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Case Report

Steroid cell tumour-a rare ovarian tumour: a case report

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ABSTRACT

Steroid cell tumours of the ovary are sex cord stromal tumours which are rare, of all ovarian tumours. Case: Here we present a case of 35-year-old lady who presented to us with complaints of amenorrhoea. With thorough history taking, diagnostic workup and postoperative histopathological evaluation found to have a rare kind of ovarian tumor steroid cell tumor.

Keywords: Steroid cell tumor, Sex cord stromal tumor, Ovarian tumor

INTRODUCTION

Steroid cell tumors belong to the class of sex cord stromal tumours which are arising from ovarian parenchyma, composed of steroid cells.[1,2] The incidence is <0.1% of all ovarian neoplasms. WHO classified steroid cell tumour into Steroid cell tumors NOS, and steroid cell tumor malignant.[3] Steroid cell tumours, NOS, incidence is around 60% steroid cell tumor subtypes.[4] In this case report, we emphasize the importance of systematic clinical, laboratory and radiological methods for evaluation of secondary amenorrhoea and hirsutism and leading to a definitive diagnosis and prompt treatment for the same.

CASE REPORT

A 35-year-old parous lady presented with complaints of 6 months amenorrhoea, excessive hair growth over face and neck and right sided abdomen pain- acute onset 1 day. Her menstrual cycles were irregular. She was a diagnosed with polycystic ovarian disease, and was on lifestyle modification and drugs. LMP in January 2021. Her obstetric status is P1L1A3. Previous LSCS, 8 years back. No comorbidities. No significant past medical, surgical, family history. On evaluation, she is well built, obese, hirsutism noted, vitals stable, systemic examination normal. Abdomen was soft, obese. No mass palpable. On local examination clitoromegaly noted. speculum examination showed healthy cervix. Bimanual pelvic examination showed normal size uterus, mobile, Fornices free. USG abdomen with pelvis showed, Uterus normal size, anteverted, Right ovary posteriorly displaced in broad ligament region contains echogenic area 60×40 mm. Left ovary polycystic. MRI (15/6/21)-solid right ovarian mass 5×4.4×4.6 cm with no malignant features. Relevant blood investigations done. Beta hCG-0.09mIU/ml, AFP-1.22IU/ml, CA 125-26.37. Testosterone level was elevated to 38.90nmol/l (normal 0.29-1.67), Bioavailable testosterone-23.800nmol/l (normal 0.059-0.756), FSH-9.2IU/l, prolactin-333mIU/l (normal 102-495), AMH-1.82pmol/l. SHBG-22.7nmol/l (normal 32.4-128). HPV DNA-negative. Proceeded to laparoscopic surgery, right ovary showed a 6×6 cm tumor filled with sebaceous material. Left ovary polycystic. Uterus bulky. Right and left fallopian tube normal. Laparoscopic right ovariotomy with left ovarian drilling done. specimen retrieved out in end Endo bag and sent for histopathological examination.

Histopathological evaluation of the tumor reported as steroid cell tumor.

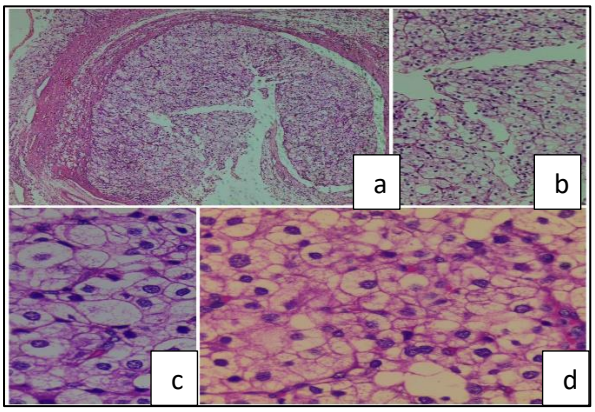

Figure 1 (a-d): Low power and high-power microscopic images of steroid cell tumor-neoplasm composed of solid nests of large polygonal cells with central round nuclei having fine chromatin and abundant clear cytoplasm.

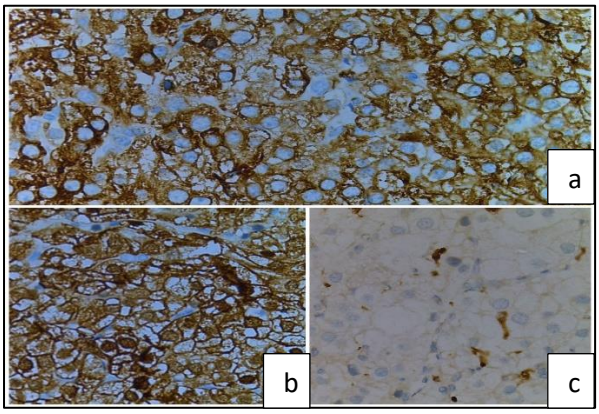

Figure 2: (a) Immunohistochemistry showing positive for inhibin and (b) calretinin and (c) negative for CD68.

