

A rare cause of virilization; Ovarian steroid cell tumor, not otherwise specified (NOS)

Nadir rastlanılan virilizasyon sebebi; overyan steroid hücreli tümör

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Abstract

Sex cord-stromal tumors account for 5% of ovarian tumors and 2% of malignant ovarian tumors. Steroid cell tumors (SCT), not otherwise specified (NOS), are rare sex cord-stromal tumors of the ovary and account for less than 0.1% of all ovarian tumors. We report a rare case of a postmenopausal woman presented with hirsutism, virilism and with findings of hyperestrogenism. (J Turkish-German Gynecol Assoc 2012; 13: 275-7)

Key words: Hirsutism, steroid cell tumor, virilization, hyperandrogenism, sex cord-stromal tumor

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Özet

Sex-cord hücreli tümörler, tüm over tümörlerinin %5 'ini, malign over tümörlerinin %2'sini oluşturmaktadır. Steroid hücreli tümörler nadir sex cord -stromal tümörler olup, tüm over tümörlerinin %0.1'ini oluşturmaktadır. Biz hirsutizm, virilizasyon ve hiperöstrojenizm bulguları mevcut postmenapozal bir bayan olgusunu sunmak istedik.

Anahtar kelimeler: Steroid hücreli tümör, virilizasyon, hirsutizm, hiperandrojenizm, sex cord- stromal tümör

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Introduction

Sex cord-stromal tumors account for 5% of ovarian tumors and 2% of malignant ovarian tumors (1, 2). SCTs account for less than 0.1% of all ovarian tumors (3). The terms 'lipid cell tumor' and 'lipoid cell tumor' have been used to designate a group of morphologically similar ovarian neoplasms of diverse cellular origin. These tumors are composed exclusively of cells-i.e., lutein cells, leydig cells, and adrenal cortical cells. The use of the above terms is misleading, however, as some tumors in this category contain little or no lipid. In view of this inaccuracy, the term 'steroid cell tumor' is proposed for these neoplasms, which can be divided into several subtypes according to their cells of origin. The designation 'steroid cell tumor' is appropriate not only because of the morphological features of the neoplastic cells, but also because of their propensity to secrete a variety of steroid hormones that often produce characteristic clinical syndromes (3). We report a rare case of postmenopausal woman presenting with hirsutism, virilism and also findings of hyperestrogenism.

Case Reports

A 51-year-old, gravida 5, para 4, abortus 1 woman (age of onset of menopause 42 years) presented with rapidly pro-

gressing hirsutism, receding hairline, male-pattern baldness, alopecia and voice deepening (Figure 1). She was diagnosed with hypertension for six years and diabetes mellitus for seven years. She had had a laparoscopic cholecystectomy operation 3 years earlier and she was diagnosed for depression 2 years previously. Physical examination revealed hirsutism involving the face, chin, upper back, chest, upper and the lower abdomen giving a score of 44 from modified Ferriman and Gallwey scoring system (Figure 2). Gynecologic examination revealed cliteromegaly which had developed over the past 6 years. Pelvic ultrasound-scan revealed a solid ovarian tumor of 35x36 mm in the left ovary. Markedly elevated serum testosterone level (8.3 ng/mL), elevated serum estradiol level (85.86 pg/mL) and suppressed gonadotrophin levels (FSH: 0.606 mIU/mL, LH<0.01 mIU/mL) were observed. Dehydroepiandrosteronesulfate level was normal (131.4 µg/dL). The levels of tumor markers were normal. Computed tomography and magnetic resonance imaging of the abdomen and pelvis revealed a 40x25mm solid tumor in the left adnexia. Adrenal glands were normal. The patient underwent total abdominal hysterectomy bilateral salpingo-oophorectomy. Frozen section of the left ovary revealed thecoma. Gross pathological examination of the left ovary revealed an 3x2.2 cm well-circumscribed, yellow-orange mass. In the uterine cavity a 0.6x0.8 mm endometrial polyp was observed.

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Figure 1. Findings of virilism. Male pattern baldness



Figure 2. Hirsutism involving the face, chin, upper back, chest, upper and the lower abdomen giving a score of 44 from modified Ferriman and Gallwey scoring system

Microscopic examination revealed steroid cell tumor, not otherwise specified for the mass in the left ovary and endometrial proliferative findings with endometrial polyp for the uterus. Total testosterone level was normal on the postoperative first month.

Discussion

Ovarian SCTs account for 0.1-0.2% of all ovarian tumors, and usually present with the findings of virilization (3, 4). There are three subtypes: stromal luteoma, Leydig-cell tumor, and steroid cell tumor, not otherwise specified (NOS). Steroid cell tumors, NOS, must be distinguished from other tumors in the steroid cell category -luteinized thecomas, pregnancy luteomas and carcinomas, both primary clear cell carcinoma and metastatic renal cell carcinoma. Both the hilus cell tumor and the rare Leydig cell tumor, nonhilus type, can be identified with certainty

only by demonstrating the presence of crystals of Reinke in the cytoplasm of the neoplastic cells (5-8). Since testicular Leydig cell tumors lack these inclusions in 60-65% of cases (9, 10), an unknown proportion of tumors in the steroid cell tumor, NOS, category are almost certainly Leydig cell tumors in which crystals have not been identified. The luteinized thecoma can be identified by the presence of a predominant spindle cell background. It is possible, however, that the steroid cell tumor, NOS, is a fully luteinized thecoma (11), since some luteinized thecomas show extensive luteinization (8, 12) and a rare steroid cell tumor, NOS, contains small areas of spindle cell proliferation (13). The focal presence of nonluteinized granulosa cells in a predominantly luteinized granulosa cell tumor helps to distinguish it from a steroid cell tumor. Electron-microscopical examination of the tumor may be of additional help in distinguishing a steroid cell tumor from a clear-cell carcinoma by demonstrating the typical abundant smooth endoplasmic reticulum in the cytoplasm of the neoplastic steroid cells (14). Steroid cell tumor, not otherwise specified, accounts for 60% of SCTs, 25-45% of which are clinically malignant. This subtype is associated with androgenic changes. SCTs often present as unilateral solid tumors but the size of tumors may be as small as 2-3 cm, thus it would be difficult to diagnose. Clinical and laboratory findings are usually exaggerated according to its dimension. Several medications such as oral contraceptives, cyproteroneacetate and spironolactone were prescribed for the presented case for hirsutism for 2 years. Also laser hair removal treatment was performed. However, symptoms of virilization did not improve. Delayed diagnosis would be important for the tumors with malignant potential. Interestingly, pathologically benign tumors can behave in a clinically malignant fashion. Estradiol secretion by these tumors is not uncommon (6-23%) (15). The presented case had endometrial hyperplasia and polyp as a result of elevated estradiol level. Excess estrogen production can result in menorrhagia, postmenopausal bleeding and rarely adenocarcinoma. SCTs should be kept in mind for the patients presenting with virilization and high serum androgen level. Meticulous clinical evaluation should be carried out before initiation of medical therapy for such patients.

Conflict of interest

No conflict of interest was declared by the authors.

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