CASE REPORT

Embryonal Carcinoma of the Ovary in a 55-year-old Woman

Başak **BAKSU**[1], İnci **DAVAS**[1], Atıf **AKYOL**[1], Sultan **ÇINAR**[1], Figen **EZEN**[1], Damlanur **SAKIZ**[2]

[1]*Şişli Etfal Training and Research Hospital, 2nd Gynecology and Obstetrics Clinic, Istanbul, Turkey*
[2]*Şişli Etfal Training and Research Hospital, Pathology Clinic, Istanbul, Turkey*

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Abstract

Germ cell tumors are the most common ovarian malignancies in females under the age of 21-years. However, they are very rare in older women. We present the eldest woman in the literature with an embryonal carcinoma. A 55-year-old woman complaining of irregular, heavy menses and pelvic pain was found to have a large pelvic mass and a positive pregnancy test. Subsequent investigation revealed a large right adnexal mass, and elevated β-hCG level. The pathological diagnosis of the mass identified at laparotomy turned out to be an embryonal carcinoma of the ovary. Although rare, the diagnosis of germ cell tumor should be considered in the differential diagnosis of an adnexal mass in older women. Regardless of age, the treatment is similar; however, fertility-sparing treatment is not a priority in the older age group.

Keywords: germ cell tumour, embryonal carcinoma, female genitalia, malignancy, advanced age

Özet

Elli Beş Yaşındaki Bir Kadın Hastada Ovaryen Embriyonal Karsinom

Germ hücreli tümörler 21 yaşın altındaki kadın hastalarda en sık görülen over kanseridir. Ancak, yaşlı hastalarda çok enderdir. Bu yazıda, literatürdeki en yaşlı embriyonel karsinom olgusu bildirilmiştir. Elli beş yaşındaki kadın hasta, düzensiz âdet ve kasık ağrısı şikâyetleri ile başvurmuş, yapılan muayenesinde büyük bir pelvik kitle saptanmış olup gebelik testi pozitiftir. İleri incelemesinde büyük bir sağ adneksiyel kitle ve yüksek β-hCG düzeyleri saptanmıştır. Laparotomide çıkartılan piyesin patolojik incelemesi ovaryen embriyonal karsinom olarak gelmiştir. Çok ender olmasına rağmen, yaşlı kadınlarda adneksiyel kitle ayırıcı tanısında germ hücreli tümörler de dikkate alınmalıdır. Yaşa bakılmaksızın, tedavi şekli aynıdır; ancak yaşlı grupta fertilite koruyucu cerrrahi ön planda değildir.

Anahtar sözcükler: germ hücreli tümör, embriyonal karsinom, kadın genital sistemi, malignite, ileri yaş

Introduction

Germ cells of the ovary are considered to be derived from the primitive germ cells of the embryonic gonad. They comprise 20% of all ovarian neoplasms; however, they are the most common malignancy of the ovary under age 21 (1). Embryonal carcinoma (EC) is commonly encountered in the testis whereas it is very rare in the ovary, with only 26 pure tumors reported till today (2).

We present the oldest case with EC. The clinical course, diagnostic tests, and histopathological basis for differentiation of this tumor are discussed.

Corresponding Author: Dr. Başak Baksu
Nato Yolu, Doktorlar Sitesi, A9 Blok, D. 9
Çengelköy, İstanbul, Türkiye
GSM : +90 542 435 74 93
E-mail : basakbaksu@yahoo.com

Case

A 55-year-old woman, gravida 8 para 2 abortion 6, presented to our outpatient clinic with a 4-month history of heavy, irregular bleeding and right lower quadrant pain. Past medical and surgical history were unremarkable. Her examination revealed a normal appearing cervix and an irregular mass on the right adnexa, approximately 7.0-8.0 cm in size, that moved with the uterus. Uterus and adnexa could not be distinguished seperately from the mass. Uterus was deviated to the left and was 9-10 gestational weeks in size. Transvaginal ultrasonography revealed endometrial thickness: 0.98 cm, anteroposterior diameter: 6.5 cm, long axis: 12.1 cm, myometrium with heterogenous echogenicity, multiple myomas with the largest measuring 2.0 cm, left ovary size: 3.2x1.6 cm, a solid pelvic mass of 6.8x8.0 cm appearing to arise from the right adnexa with regular borders but could not be differentiated from the right border of the uterus. Pelvic doppler ultrasonography showed that the mass had a resistance index (RI): 0.37.

