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BMH Med. J. 2023;10(3):79-82. **Case Report**

Steroid Cell Tumour - A Rare Ovarian Tumour Variant

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Abstract

Steroid cell tumors of the ovary are extremely rare, of all ovarian tumors. The tumors belong to the sex cord-stromal tumors and account for <0.1% of all ovarian neoplasms. Here we present a case of a young lady who presented to us with amenorrhoea, with thorough history taking, diagnostic workup and postoperative histopathological evaluation found to have a rare kind of ovarian tumour - Steroid cell tumour.

Keywords: Steroid cell tumour, sex cord stromal tumour, ovarian tumour

Introduction

Steroid cell tumors of the ovary are extremely rare, of all ovarian tumors [1]. The tumors belong to the sex cord-stromal tumors and account for <0.1% of all ovarian neoplasms [2].

Case report

A young parous lady presented with complaints of 6 month amenorrhoea, excessive hair growth over face and neck and right sided abdomen pain of acute onset, of one day duration. Her menstrual cycles were irregular. She was a diagnosed case of polycystic ovarian disease on lifestyle modification and drugs. Her obstetric status was P1L1A3 and previous LSCS, 8 years back. No comorbidities. No significant past medical, surgical or family history.

On evaluation, she was well built, obese, with hirsutism, vitals stable and systemic examination normal. Abdomen was soft, obese. No mass was palpable. On local examination clitoromegaly noted. Speculum examination showed healthy cervix. Bimanual pelvic examination showed bulky uterus, mobile, fornices free.

USG abdomen showed, uterus normal size, anteverted, right ovary posteriorly displaced in broad ligament region with echogenic area 60 x 40 mm. Left ovary was polycystic.

MRI showed solid right ovarian mass with no malignant features. Beta hCG was 0.09 mIU/ml, AFP 1.22 IU/ml and CA125 26.37. Testosterone level was elevated to 38.90 nmol/L. Bioavailable testosterone was 23.800nmol/L, FSH 9.2IU/L, Prolactin 333mIU/L, AMH 1.82pmol/L, SHBG

22.7nmol/L. HPV DNA Negative. Proceeded to Laparoscopic surgery. Right ovary showed a 6 x 6 cm tumour filled with sebaceous material. Left ovary was polycystic. Uterus was bulky. Fallopian tubes were normal. Laparoscopic right ovariotomy + Left ovarian drilling was done. Specimen retrieved out in endobag and sent for histopathological examination. Histopathological evaluation of the tumor was reported as steroid cell tumor.

