Case Report

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"An Intriguing Case of Virilising Sertoli-Leydig Cell Tumour of the Ovary"

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ABSTRACT

Sertoli-Leydig cell tumours (SLCTs) are rare sex cord stromal neoplasms of ovary. They are more commonly seen in reproductive age group and are rare in postmenopausal period. The average age of presentation is 30yrs. We report a rare case of Leydig cell tumour with an interesting presentation in a postmenopausal woman.

A 60-year-old woman, postmenopausal for 12 years, presented with hirsutism, male pattern baldness and deepening voice over the past five years. On general examination she had hirsutism, male pattern baldness with hoarseness of voice. Her blood counts and metabolic profile was normal. However, endocrinology work-up showed increased levels of testosterone (980ng/dl) (N 15-70 ng/ml).MRI Abdomen and Pelvis revealed a left adnexal mass measuring 4.1x4.0x3.3 cm which was heterogeneous with a centrally hyper intense area. No invasion in the surrounding structures was seen. No ascites was present. Total Abdominal Hysterectomy with Bilateral Salpingo-oophorectomy was done. Histological examination and immunohistochemical findings were of Sertoli-Leydig cell tumour. Postoperatively the symptoms of the patient abated and the levels of testosterone came back to normal.

Keywords: *Virilizing ovarian tumours, Sex cord-stromal cell tumour, Sertoli-Leydig cell tumour, Immunohistochemistry, testosterone*

Introduction

Tumours derived from the sex cords or ovarian mesenchyme are rare and constitute 5% to 12% of all ovarian neoplasms. [1] Sertoli-Leydig cell tumours (SLCTs) are rare sex cord stromal tumours of ovary constituting less than 0.5% of all ovarian tumours. These cases are seen in women of reproductive age group with seventy-five percent of them below 30 years. These patients come to notice with signs of virilization due to over production of testosterone.[2]

We report an interesting case in a menopausal woman who presented with signs of virilization and was diagnosed as SLCT.

Case Report

A 60-year-old postmenopausal lady presented with progressive hirsutism, male pattern baldness and deepening of voice of five-year duration. Physical examination revealed signs of virilization with hirsutism and deep voice. She had normal blood counts. Her metabolic profile was within normal limits. However, endocrinology work up showed increased levels of testosterone (980ng/dl) (N 15-70 ng/ml). Serum tumour markers (CA125, CA19.9, and α-FP) levels were within the normal range, and chest X-ray and EKG were normal. MRI Abdomen and Pelvis revealed a left adnexal mass measuring 4.1x4.0x3.3 cm which was heterogeneous with a central hyper intense area. No invasion in the surrounding structures was seen. No ascites was present.

Total Abdominal Hysterectomy with Bilateral Salpingo-oophorectomy was done under general anaesthesia. On laparotomy a left ovarian mass with intact capsule was noted. We received a specimen of total abdominal hysterectomy with bilateral salpingo-oopherectomy. Gross examination revealed a well circumscribed and solid tumour measuring 4.0 x 4.0 x3.0cm. Capsule was intact and external surface was smooth and lobulated. Cut section was solid, yellow and homogenous. (Fig-1) Uterus, fallopian tube and the right ovary were unremarkable.

Microscopic examination revealed a well circumscribed tumour [Fig-2] composed of sertoli cells arranged in cords and trabeculae separated by fibrous stroma. The individual cells were large with eosinophilic cytoplasm. Adjacent to these, Leydig cells with eosinophilic and finely granular cytoplasm were noted. No cytological atypia or mitoses seen. No necrosis or hemorrhage seen. On immunuhistochemistry the tumour cells were positive for Inhibin, Calretinin and Negative for EMA.

Based on the histomorphology and immunohistochemistry the tumour was diagnosed as Sertoli-Leydig cell tumour of left ovary well differentiated. This was stage IA (International Federation of Gynaecology and Obstetrics) tumour.

Postoperatively, the patient was regularly reviewed. The patient's symptoms abated and the levels of testosterone also reverted to normal levels.

