Case Report

A Rare Case of Steroid Cell Tumor, Not Otherwise Specified (NOS), of the Ovary in a Young Woman

Eek Chaw Tan, Chit Chong Khong, and Kazila Bhutia
Division of Obstetrics & Gynaecology, KK Women’s and Children’s Hospital, Singapore
Correspondence should be addressed to Kazila Bhutia; Kazila.Bhutia@kkh.com.sg
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Steroid cell tumour is a rare sex cord-stromal tumor of the ovary. It may produce steroids and is associated with testosterone secretion which causes symptoms like hair loss, hirsutism, and oligomenorrhea/amenorrhea due to hormonal activity and virilizing properties of tumor. In this article, we reported a 27-year-old woman who presented with hirsutism, hoarseness of voice, scalp hair fall, and amenorrhea for 8 years. Clinical and diagnostic evaluation revealed a left adnexal mass and elevated serum levels of testosterone and she was diagnosed as having a Sertoli-Leydig cell tumour of ovary. She underwent left salpingooophorectomy and both histopathologic examination and immunohistochemistry confirmed the diagnosis. Her serum testosterone levels normalized 3 days after the surgery and her menses resumed spontaneously a few months after the operation. In addition, we reviewed the literature on the epidemiology, clinical presentations, imaging and histological findings, and the treatment options on this disease.

1. Introduction

Steroid cell tumour (SCT) accounts for less than 0.1% of all ovarian neoplasm [1]. It is classified into 3 categories based on the cell origin: stromal luteoma, Leydig cell tumour, and SCT not otherwise specified (NOS) [2, 3]. Steroid cell tumour may produce steroids and is associated with testosterone secretion which causes symptoms such as hair loss, hirsutism, and oligomenorrhea/amenorrhea due to hormonal activity and virilizing properties of tumor [4]. In this report, we present a case of steroid cell tumour, NOS patient who presented with amenorrhea and symptoms of virilization.

2. Case

A 27-year-old single virgin lady presented to our clinic with hirsutism, hoarseness of voice, scalp hair fall, and amenorrhea for the last 8 years. She had menarche at the age of 10 and menses were regular until age of 16 years. Her menses became irregular after the age of 16 and was amenorrheic by 19 years old. She also complained of gradual weight gain of 10 kg over the last two years to her current weight of 107 kg.

The physical examination revealed an androgenic alopecia, acanthosis nigricans, hirsutism (Ferriman Galway Score 27), and clitoromegaly. Her BMI was 43.4 kg/m².

She did not have withdrawal bleed following progesterone challenge with oral medroxyprogesterone tablets for 7 days. The blood tests showed normal serum prolactin, estradiol level, free thyroxin, and thyroid stimulating hormone. However, serum total testosterone level was elevated at 22.5 nmol/L (normal range 0.4 – 2nmol/L) and serum LH and FSH were low with levels of 0.1 IU/L. In addition, both DHEA-S (dehydroepiandrosterone sulfate) and 17-hydroxy progesterone level were normal which made adrenal source unlikely.

The transvaginal sonography showed endometrial thickness of 3 mm with polycystic ovaries containing multiple avascular cysts (Figure 1). Hence, based on initial work-up, differential diagnosis of Polycystic Ovarian Syndrome was made. MRI of abdomen and pelvis was arranged and it revealed a 8.4 x 6.1x 8.9 cm predominantly solid enhancing mass arising from the left ovary. There was no evidence of adrenal mass or abdominal or pelvic lymphadenopathy (Figure 2). Blood test for ovarian tumour markers including
beta HCG, Ca 125, Ca 19-9, chorioembryonic antigen, and alpha-fetoprotein were normal.

In view of the raised serum testosterone level and MRI of pelvis findings, the initial impression was a Sertoli Leydig cell tumour of the ovary.

3. Treatment

The patient underwent an open laparotomy and left salpingooophorectomy. Intraoperatively, she was noted to have a 10 X 10 X 3 cm left ovarian mass (Figure 3). The uterus, bilateral fallopian tubes, and right ovary were normal. The frozen section of the left ovarian mass was reported as Sertoli Leydig cell tumor. The patient had unremarkable postoperative recovery and discharged well on the third postoperative day.

