

STEROID CELL TUMOR-NOS (NOT OTHERWISE SPECIFIED) OF THE OVARY: A CASE REPORT

Dr. Berna Dilbaz,¹ Dr. Burak Karadağ²

ÖZET

Steroid hücreli tümör-NOS (başka şekilde spesifiye edilemeyen) nadir görülen ovaryen seks kord stromal tümör grubundandır. Bu olgu sunumunda perimenopozal bir kadında hirsutizme neden olan steroid hücreli tümör-NOS sunulmuştur. 52 yaşındaki hasta kliniğimize hirsutizm yakınması ile başvurdu. Laboratuvar incelemesinde serbest testosteron düzeyi 0,84 pg/ml (normal değerleri 0,46-2,5), total testosteron düzeyi 0,91 (normal değerleri 0,06-0,82 ng/ml) idi. Transvajinal ultrasonografik incelemede sağ adneksiyal alanda 57*51 mm boyutlarında solid kitle görüldü. Laparoskopik sağ salpingo-oofektomi yapıldı. Patolojik inceleme sonucu steroid hücreli tümör-NOS olarak rapor edildi.

Anahtar Kelimeler: Steroid hücreli tümör, Hirsutizm, Ovaryen tümör

ABSTRACT

Steroid cell tumors-NOS (not otherwise specified), are rare ovarian sex cord-stromal tumors. We present a case of steroid cell-NOS, as a cause of hirsutism in a perimenopausal patient. A 52 year old patient was admitted to our clinic with hirsutism. Laboratory investigations showed a free testosterone level of 0,84 pg/ml (normal value 0,46-2,5 pg/ml), a serum total testosterone of 0,91 ng/ml (normal value: 0,06 – 0,82 ng/ml). Transvaginal examination revealed a 57x51 mm solid mass in the right adnexal area. She underwent laparoscopic right salpingo-oophorectomy. Gross pathologically and microscopically investigations showed the appearances of steroid cell tumor-NOS, a benign entity.

Key Words: Steroid cell tumor, Hirsutism, Ovarian tumor.

INTRODUCTION

Steroid cell tumors, not otherwise specified (NOS), are rare ovarian sex cord-stromal tumors. These tumors usually present in premenopausal women with symptoms of androgen excess, manifested by hirsutism and elevated testosterone levels (1). Steroid cell tumors of the ovary account for less than 0.1% of all ovarian tumors (1). Due to the rarity of these tumors, little is known about their metastatic behavior and malignant potential. We present a case of steroid cell,-NOS, as a rare cause of hirsutism in a perimenopausal patient.

CASE

A 52 year old gravida 11 para 6 patient was admitted to our clinic with hirsutism over the upper lip,

chin, temporal region, upper neck, upper and lower abdomen, thighs and perineum for approximately two years. Patient's Ferriman-Gallwey score was 15 (2). No cliteromegaly was detected. A pelvic mass of approximately 5cms was detected in the right adnexal area. Other systems examination was normal.

Laboratory work-up showed a free testosterone level of 0,84 pg/ml (normal value 0,46-2,5 pg/ml), a serum total testosterone of 0,91 ng/ml (normal value: 0,06-0,82 ng/ml) and serum 17-Hydroxyprogesterone, cortisol, thyroid hormone levels, and all other blood parameters and tumor markers were within normal limits (Table 1).

A transvaginal examination showed a 57x51 mm solid mass in the right adnexal area (Figure 1). In Doppler study pulsatility index and resistance index of the mass was found 0,68 and 0,51 respectively. She

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¹Etlik Zübeyde Hanım Women's Health Teaching and Research Hospital, ANKARA

