TÜRK JİNEKOLOJİK ONKOLOJİ DERGİSİ
2016-3, Sayfa 113-116

PERİTONEAL YAYILIM GÖSTEREN VE KOLON KANSERİNİ TAKLİT EDEN REKTOSİGMOİD KİTLE; GRANÜLOZA HÜCRELİ TÜMÖR

GRANULOSA CELL TUMOR WITH PERITONEAL DİSSEMINATION AND RECTOSIGMOID MASS MIMICKING COLON CANCER

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ÖZET

Granüloza hücreli tümör; overin seks-kord stromal tümörlerinden en sık görülenidir. Yetişkin ve jüvenil olmak üzere iki tipi vardır. Yetişkin tip tümör genellikle adneksiyal kitle ve tümörün hormon salgılamasına bağlı vajinal kanama ile erken dönemde teşhis edilir. Burada karında kitle ve rektal kanama ile hastanemize başvuran 42 yaşındaki postmenopozal olguyu sunduk. Eksploratif laparotomide yaklaşık 20 cm boyutunda mezenterik kitle ile karaciğer metastazı ve çevre dokulara yayılmış implantlar saptandı. Eksize edilen kitlenin histopatolojik incelemesinde yetişkin tipi granüloza hücreli tümör özellikleri görüldü.

Anahtar Kelimeler: Ekstraovaryan, granüloza hücreli, over tümörü

ABSTRACT

Granulosa cell tumor is the most common sex-cord stromal tumor of the ovary. There are two types of granulosa cell tumor, which were defined as juvenile and adult types. Generally, the adult type tumor is diagnosed at early stages with adnexal mass and vaginal bleeding due to hormone secretion from the tumor. We report a 42 year-old postmenopausal woman admitted to our hospital with an abdominal mass and rectal bleeding. Exploratory laparotomy revealed a large mesenteric mass measuring 20 cm with spread implants and liver metastasis. Histopathological examination of the excised mass showed features of adult-type granulosa cell tumor.

Key Words: Extraovarian, granulosa cell, ovarian tumors

Geliş Tarihi: 30/10/2016 Kabul Tarihi: 10/05/2017

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INTRODUCTION

Granulosa cell tumors (GCT) are the most common malignant sex cord—stromal tumors of the ovary [1]. They can also arise in locations other than the ovary and may be derived from the mesenchyme of the genital ridge [2]. Women who have undergone oophorectomy may have the potential to develop GCT [2]. The primary extraovarian GCT is extremely rare tumor [3]. In the literature only 15 cases have been reported to date [3-8].

There are two types of granulosa cell tumor, which are juvenile and adult types. Adult type granulosa cell tumor is mainly seen at perimenopausal period and the main symptoms are vaginal bleeding and pelvic pain. Since the tumor secrete estrogen, endometrial glands proliferate and hyperplasia or endometrial malignancy may co-exist. Approximately, 85% of the patients were diagnosed at Stage I and overall 5 and 10-year survival were 93% and 90%, respectively (9).

Here, we report a rare, advanced granulosa cell tumor presenting with an extra-ovarian mass, mimicking colon cancer.

CASE

A 42 year-old postmenopausal woman, parity 3, with hypertension and diabetes admitted to our hospital with rectal bleeding. An abdominal mass was detected during examinations. Nine years ago she had undergone left oopherectomy. The excised ovary was histologically examined as non-neoplastic cyst. Menstruating had ceased 3 years ago. When she was admitted to the hospital her hemoglobin was 8 g/dL. Sonography of abdomen showed ascites and a 16x13cm sized solid mass in left adnexal area. Using computed tomography (CT), we observed left adnexal mass mainly associated with the mesentery of the sigmoid colon, with peritoneal/omental implants and liver metastasis (Figure 1). Liver metastasis was observed

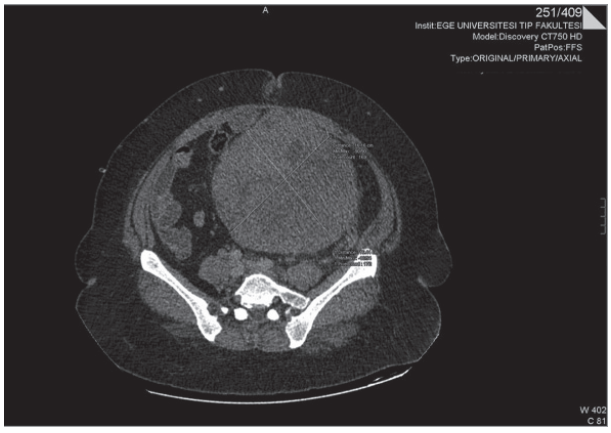

Figure 1 • Computer tomography image of the mass.

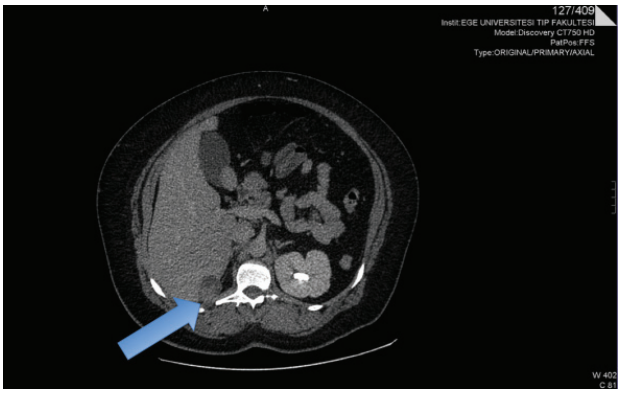

Figure 2 • Computer tomography image of metastatic subcapsular lesion on liver segment 6 (Arrow)

in segment 6 in the subcapsular space (Figure 2). The levels of serum tumor markers (CEA, CA 19-9, CA125, CA15-3) were in normal range.