Additional laboratory tests including liver function tests, electrolytes, and complete blood count were within normal limits. Of the serum tumor markers, AFP, CEA, CA-125, CA 15-3, CA 19-9 were normal. Serum β-hCG level was elevated to 19 325 mIU/ml. Of the hormone levels, estradiol: 1449 pg/ml, FSH: 0.124 mIU/ml, LH: <0.100 mIU/ml. Other preoperative studies, including chest X-ray, abdominal-pelvic CT scan and Doppler ultrasound disclosed no evidence of metastases or ascites, and Papanicolaou cytology was negative. Endometrial curettage revealed endometrium with inactive glands under very strong progesterone effect. Colonoscopy showed only internal hemoroids. Abdominal-pelvic magnetic resonance showed a solid mass measuring 7.0x10.0x8.5 cm in size, on the right adnexa, just next to the right side of the uterine corpus where the borders in between the two could not be sometimes discriminated. On T1-weighted images, the mass was iso-intense compared to myometrium with hyperintense foci. On T2-weighted images, it was iso-intense compared to myometrium with cystic-necrotic hyperintense foci and showing intense contrast holding pattern. The mass was in close approximation to iliac chain, sigmoid colon and iliopsoas muscle; however, there was no sign of invasion. There were no pathological lymphatic nodes. Between the mass and the uterus, anteriorly placed was a thickened fallopian tube. There were multiple myomas, hypointense both on T1- and T2-weighted images.

During laparotomy, left ovary and fallopian tube were normal in appearance and uterus was 9-10 gestational weeks in size. A right-sided mass between the leaves of the ligamentum latum, 6.0-7.0 cm in size, leading towards pouch of douglas with solid and cystic components was observed. Omentum was normal looking. After sampling of peritoneal fluid, the mass was removed as intact, without rupture, hysterectomy and bilateral salphingo-oopherectomy, appendectomy, omentectomy and pelvic-paraaortic lymph node dissection was performed.

Histopathological examination revealed an ovary with intact capsule and with a tumoral mass forming glandular and papillary structures among widespread necrotic and hemorrhagic areas. There were syncytiotrophoblastic type giant cells in between. During immunohistochemical examination with cytokeratin, membraneous cytoplasmic staining in tumor cells were observed. Giant cells observed among the islands of tumor cells showed immunoreactivity with β-hCG. There was no staining with CD30, AFP, epithelial membrane antigen and placental alkaline phosphatase. The combination of the morphological and immunohistochemical characteristics were consistent with an embryonal carcinoma of the ovary. Since the tumor was confined to the right ovary, and peritoneal cytology and lymph nodes were negative, the patient was surgically confirmed as Stage IA (Figure 1 and 2).

The patient was placed on chemotherapy for the treatment of germ cell tumor of the ovary, consisting of BEP: bleomycin (30 units iv) weekly for 9 weeks, and etoposide (100 mg/m^2/day) and cis-platinum (20 mg/m^2/day), both for 5 days, repeated every 21 days for 3 courses. β-hCG level returned to normal. One month after the operation, β-hCG level was 4.42 mIU/ml.

