Case Report

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Sertoli–Leydig Cell Tumor of Ovary an Incidental Finding: A Rare Case Report

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Abstract

Sertoli–Leydig cell tumor (SLCT) is a group of tumors composed of variable proportions of Sertoli cells, Leydig cells, and sometimes heterologous elements. Most tumors are unilateral, confined to the ovaries, and are seen during the second and third decades of life. These tumors are characterized by the presence of testicular structures that produce androgens. Hence, many patients have symptoms of virilization depending on the quantity of androgen production. We are presenting a case of incidental finding of ovarian tumor in a 54-year-old lady who underwent hysterectomy with bilateral salpingo oopherectomy for dysfunctional uterine bleeding. A diagnosis of SLCT of ovary was rendered for the same.

Key words: Androgen secreting neoplasm, Sertoli–Leydig cell tumor, Virilizing ovarian tumors

INTRODUCTION

Sertoli–Leydig cell tumor (SLCT) is a rare ovarian tumor that belongs to the group of sex-cord stromal tumors. These constitute <0.1% of ovarian tumors.[1,2] The tumor is subdivided into many different subtypes. The most typical is composed of tubules lined by Sertoli cells and interstitial clusters of Leydig cells. Patients with SLCT present with signs of defeminization followed by masculinization. Age of the patient, stage of the disease and degree of tumor differentiation based on morphology are the most important factors to consider in the management of the case.[3]

CASE REPORT

Multiparous 54-year-old lady presented with dysfunctional uterine bleeding. History of dilatation and curettage done twice and showed an anovulatory endometrium with no atypia or malignancy. Pelvic ultrasound showed no significant findings. Pap smear of cervix showed inflammation with no intraepithelial lesion. Later, total abdominal hysterectomy with bilateral salpingo ophorectomy was performed.

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Gross Examination

Panhysterectomy specimen received in 10% formalin (Figure 1a). Cut section of cervix was unremarkable and uterus showed myometrial hyperplasia with coarse trabeculations. Cut section of left ovary showed a single cyst measuring 0.5 cm in diameter and right ovary showed a well-circumscribed tumor measuring 1.5 cm × 1 cm with lobulated tan yellow solid and micro cystic areas. Solid areas were firm in consistency (Figure 1b).

Microscopic Examination

Showed chronic cervicitis with nonsecretory proliferative endometrium and adenomyosis with myometrium showing few thick walled blood vessels. The left ovary showed ovarian tissue with a cyst with features of paraovarian cyst.

Right ovary showed ovarian tissue with a tumor composed of tubules and glandular structures lined by Sertoli like cells (Figure 2a) separated by stroma with nests of Leydig type cells (Figure 2b).

The diagnosis of right ovarian lesion was SLCT, (well-differentiated) Immunohistochemistry (IHC) was suggested for confirmation. IHC report showed, (Figure 3).

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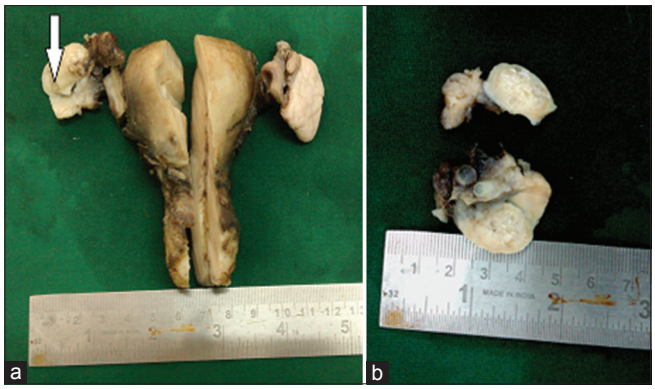

Figure 1: (a) Panhysterectomy specimen with right ovary showing a neoplasm, (b) C/S of right ovary showing a well circumscribed tumor measuring 1.5 cm × 1 cm with lobulated tan yellow solid areas and micro cystic spaces

Inhibin: Positivity.
Calretinin: Positivity.
Epithelial membrane antigen (EMA): Negativity.
CK 20: Negativity.
CK 7: Negativity.
CD10: Negativity.

