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Pathology

RETIFORM SERTOLI LEYDIG CELL TUMOR OF OVARY IN A 55 YEAR OLD WOMEN: A CASE REPORT AND REVIEW OF THE LITERATURE

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ABSTRACT **Background-** Sertoli-Leydig Cell Tumor is a rare ovarian tumor that belongs to the group of sex-cord stromal tumor. Ovarian sex cord stromal tumors arises from the stromal cells that surround and support the oocytes. These Neoplasm accounts for less than 0.5% of ovarian neoplasm and occurs at an average age of 25 years , except for the Retiform variant that trends to present at younger age (approx. 15 years).

Case Report- A 55 years old woman presented with pelvic pain was found to have large complex left adnexal mass on CT. Total Abdominal Hysterectomy with Ovarian Cystectomy was done and the specimen was received in our Department of Pathology Dr S N Medical College, Jodhpur for Histopathological examination. The Ovarian mass was diagnosed as Sertoli-Leydig Cell Tumor, Retiform variant StageIA. While the usual age of presentation of these tumors are around 15 years of age.

Conclusion-This case report is presented because of the rarity of The Retiform Sertoli-Leydig Cell Tumor of Ovary and the unusual older age of 55 years at the time of diagnosis. Sertoli-Leydig cell tumors, including Retiform variant, often present a diagnostic challenge due to the various morphological patterns that may be assumed by the tumor.

KEYWORDS : Retiform variant, Sertoli-Leydig cell tumor, Sex cord stromal tumor.

INTRODUCTION

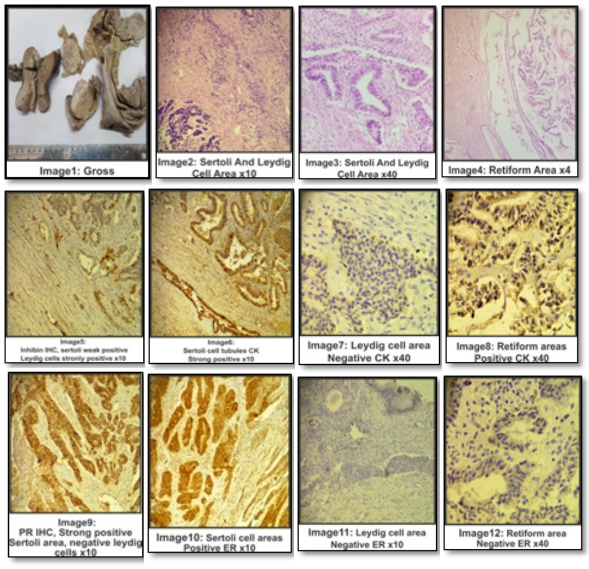
Ovarian sex cord-stromal tumor include pure form like fibroma, thecoma, granulosa cell, sclerosing stromal tumor, Sertoli cell tumor etc. Mixed form also exists, a category comprised of Sertoli-Leydig cell tumor (SLCTs) and sex cord stromal tumor NOS(1). Malignant sex cord stromal tumor of ovary are rare comprising 1-2% of all primary ovarian tumor(2). SLCTs are even more uncommon accounting for less than 0.5% of all primary ovarian neoplasm. SLCTs are classified in five major classes:-well differentiated, intermediate differentiated, poorly differentiated, heterogenous and retiform. Retiform variant account for 10% of SLCTs and are most frequent in second decade. Usual SLCTs produce androgens, resulting in signs of virilization, which is less common in retiform variant. This case is presented because retiform variant SLCT are Per se less common and it is presented in a 55 year old women which is unusual age for this tumor.

CASE REPORT

A 55 years old postmenopausal female came to gynae OPD with complains of pain and heaviness in lower abdomen with spotting PV on and off from six months. No other complain given by patient. On clinical examination a mass was felt per abdomen consistent with 32 weeks of gestational size. PV examination shows a mass in left adenaxal region. CT abdomen and pelvis shows a large well defined solid cystic mass measuring 12x10x06cm in left adenaxal region ? cystadenocarcinoma. CA 125 was 23.06 units/ml, AFP levels not done. Rest of hematological and biochemical investigations were within normal range except for mild microcytic hypochromic anemia. Total abdominal hysterectomy with bilateral salpingo-oophorectomy was done and specimen sent for histopathological examination in our department of pathology DR S N MEDICAL COLLEGE JODHPUR. Specimen of uterus cervix with bilateral adenaxa was received, uterus measuring 6x3x3 cm, endocervical canal patent, endomyometrium thickness was 1.5cm. Right ovary was 3x1.5x0.5 cm. Left ovary was measuring 12x10x06 cm, on cutting it was solid cystic, solid area measuring 7x4x4 cm, cyst was filled with mucinous, haemorrhagic grey black fluid. Solid area showed papillary excrescences, nodularity along with vascularity, capsule was intact with peritoneal fat attached. The thickness of cyst wall was 0.3 to 0.5 cm .

