Ovarian hemangioma: A rare cause of hirsutism in postmenopausal woman

Ovaryen hemangioma: Postmenapozal kadında hirsutizmin nadir bir nedeni

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Abstract

69-years-old postmenopausal woman was referred to our clinic with a solid ovarian tumor. On examination she had significant facial and dorsal hirsutism and especially male pattern pubic hair growth. While DHEA-S level was within a normal range, testosterone measurement was markedly increased. Ultrasonography showed a well-defined hyperechoic lesion of 2 cm diameter in the left ovary. MRI scans demonstrated adrenal glands without tumor. She underwent laparotomy for ovarian mass. Intraoperative frozen section examination of the left ovary revealed a 2 x 2 x 1 cm, well-circumscribed tumor on the cut surface and no evidence of malignancy. Histopathological findings were an ovarian hemangioma with stromal luteinization. The serum androgen levels reduced to a normal value after surgery. Furthermore, the clinical signs of hyperandrogenism improved significantly. Although its low incidence, it is a possible that must be considered in all postmenopausal women with signs of hirsutism.

Keywords: postmenopausal, ovarian hemangioma, hirsutism, stromal luteinization.

Özet

69 yaşında postmenopozal bir hasta solid ovaryan tümör nedeniyle kliniğimize refere edildi. Hastanın yüzünde ve sırtında belirgin kıllanma ve özellikle pubik bölgede erkek tipi kıllanma görüldü. DHEA-S seviyesi normal sınırda iken, testosteron ölçümü belirgin şekilde yüksekti. Ultrasonografide, sol overde 2 cm. çapında iyi sınırlı, hiperekojen kitle görüldü. MRI taraması adrenal glandlarda herhangi bir tümörün olmadığını ortaya çıkardı. Hastaya ovaryan kitle tanısıyla laparatomi yapıldı. Sol overin operasyon sırasında frozen incelemesi 2 x 2 x 1 cm. boyutlarında, malignite belirtisi olmayan ve kesi yüzeyi düzgün sınırlı bir tümörü ortaya çıkardı. Histopatolojik bulgular stromal luteinizasyon ile birlikte bir ovarian hemangiomayı gösteriyordu. Cerrahi sonrasında hastanın serum androjen seviyeleri normal değerlere geriledi. Dahası, hiperandrojenizmin klinik bulguları da önemli derecede düzeldi. Ovaryan hemangioma nadir görülen bir hastalık olmasına rağmen, hirsutizm bulguları gösteren postmenopozal tüm kadınlarda bir olasılık olarak göz önünde bulundurulmalıdır.

Anahtar kelimeler: postmenopozal, ovaryan hemangioma, hirsutizm, stromal luteinizasyon

Introduction

Hemangiomas are benign vascular tumors that are extremely rare found in the ovaries. Cavernous type of ovarian hemangiomas may present as isolated ovarian masses (1,2), which are discovered incidentally or with diffuse abdominopelvic hemangiomatosis leading to abdominal and pelvic symptoms (3). Ovarian hemangiomas may rarely be associated with systemic manifestations. It is commonly nonfunctional neoplasms. However, it is well known that luteinization of stromal cells commonly occurs as a relative phenomenon. Luteinization of ovarian stromal cells may result in androgenic effects, estrogenic effects, or both (4,5). Herein we report a case of a cavernous hemangioma in a 69-years-old woman presenting with high androgen levels, and hirsutism.

Case report

A 69-years-old postmenopausal woman (gravida 5, parity 3) was referred to our clinic, due to a solid well-circumscribed ovarian mass detected by abdominopelvic ultrasonography. The patient had been menopausal for 18 years. She had never used hormone replacement therapy or any medical treatment. On physical examination, the patient had significantly dorsal hirsutism and male pattern pubic hair growth for many years. She have also not systemic multiple hemangiomatosis.

All routine laboratory values including CA-125 (carbohydrate antigen 125), CA15-3, and CEA (carcinoembriogenic antigen) levels were within normal limits. Preoperative hormone levels were as follows: total testosterone 295 ng/dl (normal range 15-80 ng/dl), free testosterone 4.5 pg/ml (normal range 0.29-3.18 pg/ml), androstenodione 3.7 ng/ml (normal range 0.3-3.5 ng/ml), DHEA-S (dehydroepiandrosterone sulfate) 54 ng/dl (normal range 35-430 ng/dl), and estradiol 51 pg/ml (normal range 0-30 pg/ml in postmenopausal women). Transvaginal ultrasonography showed a normal uterus, right ovary with normal appearance, left ovary measuring 3 x 2 cm with a well-defined hyperechoic lesion measuring 2 x 2 cm. These findings were confirmed by abdominopelvic magnetic resonance imaging (MRI) (Fig. 1). MRI of the adrenal gland performed for elevated androgens levels was revealed any tumour. An ovarian source clinical and biochemical hyperandrogenism was suspected.

