

Sertoli leydig cell tumour - a rarity in reality

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Abstract

Introduction: Sertoli Leydig Cell Tumour of the ovary is an extremely rare tumour that belongs to a group of Sex Cord Stromal tumours of the ovary. It accounts for less than 0.5% of all primary ovarian neoplasms. Very few cases are documented in the literature so far. Hence we report a case of Sertoli Leydig Cell Tumour of intermediate grade differentiation involving the left ovary in a 26 years old nulliparous woman who presented with menstrual irregularity and virilizing features.

Keywords: Sertoli Leydig Cell Tumour, Sex cord stromal tumour, Ovarian tumours.

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INTRODUCTION

Sertoli-Leydig cell tumor (SLCT) of ovary is an exceedingly unusual neoplasm that belongs to a group of sex cord-stromal tumors of ovary and accounts for less than 0.5% of all primary ovarian neoplasms¹ and is the most common virilizing ovarian tumour. It is characterized by uncontrolled proliferation of naturally occurring testicular structures (Sertoli and Leydig cells) of varying degrees of differentiation in ovary. The neoplastic Sertoli and Leydig cells exhibit varying degrees of differentiation (grading) which include well differentiated, moderately differentiated, poorly differentiated, and with heterologous elements². The case report of SLCT is presented in view or rarity of the condition, limited number of case reports, series and clinical trials documented and lack of standard treatment guidelines.

CASE REPORT

A 26 years old nulliparous woman married for 5 years presented with menstrual irregularity of oligomenorrhoea for 2 years followed by amenorrhoea of 2 years duration with primary infertility and excessive

hair growth. Her past medical and surgical histories were unremarkable. Her physical examination revealed hirsutism, breast development of Tanner stage IV and pubic hair of Tanner stage III. There was moderate free fluid with no palpable mass per abdomen. Local examination revealed clitoromegaly with male pattern of distribution of pubic hair. Bimanual manual examination showed a normal sized uterus, left adnexal mass solid to cystic of 7x8cm, mobile and non tender, other fornices were free. Laboratory tests showed a raise in total testosterone of 586ng/dl (N - 14- 76ng/dl), DHEAS of 255.9 mg/dl (N - 35-430mg/dl), CA125 of 20.7U/ml (N - 21U/ml), FSH < 0.3 mIU/ml, LH - 13.04 mIU/ml, Estradiol - 85.6pg/ml. Thyroid profile, serum Prolactin other tumour markers like AFP, CEA were normal. Ultrasound pelvis showed a large midline cyst 9.3x8.3x5.3 cms with solid and cystic areas with vascularity in the substance of the cyst, venous Doppler normal, moderate free fluid seen in the pelvis. MRI abdomen and pelvis showed a well circumscribed lobulated heterogenous multiseptated cystic lesion of 8 x 7 x 9.5 cms with solid components and hemorrhage in the left adnexa, left ovary is not seen separately, no lymphadenopathy, moderate ascites, no focal lesions in liver or adrenals. In view of left ovarian mass, moderate ascites, virilizing features, patient was subjected for Staging Laparotomy and procedure. Intraoperatively, there was moderate ascites, left ovarian mass of 8x7 cm with solid and cystic areas. Uterus, fallopian tubes and right ovary were normal. Hence left sided oophorectomy was done. Frozen section biopsy of left ovarian mass showed possibly sex cord stromal tumour. Macroscopically, the mass weighed 235g with smooth surface measuring 8x7x3.5cm. The cut section showed

solid and cystic areas grayish brown to yellow with focal areas of necrosis. Microscopically, it was sex cord stromal tumour - Sertoli Leydig Cell Tumour (SLCT) of intermediate grade with capsule intact and ascitic fluid negative for malignant cells. The pathological staging was T1a N0 M0. Immunohistochemically, tumour cells were positive for Vimentin, Inhibin, and WT1. They were negative for Glypican and AFP. The patient had a quick and uneventful post operative recovery. The hormonal assay prior to discharge showed a dramatic fall in testosterone to normal range (from 586.06 to 30.94 ng/dl) and 17 OH progesterone from 3.05 to 1.92ng/ml. She was asymptomatic with spontaneous onset of menstrual cycles after four weeks.

DISCUSSION

Sertoli Leydig cell Tumour (SLCT) is an extremely rare tumour of the ovary accounting for less than 0.5 % of all primary ovarian neoplasm¹. Even though it can occur in any age group, it is common around 25 years as in our patient. Less than 10% can have prior to menarche or following menopause¹. Majority of SLCT are unilateral, mostly confined to ovaries and nearly 90% are classified as stage I at the time of clinical diagnosis as in our patient. The tumour cells show varying degrees of differentiation (grading) which include well differentiated, moderately differentiated, poorly differentiated and heterologous elements³. The clinical features can be related to either hormone production (testosterone) or due to abdomino pelvic mass. Our patient presented with virilizing features of hirsutism, (figure 1) receding hair line, clitoromegaly, (figure 2) oligomenorrhoea and amenorrhoea but no hoarseness of voice. As she had moderate ascites mass could not be palpated abdominally. Elevated serum levels of testosterone and androstenedione is seen in 80% of SLCT with virilizing manifestations^{4,5}. 17 -OH keto steroids are often normal or mildly elevated unlike virilising adrenal tumours which shows extreme elevation. Imaging studies can be utilized for the diagnosis. Trans vaginal ultra sound remains the best imaging modality for the initial assessment of adnexal masses. Other imaging modalities like Computerized Tomography (CT), Magnetic Resonance Imaging (MRI) can be used for better characterization of ovarian SLCT, detection of metastasis and other primary neoplasms. Management of ovarian SLCT remains challenging due to the lack of standard management protocol. Surgical resection remains the main stay of the management⁶ Fertility sparing surgery of salphingo oophorectomy can be considered in majority of patients as they are usually in stage I grading at the time

of diagnosis. So we proceeded with unilateral oophorectomy in our patient. The need for pelvic lymphadenectomy is debatable. The prognosis of SLCT depends on the degree of differentiation and tumour extent. The overall 5 year survival rate for well differentiated tumour is 100% whereas for moderately and poorly differentiated SLCT's is collectively 80%⁷

CONCLUSION

Sertoli Leydig Cell Tumour even though rare should be thought of in a young women presenting with virilizing features and conservative surgery is sufficient in majority of cases.



Figure 1: Patient with hirsutism and Masculine features



Figure 2 Showing Clitoromegaly and male pattern of hair distribution

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