Therefore, a final diagnosis of SLCT – well-differentiated (Meyer's Type I) was made.

DISCUSSION

The SLCT (androblastoma, arrhenoblastoma) is a gonadal tumor of sex-cord- stromal type in which the components to a greater or lesser extent recapitulate the cells of the testis at various stages of development.[4] WHO defines them as tumors containing various combinations of Sertoli–Leydig (S–L) cells and cells resembling rete epithelial cells and fibroblast in variable degrees of differentiation.[5]

SLCTs are uncommon tumors of ovary - accounting for <0.1% of all ovarian neoplasms. The majority of these patients are seen during the second and third decades of life, with the average age at diagnosis being 25 years. It is very rare after menopause. Some are diagnosed during pregnancy.[6] 50% of cases come to clinical attention because of progressive defeminization, virilization, and pelvic mass. Androgenic excess is manifested by defeminization, like amenorrhea, breast atrophy, loss of subcutaneous tissue deposits and later by musculinization like deepening of the voice, hirsutism, temporal alopecia and hypertrophy of clitoris and acne. Non-virilized patients present with non-specific symptoms like abnormal vaginal bleeding, abdominal distention, abdominal mass, or abdominal pain. Serum levels of testosterone and urine levels of 17-ketosteroids

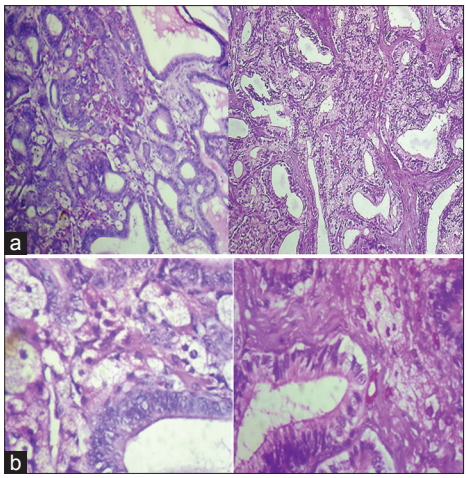

Figure 2: (a) Sertoli cells arranged in hollow tubules and glands with clumps of Leydig cells present in an edematous background. Another area showing clusters of Sertoli cells with clear cytoplasm lining the tubules with little intervening stroma (H and E stain, ×100 magnification), (b) Intermixed Leydig and Sertoli cells in the loose stroma at a higher magnification (H and E stain, ×400)

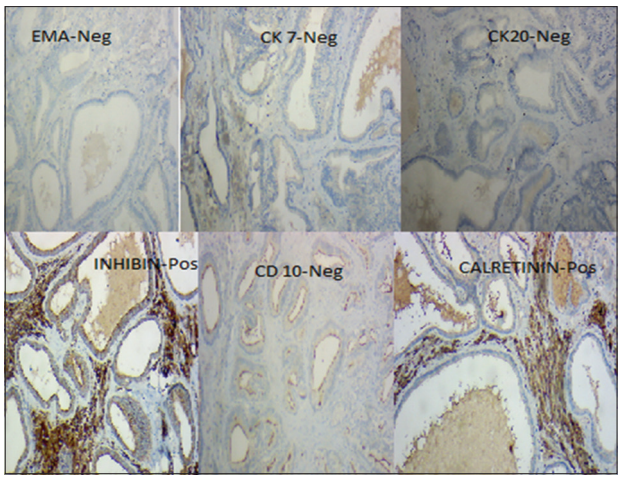

Figure 3: Immunohistochemistry – right ovarian tumour

are increased in virilized pts. Some cases with foci of heterologous hepatic differentiation are associated with elevation of serum alpha fetoprotein (AFP).[7] Rarely SLCTs are discovered in asymptomatic women.[7] Our patient who was 54-year-old multiparous lady had an uncommon presentation as it was an incidental finding and she had abnormal vaginal bleeding. The abnormal vaginal bleeding is due to the estrogen and progestrone imbalance created by the tumor.[6]