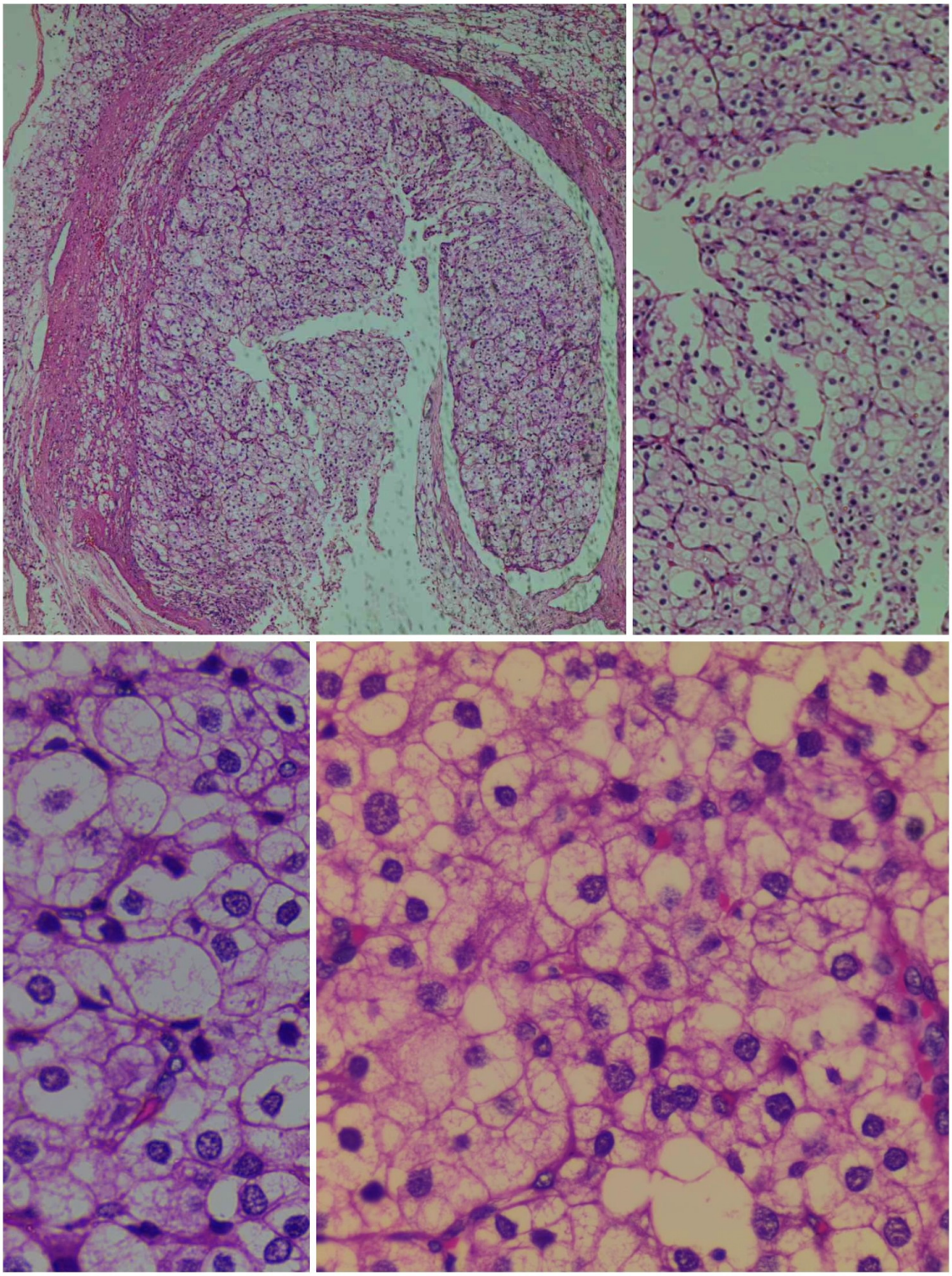

Figure 1: Low power and high power microscopic images of steroid cell tumor. Neoplasm composed of solid nest of large polygonal cells with central round nuclei having fine chromatin and abundant clear cytoplasm.

At one month post surgery, testosterone level came down to 1.08 nmol/L, SHBG increased to 45.8 nmol/L. She had periods 25 days post surgery. After that, she had 3 regular menstrual cycles and became pregnant 4 months post surgery.

