**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 neoplasm. SLCTs are even more uncommon accounting for less than 0.5% of all primary ovarian neoplasms. 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 adnexal region. CT abdomen and pelvis shows a 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 Stage IA. While the usual age of presentation of these tumors are around 15 years of age. 

**DISCUSSION**

SLCTs are rare, making up to 0.5 % or less of 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. 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 manifestation like development of muscle bulk, hirsutism, deepening of voice, amenorrhoea, breast atrophy, 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, solid-cystic, 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 conspicuous tubules, presence of Leydig cells and absence of squamous differentiation in SLCT helps to differentiate from endometroid adenocarcinoma. SLCTs are positive for inhibin, Calretinin and FOXL2, ovarian endometroid adenocarcinoma are negative for these and are positive for CK7 and EMA. Moderately to poorly differentiated SLCTs are difficult to distinguish from germ cell tumor, serous neoplasms, carcinosaarcoma 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 multicytic 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 etc.

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 differentiated tumors. The presence of retiform growth pattern is thought to confer slightly worse prognosis in case of moderately 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 whether 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 whether chemotherapy has a role in managing patients with retiform SLCT.

REFERENCES