Discussion

Sex cord stromal tumours are uncommon and presentations are varied and intriguing. These are composed of various combinations of granulosa cells, theca cells, Leydig cells, Sertoli cells, and fibroblasts of stromal origin [3]

These patients either present with abdominal pain due to space occupying lesion or with androgenic or estrogenic expression. Androgenic manifestations are hirsutism, clitoromegaly, acne, hoarseness of voice and estrogenic manifestations are abnormal uterine bleeding, precocious puberty, menstrual irregularities, breast hypertrophy, weight gain, generalised oedema, endometrial hyperplasia, endometrial polyps and endometrial carcinoma. [3]

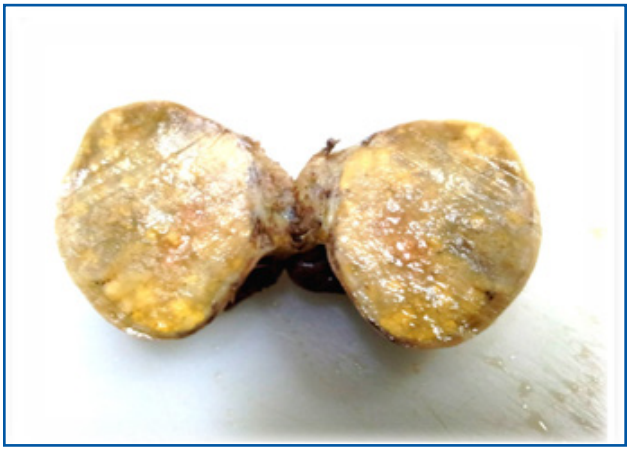

Fig. 1: Gross appearance showing well circumscribed & solid tumour with smooth external surface & solid, yellow and homogenous cut surface.

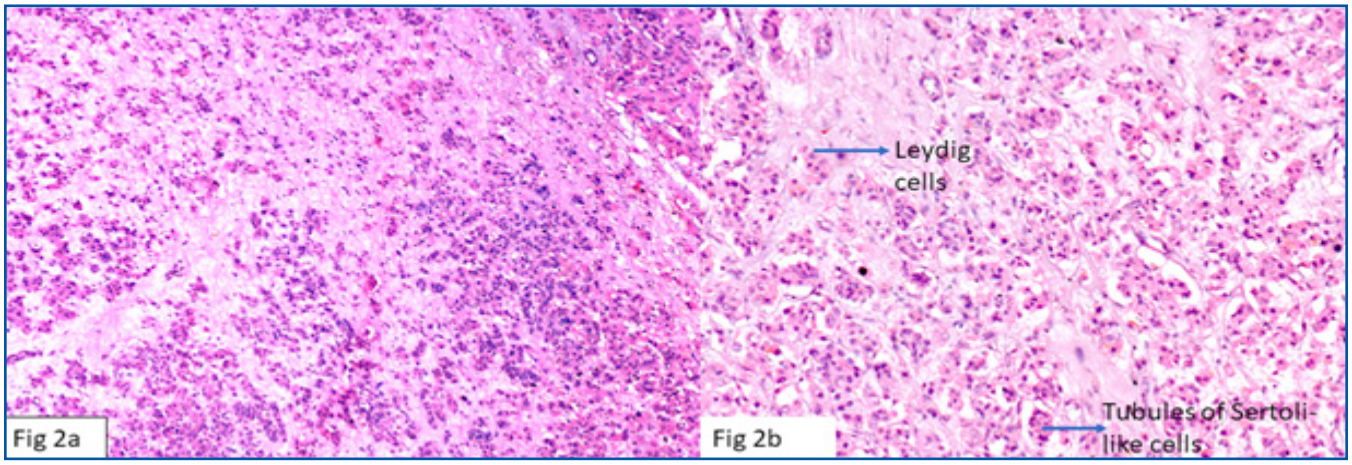

Fig. 2: (a & b). Photomicrograph showing cords of mature Sertoli cells and clusters of Leydig cells. Haematoxylin& eosin (a-100X,b-400X).