Endoscopy and colonoscopy were performed. Internal hemorrhoids and varicose veins were observed in colonoscopy. Endoscopy was reported as normal.

Exploratory laparotomy revealed a mass, measuring 20 cm, at descending colon and sigmoid colon junction level originating from mesentery of the colon. Probably depend on previous operations left ovary was not observed. Implants were seen on mesentery of the small intestine, appendix and right ovary. During the operation, biopsy was sent for frozen section pathology. The frozen section result indicated a mesenchymal malignant tumor with atypia and increased mitosis. In the operation, the patient was thought as a colon tumor and peri-operative consultation was requested from general surgeons. The patient underwent right salphingo-oophorectomy, appendectomy, left hemicolectomy with sigmoid colon tumor excision. Suboptimal cytoreduction could be performed because of disseminated implants on the mesentery of the small intestine and peritoneum.

Final pathological examination revealed granulosa-theca cell tumor (24 mitotic figures under 10 high power fields) and granulosa cell tumor metastasis in the appendix, peritoneum, omentum, intestinal mesentery and right ovary (Figure 3). The tumor on the right ovary was 1,5 cm. The 20 cm mass in the sigmoid mesentery was totally composed of tumor tissue. The main characteristics of GCT; presence of Call-Exner bodies as microfollicular structures on microscopy, and immunohistochemistry results showing positive CD56, inhibin-α and S-100 staining, but negative CK19 staining were seen in our case.

As a result of this report, tumor board decision was adjuvant chemotherapy with EP regimen (etoposid, cisplatin) for the patient (etoposid 100 mg/m^2 on day

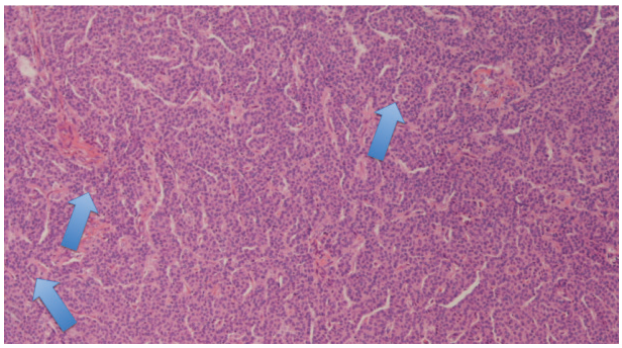

Figure 3 • Microscopic evaluation of granulosa cell tumor (x40 HPF), (Arrow: Call-Exner bodies)

1-2-3; cisplatin 35mg/m^2 on day 1-2-3 every 21days). She received first dose of the chemotherapy 6 weeks after the operation. PET-CT scans taken after the fourth dose chemotherapy shows the progression of the lesion in the liver segment 6. Control tumor markers' results (CEA, CA 19-9, CA125, CA15-3) were in normal range.

After the sixth cycle, metastatic lesion on the liver segment 6 had still persisted, therefore it was decided for secondary cytoreductive surgery in the tumor board. In the second operation, subcapsular implant about 5 cm size located at segment 6 was totally excised. Hysterectomy was performed. Colostomy was closed and the colonic passage was reassured with the anastomosis. The peritoneal implants were near-totally regressed after the chemotherapy.

In this period, the patient is continuing chemotherapy without any residual or recurrent disease.

DISCUSSION

Granulosa cell tumor (GCT) of the ovary is a rare tumor representing 2–5% of all ovarian neoplasms and 70 % of sex-cord stromal tumors [11-12]. They occur more often in menopausal and postmenopausal women with a peak age incidence between 50 and 55 years, but they may be seen at any age. They are the most common ovarian tumors with estrogenic manifestations. Granulosa-theca cell tumors have clinical importance for two reasons: 1. Their potential elaboration of large amounts of estrogen, and 2. The small but distinct hazard of malignancy in the granulosa cell forms. Due to the estrogen production 25-50% of these tumors associated with endometrial hyperplasia and 5-13% of these associated with endometrial carcinoma. Slow growth and late recurrence are main characteristics of these tumors. Rarely, GCT can develop at an extra-ovarian site, even in an oophorectomized patient [3]. Clinically, this impacts the evaluation of the oophorectomized patient with a pelvic mass. Extraovarian granulosa cell tumor can develop in retroperitoneum [4,10], broad ligament [5], mesentery, omentum, liver, adrenals, and so forth [4]. Histogenetic origin is thought to be from ectopic gonadal stromal tissue from the mesonephros [4]. In our case, there are three possibilities about the origin of the large mass located on the sigmoid colon mesentery. The first one is that, it can be a metastatic implant of the tumor located on the right ovary. This option is not realistic because the tumor on the right ovary is 1.5 cm, but the mass on the sigmoid mesentery was 20 cm. The second probability may be that, it can be originated from remnant of the left ovary that was excised many years ago. The third possibility is that, it may be extra-ovarian granulosa tumor that was originated from ectopic gonadal stromal tissue from the mesonephros.

Our case was typically presented as a colon tumor with the mass at the mesentery, peritoneal dissemination, and ascites. The left ovary was already excised and there were only a small implant on the right ovary like a Krukenberg tumor.

Hepatic metastases rarely occur, with an incidence of 1-2% of all metastatic GCT [13]. The occurrence of these metastases in only one segment is also rare, as they are almost always large in size and occupy a wide region of the liver parenchyma [14]. In our case liver metastasis was observed only in the segment 6 in the subcapsular space.

The case is reported for its rarity and to describe its importance in clinical practice. Women with oophorectomy may develop extraovarian GCT. Diagnosis is made by characteristic histologic features and by excluding previous GCT of ovary.

Conflict of Interest: The authors declare that there is no conflict of interest.

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