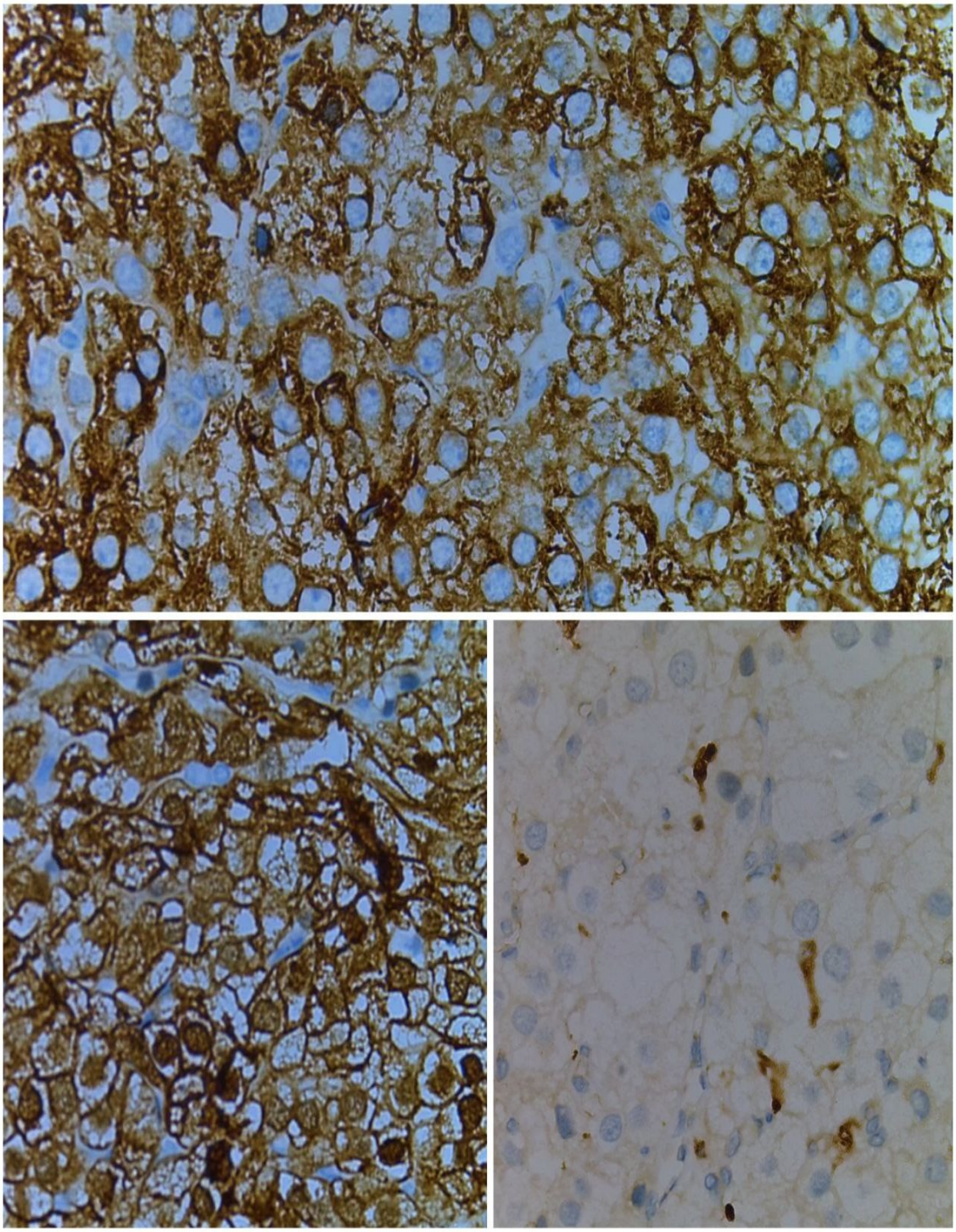

Figure 2: Immunohistochemistry showing positive for inhibin and calretinin and negative for CD68.

Discussion

Steroid cell tumors belong to the class of sex cord stromal tumours, account for <0.1% of all ovarian neoplasms. It is an ovarian parenchymal tumour composed of steroid cells. WHO Classification of Tumours, 5th Edition, Volume 4 classified steroid cell tumour into Steroid cell tumors NOS, and steroid cell tumor malignant [3]. Steroid cell tumors, NOS, account for approximately 60% steroid cell tumor subtypes [4]. Mean age of occurence of Steroid cell tumors NOS is 43 years. These tumors are usually unilateral, only 6% of cases are found to be bilateral. The clinical manifestations of the tumor are associated with its hormonal activity, tumour progression and virilizing properties.The usual clinical presentation includes abdominal pain, distention, irregular menstrual cycles, hirsutism

and features of hyperandrogenism. Serum testosterone and DHEA-S assays are the first tests used for the evaluation of either an adrenal or ovarian source of pathology for the hyperandrogenism. Serum testosterone level above 200 ng/dl is the important diagnostic threshold level for the discrimination of neoplastic source from other nonneoplastic causes of hirsutism. Transabdominal and pelvic ultrasonogram also aids in diagnosis. The final diagnosis of steroid cell tumor is by histopathological examination of tumor.

Table 1: Comparison of preoperative and postoperative hormone assay.

	preoperative	1 month post surgery
Testosterone	38.90nmol/L(normal 0.29-1.67)	1.08nmol/L
SHBG	22.7nmol/L(normal 32.4-128)	45.8nmol/L
Prolactin	333mIU/L(normal 102-495)	390mIU/L

Treatment decision depends on many prognostic factors including the stage of the tumor, the presence of malignant features, age of the patient, and fertility desire. In young patients who want to preserve their fertility, unilateral salpingo-oophorectomy is the preferred method of treatment [5]. In postmenopausal women the appropriate option is hysterectomy with bilateral salpingo-oophorectomy. Malignant NOS steroid cell tumors should be managed with surgical removal followed by a combination of chemotherapy and radiotherapy. The adjuvant chemotherapy regimens currently recommended for treatment are as follows: BEP; cisplatin, doxorubicin, and cyclophosphamide; taxane and platinum; and bleomycin, vinblastine, and cisplatin [6]. Gonadotropin-releasing hormone agonist could be used as postoperative adjuvant therapy. Regular follow-up evaluation with measurement of serum testosterone level is mandatory.

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