²T.C.S.B. Erbaa Devlet Hastanesi Kadın Hastalıkları ve Doğum Kliniği TOKAT

İletişim: Dr. Burak Karadağ

T.C.S.B. Erbaa Devlet Hastanesi Kadın Hastalıkları ve Doğum Kliniği TOKAT

Tel: 0505 586 38 56

E-Posta: drburakkaradag@gmail.com

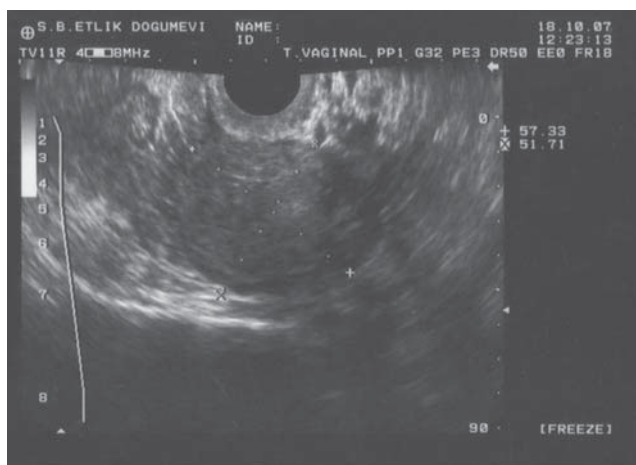


Figure 1—A transvaginal examination showed a 57x51 mm solid mass in the right adnexal area.

underwent laparoscopy. During laparoscopic evaluation a cyst of 6.5x5 cm arising from the right ovary was observed, otherwise the pelvic and abdominal organs observed were normal. A peritoneal washing was taken for cytological evaluation and right salpingoophorectomy was preformed. Frozen section evaluation at the time of the surgery examination was reported as benign. The operation and postoperative recovery we uneventful. Final histopathological evaluation of the specimen that was defined during gross examination as yellow-orange solid mass 65mmx55mm in diameter with intact capsule was reported as a benign ovarian steroid cell tumor-NOS. No necrosis, hemorrhage or atypia was detected and one mitotic figure was counted in 10 high power field.

Table 1—Laboratory Findings

Parameters	Values
Hb (gr/dl)	14,4
Fasting blood sugar (mg/dl)	93
Estradiol (pg/ml)	22,68
Serum Cortisol (µg/dl)	15,2
DHEA-SO4 (µg/dl)	162,4
Total Testosterone (ng/ml)	0,91
Free Testosterone (pg/ml)	0,84
17 – hydroxyprogesterone (ng/ml)	0,66
Prolactin (ng/ml)	5,31
TSH (uIU/ml)	0,972
Ca 125 (IU/ml)	8,47

DISCUSSION

Steroid cell tumors was first described by Scully in 1979 and account for less than 0.1% of all ovarian tumors (1,3). There are three subtypes of steroid cell tumors: NOS, stromal luteomas, and Leydig cell tumors (4). Steroid cell tumors-NOS are the most common subtype within this group and typically present in younger reproductive-aged women (mean age 43) (4).

Hirsutism and virilization are the most common symptoms found in 56% to 77% of patients (5). These tumors typically are associated with androgenic excess, but estrogenic or cortisol overproduction (i.e. Cushing syndrome) also has been reported (5). Estradiol secretion found in 6% to 23% of patients, and 6% to 10% of patients have been associated with Cushing sendrome (5). However, 1/4 of these tumors may not produce any hormones (5).

Steroid cell tumors-NOS, typically are solid, usually benign and slow growing, and the symptoms are frequently present for many years before the diagnosis is made (6). A third of steroid cell tumors NOS are clinically malignant (7). Some pathological features indicate malignancy potential. Hayes et al. after evaluating 68 steroid cell tumors-NOS reported that the features that predict malignant behavior were grade 2 or 3 nuclear atypia (%62 malignant) hemorrhage (77% malignant), tumor size >7 cm (78% malignant), necrosis (86% malignant), two or more mitotic figures in 10 high power fields (92% of malignant) (5).

The treatment of resectable benign type is surgery while malignant steroid cell tumor type is similar to malignant epithelial type tumor of the ovary. Steroid cell-NOS tumors of the ovary are mostly unilateral, so removal of the contralateral ovary is not required if there is no involvement (8). The efficacy of chemotherapy in malignant cases is unknown because of the rarity of these tumors but regimes are similar to those used in malign epithelial and germ cell tumors (9).

In the reported patient blood total and free testosterone levels were not remarkably high, estradiol and other laboratory findings were normal. No additional treatment was not given.

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