sent for cytological examination in heparinized vial. Right ovary was replaced by approximately 15×10 cm solid grey white mass, and left ovary was also replaced by approximately 8×6 cm tumor of same appearance. Uterus was found normal, no signs carcinosis seen on parietal peritoneum, intestines, liver, no surgical spillage, (Figure 1) making the surgical stage: 1B. after discussing the risk and prognosis with patients' attendants her total hysterectomy and bilateral salpingoophorectomy was done and sample was sent for histopathological examination.

Patient's postoperative period was uneventful. She was started with hormone replacement therapy from postoperative day two. Abdominal stitches were removed on postoperative day 8 and patient was discharged satisfactorily from hospital with advice for regular follow up.

Gross examination of specimen showed ovarian masses with smooth outer surfaces. On cut section the masses were solid with few cystic spaces filled with clear fluid. The histopathology of tumor showed solid masses (Figure 2) comprise of poorly formed cords, nests and tubules of tumor cells which had hyperchromatic nuclei with moderate amount of cytoplasm (sertoli cells). In between these cells there were nests of polygonal cells with round nuclei and plenty of granular cytoplasm (leydig cells) (Figure 3). Uterus, cervix and both tubes were found normal histologically. So, the diagnosis of well differentiated Sertoli-Leydig cell tumor of ovary was made.

DISCUSSION

Less than 0.5% of all primary ovarian tumors are SLCTs, an extremely rare kind of ovarian tumor that falls under the category of sex cord stromal tumors.[1] The reproductive age group (second and third decades of life) is typically affected.[1-4] The presence of a mass-occupying lesion or hormonal production (mainly androgen, seldom estrogen) are important feature of clinical presentation.[1-3,5,6] Virilism, hirsutism, hoarseness of voice, clitoromegaly, oligomenorrhea, and amenorrhea are main signs of androgen excess.[7,8] Ovarian SLCTs can be diagnosed with imaging techniques. Sonography's great sensitivity, applicability, and affordability make it the preferred imaging modality for the preliminary evaluation of adnexal masses.[6,9] Sonographically, SLCTs seem solid most of the time and are usually unilateral tumors.[1,3,5,6,9] About 60% of all ovarian SLCTs include mixed (solid and cystic) components. Color Doppler provides additional malignant mass classification and assessment. Low resistance indices and moderate-to-rich ovarian vascular masses strongly point to malignant rather than benign lesions. To detect extra ovarian illness or metastases and to better characterize ovarian SLCTs, additional imaging modalities such CT, MRI, and PET scans can be employed. On CT or MRI, SLCTs appear as hypervascular

mixed solid-cystic lesions inside the adnexa. Preoperative tumor staging can be accomplished by cross-sectional imaging. Macroscopically, SLCTs are yellow-gray, substantial, firm, and well-encapsulated masses.[10] The surface of the cut portion shows different levels of greasy/fleshy consistency, fluid with a straw hue, necrosis, bleeding, and cystic areas divided by fibrous septae.

Under a microscope, SLCTs are composed of intervening Leydig cell nests and the unchecked growth of tubules bordered by Sertoli cells that have undergone differentiation.[10] Leydig cells have polygonal cells with clearly defined borders, central nuclei, noticeable nucleoli, and eosinophilic cytoplasm. They are generally found in clusters in the interstitial stroma. Sertoli cells usually form tubular structures that are lined with eosinophilic or vacuolated cytoplasm, oval black nuclei, well-bounded edges, and single or multiple layers of cuboidal-columnar cells. For ovarian SLCTs, surgical resection is the cornerstone of treatment.[4] For all individuals with well differentiated ovarian SLCTs, fertility sparing surgery (unilateral salpingo-oophorectomy) may therefore be considered. Postoperative chemotherapy is typically evaluated for patients with poor prognostic characteristics, such as moderate to poor tumor grading, advanced disease staging, high mitotic figures, tumor rupture, and existence of heterologous elements.[1-3] Grading and staging-two measures of tumor area and degree of differentiation-have a substantial correlation with the prognosis of ovarian SLCTs.[1] Long-term follow-up is highly encouraged in all patients.

CONCLUSION

SLCT is a rare neoplasm seen in young women with mean age of 25 years. Size may range from 5-25 cm. It is generally solid or solid cystic. Prognosis of SLCT depends on stage and degree of differentiation; but usually carries good prognosis. The 5-year survival rate is 70% to 90 %. Adjuvant chemotherapy is recommended in advanced stages, poorly differentiated tumor, retiform pattern and presence of heterologous elements.

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