After 1-month post-surgery, testosterone level came down to 1.08nmol/l, SHBG increased to 45.8nmol/l (normal 32.4-128). She got her periods 25 days post-surgery. Followed that, she got 3 regular menstrual cycle. She became pregnant 4 months post-surgery.

Table 1: Comparison of preoperative and postoperative hormonal assay.

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

DISCUSSION

Steroid cell tumors are sex cord stromal tumors composed of steroid cells.[1,2]. Steroid cell tumours, NOS, can be seen at all ages, and at a mean age of 43 years. Mostly these tumours are unilateral, only 6% of cases are found to be bilateral. The majority are benign.[2] SCT are frequently hormonally active, leading to elevated serum and/or levels of androgenic hormones and their metabolites. The symptoms of this tumor ranges from abdominal pain, distention, irregular menstrual cycles, and hirsutism .The symptoms are caused due to the increased hormonal secretion from tumor-mostly testosterone, and have virilizing properties. Steroid cell tumours cause androgenic and estrogenic manifestations in 50% and 10% of patients, respectively.

Majority of patients presents with hirsutism and virilization symptoms.[5] Approximately 25% shows estrogenic effects, such as menorrhagia, postmenopausal bleeding or even endometrial cancer. Rarely some presents with Cushing's syndrome (< 10%) due to ACTH secretion, and some cases have association with Von Hippel Lindau syndrome.[6] While approximately 25% are not associated with hormonal disturbances.[2]

Evaluation of cause of hirsutism may follow a systematic approach. We need to differentiate between, ovarian, adrenal, exogenous hormone secretion as cause of hirsutism. In contrast with PCOS androgen-secreting tumours, presents with progressive and severe hyperandrogenism leading to signs of virilization. Serum testosterone and DHEA-S assays are the first tests used for the evaluation of either an adrenal or ovarian cause for the hyperandrogenism. Serum testosterone level above 200 ng/dl is the important threshold level for diagnosing neoplastic causes of hirsutism. Adrenal tumors causing virilization are very rare.[7]

In patients with evidence of virilization, serum testosterone levels higher than 2 ng/ml, normal dehydroepiandrosterone-sulphate, and absence of 21-hydroxylase deficiency are suggestive of ovarian virilizing tumor or ovarian hyperthecosis.[8] In young woman, serum 17-hydroxyprogesterone should be obtained to screen for late-onset congenital adrenal hyperplasia. Various other serum tumor markers (including AFP and CA-125) facilitate the differential diagnosis of ovarian adenocarcinoma.

Transabdominal and pelvic ultrasonogram is used for assessing the size and morphology of the ovary.[8] MRI is useful for defining tumor character. The size of the tumor rarely go above 10 cm. It is a solid tumor with areas of haemorrhage and necrosis. Microscopically these tumors comprise large polygonal cells, but nests cords and pseudo glandular and follicle like arrangements of tumour cells may also be seen. The tumour cells have abundant cytoplasm and vacuolated. The nuclei are round, with prominent central nucleolus, with nuclear atypia and

necrosis are sometimes present.[3] The absence of crystals of Reinke is helpful to distinguish SCT-NOS from leydig cell tumours. Also, the lack of spindle cells and fibromatous background is useful to differentiate it from luteinized thecoma pregnancy luteomas are one of the differential diagnoses wherein there is proliferation of lutein cells Nearly half of the cases present as bilateral tumor. Other differential diagnoses include stromal luteomas that are characterized by stromal hyperthecosisans 33 % cases have bilateral incidence.[9]

Immunohistochemistry evaluation shows, steroid cell tumours are typically positive for sex cord-stromal markers, such as inhibin, calretinin and melan-A, and are negative for FOXL2. Treatment of steroid cell tumour is primarily surgical. Treatment decision depends on many prognostic factors including the stage of the tumor, the presence of malignant features, age of the patient, and need for future fertility. In a young patient who want to preserve their fertility, unilateral salpingo-oophorectomy is the preferred method of treatment.[10]

In postmenopausal women the appropriate option is hysterectomy with bilateral salpingo- oophorectomy. Malignant NOS steroid cell tumours should be managed with surgical removal followed by a combination of chemotherapy and radiotherapy.[7] The adjuvant chemotherapy regimens currently recommended for treatment are as follows, BEP, cisplatin, doxorubicin, and cyclophosphamide, taxane and platinum and bleomycin, vinblastine, and cisplatin.[11]

Gonadotropin-releasing hormone agonist could be used as postoperative adjuvant therapy. Regular follow-up evaluation with measurement of serum testosterone level is mandatory. Patients with large ovarian tumors and with an advanced stage have worse prognosis.

CONCLUSION

All cases of hirsutism and features of virilization need to be evaluated in a systematic manner. This tumor belongs to one among the rare ovarian tumours. With prompt diagnosis and proper treatment, the patient improved symptomatically postoperatively and she conceive in immediate cycle.

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