An exploratory laparotomy was performed. Abdominal and pelvic peritoneal washings were obtained. Total abdominal hysterectomy and bilateral salpingo-oopherectomy was performed. Intraoperative frozen-section examination revealed that on the cut surface of the left ovary (3 x 2 x 1.5) contained a 2 x 2 x 1 cm, well-circumscribed, maroon colored tumor (Fig. 2). Intraoperative

Figure. 1. Spin-echo T1-weighted axial MRI of the pelvis demonstrated a low-signal-intensity mass in the left ovary.

microscopic examination of this tumor reported an ovarian benign tumor. She made a satisfactory post-operative recovery. The final diagnosis of the left ovary was a benign cavernous hemangioma, consisting of multiple thin walled blood vessels, filled with red blood cells, and a single layer of flattened endothelium lined all of the vessels. There was no necrosis and mitotic activity. The surrounding ovarian stroma was contained luteinized stromal cells (Fig. 3). Immunohistochemical staining showed negative for estrogen and progesterone receptors.

Pre-operative elevated androgen levels were within normal value after surgery. At 6-months folow-up, the clinical signs of hyperandrogenism improved significantly.

Discussion

Vascular tumors are extremely rare in the ovary. Hemangiomas of the ovary are only approximately 50 cases reported in the literature to date. Ovarian hemangiomas have been reported in both adults and children without predominance in any decade (6). Histologically, ovarian hemangiomas may be cavernous or capillary. Most ovarian hemangiomas are of the cavernous type (1,2). Their size ranged from 0.3 to 24 cm in the largest diameter in previous reports (7). Hirsutism or other signs of hyperandrogenism are usually caused by ovarian or adrenal disorders. With all androgen-producing ovarian tumors, serum testosterone is elevated in conjunction with normal or mildly elevated serum DHEA-S level. The lack of a significant increase in DHEA-S level distinguishes ovarian from adrenal androgen-producing tumors. In this case, while DHEA-S levels were within a normal range, testosterone measurement were markedly increased, hence suggestion an ovarian origin. Ovarian causes of hyperandrogenisms including primary and metastatic ovarian neoplasms. However, in nonfunctional ovarian neoplasms, such as ovarian hemangiomas, association with hirsutism has been rare.

Ovarian hemangiomas are commonly nonfunctional tumors. However, nonendocrine benign and malign ovarian neoplasms can cause hormonal effects (1,4). Functioning ovarian stroma with luteinized stromal cells may be presented in the adjacent or surrounding ovarian tissue of hemangiomas. Nevertheless, in nonepithelial neoplasms such as our case, the association with stromal luteinization has been rare (6). A search of MEDLINE (English language; 1991-2008; search terms: "ovarian hemangioma", "stromal luteinization") revealed only 7 cases. These luteinized ovarian stromal cells produce androgens. They are subsequently converted to estrogens in the adipose tissue. The end result of this cascade may be estrogenic (breast tenderness, endometrial stimulation), androgenic (alopecia, virilization), or a combination of both (8). Our patient demonstrated hyperandrogenism both a clinically (hirsutism) and biochemically (elevated androgen levels).

There have been speculations in the literature regarding the pathogenesis of ovarian hemangiomas and their hormonal status. One hypothesis has been that the encouraging incident in the development of ovarian hemangiomas is hyperestrogenism resulting from stromal hyperplasia or hyperthecosis (4,9,10). It is based on that estrogens have known growth stimulatory effects on the vasculature and that most hemangiomas carry estrogen receptors. Another hypothesis is that the presence of an ovarian hemangioma is the primary event in the pathway leading to hyperandrogenism or hyperestrogenism. An expansible hemangioma has been suggested as inducing stromal luteinization by mass effect. Stromal luteinization cause by an expansible ovarian hemangioma is usually limited to the stroma of the neoplasm or to the ovarian stroma adjacent to neoplasm (8). In our case, stromal luteinization was markedly in the stroma adjacent to the tumor. Moreover, immunohistochemical staining was negative for estrogen and progesterone reseptors in the endothelial cells of the hemangioma. Absence of estrogen and progesterone receptors in the endothelial cells of the hemangioma suggested that ovarian hemangiomas may develop independent from their stimulation.

Consequently, ovarian hemangiomas are extremely rare, most are nonfunctional, and they require surgical treatment. However, in spite of its low incidence, it is a possible that must be considered in all postmenopausal women with signs of hirsutism.

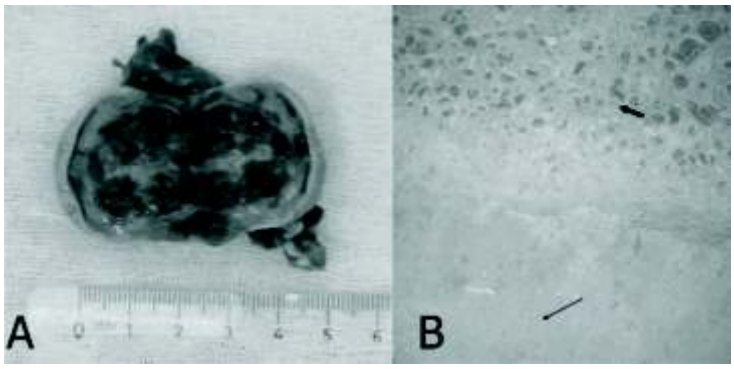

Figure. 2. A: Tumor is well circumscribed with brown appearance on sectioning. B: Luteinized stromal cells (thin arrow) and hemangioma (thick arrow) are shown (hematoxylin-eosin, x40).

Yazarlarla ilgili bildirilmesi gereken konular (Conflict of interest statement) : Yok (None)

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