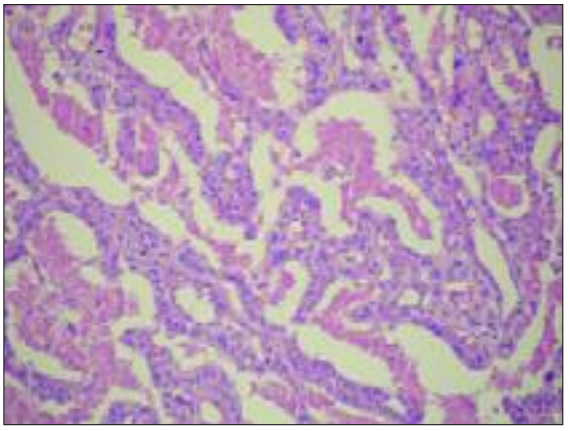

Figure 1. Tumor cells forming glandular and papillary structures (HE, x100)

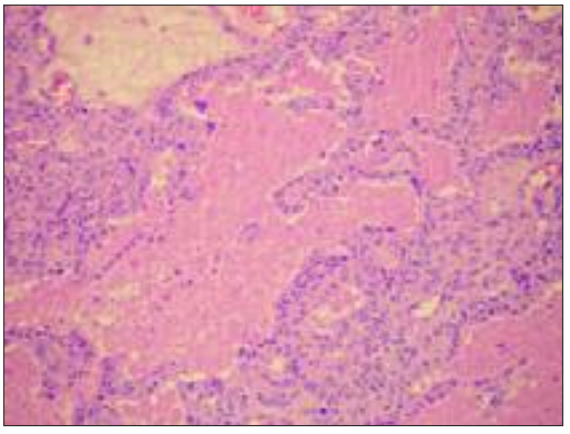

Figure 2. Tumor tissue and syncytiotrophoblastic giant cells among widespread necrotic areas (HE, x100)

Discussion

Ovarian germ cell tumors are special types of ovarian cancers due to their histology, age of presentation, prognosis and treatment. The percentage of germ cell tumors decline with increasing age (3). The patients usually present with abdominal pain, palpable mass, abdominal distention and/or menstrual irregularities (4). Our case presented with irregular, heavy bleeding and abdominal pain. The majority of these tumors are confined to one ovary (Stage I) (4). The case presented here was also Stage I. Diagnosis depends mostly on age, symptoms, size and consistency of the tumor and tumor markers (4). In this case, the woman was 55 years old with pelvic mass of 6.0-7.0 cm in size, producing β-hCG. Since these tumors continue to be diagnostically challenging, their correct diagnosis has major important therapeutic and prognostic implications.

On the other hand, most patients with epithelial ovarian tumors are postmenopausal and present with clinical ascites, dyspepsia and at laparotomy with peritoneal spread (Stage III) (1).

Embryonal carcinoma was first described as a separate germ cell tumor by Kurman and Norris (5). Embryonal carcinomas, probably arising from the primordial germ cell with little differentiation toward embryonic or extraembryonic tissue, are rarely pure, and usually in association with other constituent mixtures (6). The cells can produce both AFP and β-hCG, and may contain giant or syncytiotrophoblastic cells (6). In this case, there were syncytiotrophoblastic type giant cells among tumor cell islands. The cells staining positive for cytokeratin, and hyaline bodies may be present (6).

Like all germ cell tumors, ECS are encountered in children and young women, mostly under age of 30 years (1,6). The oldest reported women with EC is 53 years old (2). To our knowledge, this case is the oldest patient to date with EC.

The treatment of EC is unilateral oopherectomy followed by combination chemotherapy with BEP (4,7). However, ovarian preservation is not recommended in the elderly patient group (8). Since our patient was 55-years-old, we perfomed hysterectomy and bilateral salphingo-oopherectomy. Radiation does not seem to be useful for primary treatment (7).

Survival in germ cell tumors is associated with prognostic factors like histological type, clinical staging operation, lymph node and residual tumor (4). Overall survival for all stages of EC is reported as 39%, and for Stage I as 50% (9). In general, the survival rates for older women with germ cell tumors were reported to be lower than the younger population (10).

Although rare, the diagnosis of germ cell tumor should be considered in the differential diagnosis of an adnexal mass in older women. Regardless of age, the treatment is similar; however, fertility-sparing treatment is not a priority in the older age group.

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