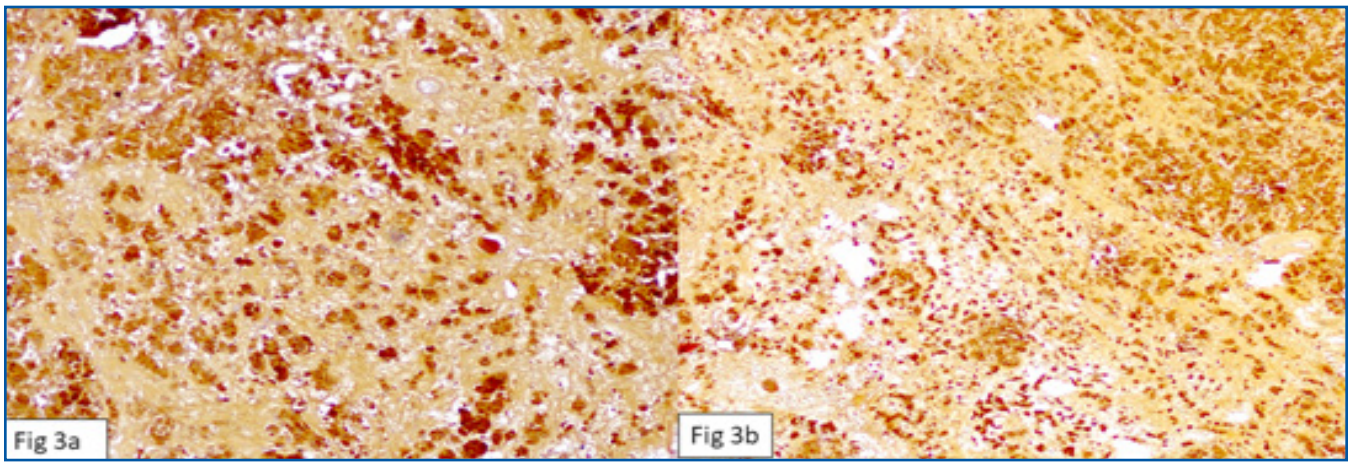

Fig. 3: (a & b). Photomicrograph showing immunohistochemistry with tumour cells positive for calretinin and alpha-inhibin. (3a- Calretinin,100X, 3b- Alpha inhibin, 100X).

Mostly these are confined to ovaries, unilateral, and usually stage I at the time of clinical diagnosis. [3,4]

Grossly, they are predominantly solid, may have cystic areas. Microscopically, they are composed of a mixture of cells resembling male Sertoli and Leydig cells. Well differentiated tumours are composed of tubules lined by Sertoli-like cells and variable numbers of Leydig-like cells. Intermediate grade tumours are characterized by sheets and cords of Sertoli-like cells, separated by spindle stromal cells and recognizable Leydig cells. Poorly differentiated tumors show a sarcomatoid pattern. [5] Degree of differentiation corresponds to patient prognosis [2]

Many tumours mimic the appearance of these tumours, therefore it becomes important to thoroughly sample the tumours and correctly diagnose them. Most importantly,well-differentiated endometrioid carcinoma may mimic Sertoli Leydig cell tumours as it may show tubules similar to Sertoli Leydig cell tumours. Histologically presence of confluent pattern of endometrioid carcinoma and presence of apical mucin may help in diagnosing endometrioid carcinoma.[6]

Immunohistochemical expression of these tumours includes positive staining for inhibin and caltretinin and negative staining for epithelial membrane antigen (Weng et al., 2013). [7] Endometrioid tumors have immunohistochemical staining positive for EMA while their neoplastic glands are negative for inhibin and caltretinin.[8] Immunohistochemistry thus can supplement the diagnosis in challenging cases.

Most patients present with tumors confined to the ovary and have a favorable prognosis. These patients are treated surgically. Conservative surgery with unilateral salpingo-oophorectomy with staging is done in women of reproductive age group. In case of older patients, advanced disease/bilateral involvement the treatment of choice is abdominal hysterectomy and bilateral salpingo-oophorectomy with surgical staging.[9]

Following total abdominal hysterectomy and bilateral salpingo-oophorectomy, there is regression of symptoms and serum testosterone returns to normal.[1,4] Therefore appropriate diagnosis and treatment is crucial in these cases.

Conclusion

As these patients may have varied clinical presentations one should be aware of the same. An androgen producing tumour should be excluded in women presenting with hirsutism and markedly elevated testosterone levels. Appropriate investigations, thorough histopathological examination complemented by immunohistochemical studies are crucial in management of these patients.

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