Microscopic examination of different areas from ovarian mass showed three different patterns. The predominant pattern was of well formed tubules, ill defined tubules, trabecular pattern as well as aggregates and nest of sertoli cells. Sertoli cells are columnar to cuboidal, have round to oval nuclei, the cytoplasm showed vacuoles. The other component showed aggregrates of leydig cells, present in stroma having abundant eosinophilic cytoplasm, centrally placed round to oval nuclei, no cytological atypia or abnormal mitosis seen. The third component present at the periphery of the nodules comprising of retiform tubules which were long branched with focal papillary appearance. They are lined by low columnar to cuboidal cells with scanty cytoplasm and oval hyperchromatic nuclei. A diagnosis of sertoli leydig cell tumor of intermediate differentiation with retiform component, STAGE IA was given.

IHC in solid areas showed strong and diffuse staining for pancytoker atin, positive staining for Inhibin, ER and PR. Leydig cells showed strong Inhibin positivity, ER and PR was negative. Retiform area showed positivity for cytokeratin and PR, Inhibin was weakly positive and ER was negative .



DISCUSSION

SLCTs are rare, making upto 0.5 % or less of all all primary ovarian neoplasm(3), with a mean age of 25 at diagnosis, the average age for

retiform SLCT is much younger at 15 years(3). On reviewing the literature approx 65 cases of retiform SLCT published in different case reports and case series(4,5,6). The age of presentation is 2-39 years(7), 11mnt to 23 years(8), with the average being 15 or 17 years respectively. Only one distinct case report of this variant of SLCT published in 70 year old at the time of diagnosis(9). The presenting complain of SLCTs was related to abdominal mass effect with endocrine manifestation in only minority of patients. Laura C. et al(10) found only one case of retiform SLCT in his series of 532 neoplastic ovarian cases .

40-60% cases of SLCT show androgenic menifestation like devolopment of muscle bulk, hirsutism, deepning of voice, amenorrhea, breast atropy, acne etc. In few, estrogenic manifestation have been reported. Patients without endocrine manifestation show complaints attributed to mass effect. Elevated AFP seen in some cases, but they are generally lower than seen in yolk sac tumor(11).

SLCTs are typically unilateral with bilaterality in only 2%. The average diameter is 12-14 cms. They may be solid, solidcystic, and purely cystic. Solid areas appear fleshy, lobulated, tan pink, pale yellow or grey. Papillary excrescences may be grossly identified on the cyst wall.

SLCTs are divided into five histologic subtypes including well differentiated , moderately differentiated , poorly differentiated, SLCT with heterologous elements and retiform SLCT. The well, moderate and poorly differentiated forms are assigned based on the degree of tubular differentiation of sertoli cell component, which decreases with increasing grade, and the amount of primitive gonadal stroma present within the tumor, which increases with increasing grade. Leydig cells also trends to decrease with increasing tumor grade(1).

The differential diagnosis for well differentiated SLCTs includes endometroid adenocarcinoma with sertoliform glands. The conspicious tubules, presence of leydig cells and absence of squamous differentiation in SLCT helps to differentiate from endometroid adenocarcinoma. SLCTs are positive for Inhibin, Calretenin and FOXL2, ovarian endometroid adenocarcinoma are negative for these and are positive for CK7 and EMA. Moderately to poorly differentiat ed SLCTs are difficult to distinguish from germ cell tumor, serous neoplasm, carcinosarcoma and rare primary ovarian wilms tumor.

The retiform variant is diagnosed when a significant portion of the tumor demonstrate network of anastomosing, slit like spaces and tubules lined by flattened to cuboidal sertoli cells in a single layer or stratified pattern(11). According to mooney et al(11) a primary ovarian neoplasm to be consider as retiform variant, should contain more than 5% of tissue area on slides composing of retiform pattern. Other pattern that may be seen in retiform SLCT includes area with papillary architecture and multicystic areas with slit like spaces lined by low cuboidal to flattened sertoli cells. Stromal edema and glomeruloid structures may be seen. The retiform pattern may focally be present in 10% of moderately to poorly differentiated SLCTs.

IHC:-Solid areas composed of Sertoli cells are positive for Pancytokeratin, Beta-catenin, Calretinin, WT-1, ER, PR. While negative for CK7, Cyclin-D1, Napsin, CK20, p53, AFP, Cd10.

Leydig cells are positive for Inhibin (strongly), Calretinin, Beta-catenin. While negative for Pancytokeratin, CK7, ER, PR, Cyclin-D1, P53, AFP, CD10 etc.

Retiform areas are positive for Pancytokeratin, Beta-catenin, Calretinin, WT-1, PR, CK7, Inhibin (weak). While negative for ER, Glypican-3, Cyclin-D1, CK20, p53, AFP, CD10, Hepar-1.

SLCTs have an overall favourable prognosis. About 100% survival has been reported in well differntiated tumors. The presence of retiform growth pattern is thought to confer slightly worse prognosis in case of moderetely differentiated SLCT.

CONCLUSION

This case is presented because of rarity of this type of tumor and its presentation in atypically older patient of 55 years age at time of diagnosis. Retiform SLCTs often presents diagnostic challenge because of various morphologic patterns . IHC is required to facilitate accurate diagnosis and inclusion of Inhibin is essential in panel of stains. Unilateral salpingo-oophorectomy is treatment of choice, but it remain unclear wheather complete staging or postoperative adjuvant chemotherapy is necessary for the management of retiform SLCT. Further large sample size and longer follow up is needed for better prognostication and to determine wheather chemotherapy has a role in managing patients with retiform SLCT.

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