4. Outcome and Follow-Up

The final histology confirmed steroid cell tumour, not otherwise specified. Her serum testosterone levels normalized 3 days after the surgery and her menses had resumed spontaneously a few months after the operation. She is currently free of disease and is on regular follow-up with Gynaecological Cancer Centre.

5. Discussion

Steroid cell tumour was first described by Scully to contribute for less than 0.1% of all ovarian neoplasms [1, 5]. 60% of SCT are NOS category [6] and more than 90% of the NOS are unilateral [1, 7, 8]. NOS tends to affect younger women (mean age: 43 years) [7] and 25-40% of NOS tumors are malignant [7].

NOS are associated with androgenic changes with variable frequency ranging from 12% to over 50% and they usually stay for many years [9–12]. They generally present with virilizing symptoms such as gradually progressive hirsutism, acne, deepening of voice, temporal baldness, and amenorrhea [13]. 25% of the patients with NOS remain asymptomatic [14].

In this case, the patient was presented with irregular menses after the age of 16 years and became amenorrhoeic by the age of 19 years. She also had virilizing signs of androgenic...
tumours by absence of Reinke’s crystals [25]. Nucleoli are seen [21–24]. It is differentiated from Leydig cells with distinctive borders and central and prominent appearance. In cytological examination, polygonal or round columns or cords like zona glomerulosa and zona fasciculata tumor has a nested arrangement but can be organized into occasional cystic changes [14, 19, 20]. Microscopically, the section appears as yellow-orange surface with scribed, solid and noncalcified with a lobulated appearance. The gross appearance of NOS generally is well circumcised, solid and noncalcified with a lobulated appearance. In cytological examination, the sections of the main tumor showed a predominantly diffuse proliferation of polygonal cells with ample, pale, vacuolated cytoplasm and round central nuclei. Small areas of a more nested appearance were also seen. Foci showing cells with more eosinophilic cytoplasm were also evident, but no Reinke crystals were evident. The nuclei were central and round, with no significant atypia. Immunohistochemical stains for calretinin and alpha-inhibin were diffusely and strongly positive. CD10 is negative. An area showing multiple cystic structures lined by bland cuboidal to columnar epithelium with focal ciliation in a fibromatous stroma was seen. No significant epithelial proliferation or atypia was seen.

Immunohistochemical markers for inhibin and calretinin are sensitive markers for steroid cell tumors NOS [26]. The sensitivity of positive calretinin is 60 to 90% whereas the sensitivity of inhibin reactivity ranges from 5 to 90% [27, 28]. Other markers such as EMA, cytokeratin, CD 99, and SI100 have been reported to be positive. HMB45, Chromogranin-A, LeuM1, AFP, carinoembryonic antigen (CEA), and periodic acid Schiff (PAS) are other markers which have been studied [29].

The recommended treatment of steroid cell tumor, NOS is primarily surgical. For patients with benign tumor confined to one ovary and family is completed, unilateral salpingooophorectomy should be recommended. For patients with unilateral malignant tumor desiring fertility, staging laparotomy with unilateral salpingooophorectomy with preservation of contralateral ovary and uterus is a reasonable option [30]. In postmenopausal patient or bilateral tumors, hysterectomy with bilateral salpingooophorectomy can be performed [22]. For metastatic cases, debulking surgery and chemotherapy or radiotherapy are recommended [13, 22]. In this case, the patient underwent laparotomy and left salpingooophorectomy. Serum testosterone levels normalized 3 days after the surgery and her menses resumed spontaneously a few months after the operation.

6. Conclusion
Steroid cell tumour, NOS is a rare tumour which can be onerous to diagnose. We highlight the importance of careful laboratory and imaging investigations with involvement of the multidisciplinary team in making the diagnosis. Any young patient presenting with severe virilisation and/or menstrual problems, neoplastic etiology should always be kept in mind.

Disclosure
This paper had been presented as a poster in the RCOG World Congress 2017, 20–22 March 2017, CTICC Cape Town, South Africa.

Conflicts of Interest
The authors declare that they have no conflicts of interest.

References