Gross appearance of the tumor is predominantly solid and partly cystic. Well-differentiated tumors are unilateral, solid and encapsulated (average size 5 cm). Intermediate and poorly differentiated tumors are larger (average size 15 cm) with areas of cystic change and necrosis. Papillae are sometimes visible in tumors with retiform differentiation.[7]

Microscopically they have an extremely variable histological pattern. Five categories are distinguished.[8] Meyer has graded this tumor according to differentiation into three types. Well differentiated (Meyer Type I) 11%, intermediate (Meyer Type II) - 54%, poorly differentiated (sarcomatoid, undifferentiated, Meyer Type III) - 13%. Other two categories include those with heterologous elements (teratoid androblastoma) - 22% and retiform - 15%.[8] Heterologous elements are represented by endodermal elements such as cysts and glands and mesenchymal elements such as bone, cartilage or skeletal muscle. The SLCT with heterologous mesenchymal elements are usually poorly differentiated in contrast to neoplasms with endodermal elements, which typically are of intermediate differentiation.[9] Retiform pattern simulates rete testis with highly branching ribbon like tubules with formation of papillary structures with hyalinized core.[10]

Differential diagnosis of SLCTs varies with differentiation. Well-differentiated ones have to be ruled out from gonads of the testicular feminization syndrome. Because of their wide range of morphologic patterns they may be confused histologically with a variety of other primary as well as metastatic ovarian tumors. Important differentials include sertoliform endometrioid carcinoma and metastatic carcinoma, especially adenocarcinoma. Those with heterologous elements, intermediate and sarcomatoid pattern have to be differentiated from teratoma, malignant mixed mullerian tumor, carcinosarcoma, malignant mesodermal mixed tumor, and carcinoid tumors (especially trabecular variant of carcinoid).[11]

IHC

S–L cells stain positive for inhibin alpha, calretinin, AFP (if heterologous hepatoid cells are present), testosterone, estradiol, CK (weak positivity), CD99 (membrane positivity) and WT-1. Recently, SLCTs are being added into the list of WT-1 positive tumors. They are negative for EMA, placental alkaline phosphatase, carcinoembryonic antigen, CA19.9, CA-125, S-100. These help to rule out other metastatic malignancies. From the practical viewpoint, the most helpful immunohisto chemical findings are the negative staining of sex cord tumors for EMA, and positive staining for inhibin and calretinin; findings that are converse to those seen in endometrioid carcinomas of the ovary, which commonly have formations that may simulate sex cord tumors.[12]

Recent studies have shown that many cases of SLCT of the ovary are caused by germline mutations in the DICER1 gene. These hereditary cases tend to be younger, often have a multinodular thyroid goiter and there may be a personal or family history of other rare tumors such as pleuropulmonary blastoma, Wilms tumor and cervical rhabdomyosarcoma.[13,14]

Prognosis and treatment correlates with stage and degree of differentiation of tumor.[6] Well-differentiated tumors are benign with no recurrence after complete excision.[7] The most reliable indication of malignancy is evidence of local extraovarian spread or metastases at the time of staging laparotomy.[6] The incidence of clinical malignancy in Sertoli stromal cell tumors is 18%. Well-differentiated tumors have 0% chance of malignancy and intermediate have 11%, poorly differentiated tumors are 59%, clinically malignant. 19% of tumors with heterologous elements, and 25% of retiform tumors with intermediate differentiation behave clinically malignant.[6] Treatment for young women with Stage I tumors are unilateral salpingoophorectomy. If poorly differentiated elements or heterologous elements are present, adjuvant therapy with radiation or combination chemotherapy is indicated. Stage II or higher require total abdominal hysterectomy with bilateral salpingo ophorectomy. Adjuvant therapy may be considered according to differentiation.[7]

CONCLUSION

SLCTs are uncommon tumors of ovary which as an incidental finding in old age group without history of mass, abdominal pain, abdominal distention, masculinization, and infertility is very rare and this case is presented for its rarity.

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