Malignant Mixed Mullerian Tumor with Heterologous Component of the Fallopian Tube: Case Report

Fallop Tüpünün Heterolog Bileşenli Malign Miks Müllerian Tümörü

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ABSTRACT Malignant mixed Mullerian tumors of the fallopian tube are rarely seen neoplasms. A 58 years old, gravida 2, para 2 postmenopausal woman was admitted to our clinic, complaining about pelvic pain. In the ultrasound examination of the patient, left adnexal mass measuring 66 x 35 mm in diameter with solid and cystic components were disclosed and CA-125 levels of the patient were elevated. With a presumptive diagnosis of ovarian neoplasm, the patient underwent an exploratory laparotomy. When undifferentiated carcinoma originating from fallopian tube was detected with frozen examination, extensive staging laparotomy was preferred. Postoperatively this pelvic mass was described as malignant mixed Mullerian tumor of the fallopian tube with heterologous component. We concluded that malignant mixed Mullerian tumor of the fallopian tube should be consider in differential diagnosis of pelvic mass in postmenopausal women who were presented with the symptoms, elevated CA-125 levels and ultrasonographic appearance.

Key Words: Fallopian tube neoplasms; adnexal diseases

ÖZET Fallop tüpünün malign miks Müllerian tümörü nadir görülen neoplazmdır. Elli sekiz yaşında, gravida 2, para 2 postmenopozal kadın ağrı şikâyetiyle kliniğimize başvurmuştur. Hastanın ultrasound muayenesinde 66 x 35 mm çapında ölçülen solid ve kistik bileşenlerden oluşan sol adneksiyal kitle gösterilmiştir ve hastanın CA-125 seviyeleri yüksektir. Ovariyan neoplazm ön tanısıyla hastada eksploratif laparotomiye gidilmiştir. Fallop tüpünden kaynaklanan farklılaşma göstermeyen karsinom frozen çalışması ile tespit edildiğinde, geniş evreleme laparotomi tercih edilmiştir. Operasyon sonrası bu pelvik kitle, heterolog bileşenli fallop tüpünün malign miks Müllerian tümörü olarak tanımlanmıştır. Fallop tüpünün malign miks Müllerian tümörü semptomlarıyla, artmış CA-125 seviyeleri ve ultrasound görünümü ile sunulan postmenopozal kadınlardaki pelvik kitlenin ayırıcı tanısında göz önünde tutulabileceği sonucuna vardık.

Anahtar Kelimeler: Fallop tüpü neoplazmları; adneksiyal hastalıklar

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Malignant mixed Mullerian tumors (MMMTs) of the fallopian tube are very rare neoplasms.[1] The sarcomatous component of MMMT exhibits the appearance of fibrosarcoma, high grade endometrial stromal sarcoma, chondrosarcoma, rhabdomyosarcoma, osteosarcoma or liposarcoma.[2] We present such an unusual case with a pelvic mass that was postoperatively described as MMMT of the fallopian tube with heterologous component and successfully managed.

CASE REPORT

A 58 years old woman, gravida 2, para 2 postmenopausal woman was admitted to our clinic, complaining about pelvic pain. Her past medical, gynecological, and surgical histories were unremarkable. On physical examination there was not found any abnormalities. Ultrasound exam disclosed left adnexal mass measuring 66 x 35 mm in diameter with solid and cystic components (Figure 1). The right ovary and uterus appeared normal on vaginal ultrasonography. Laboratory studies showed elevated levels of CA-125 (187.3 U/mL; normal <35 U/mL) and normal levels of CA-19.9 (14.2 U/mL), carcino embryogenic antigen (CEA) (0.27 ng/mL) and alpha-fetoprotein (18.13 ng/mL).

With a presumptive diagnosis of ovarian neoplasm, the patient underwent an exploratory laparotomy. At surgery, the main tumor was located in the left fallopian tube measuring approximately 5 cm in diameter and was adhered to left ovary and uterus. The other ovary and fallopian tube as well as other intra-abdominal organs appeared normal on inspection and palpation. Additionally, there was not found any evidence about salpingitis during inspection. The patient underwent to a total abdominal hysterectomy (TAH) and bilateral salpingo-oophorectomy (BSO). Undifferentiated carcinoma originating from left fallopian tube was detected with frozen examination. Therefore total omentectomy, appendectomy, and bilateral pelvic and paraaortic lymph node excision and cytological sampling were performed.

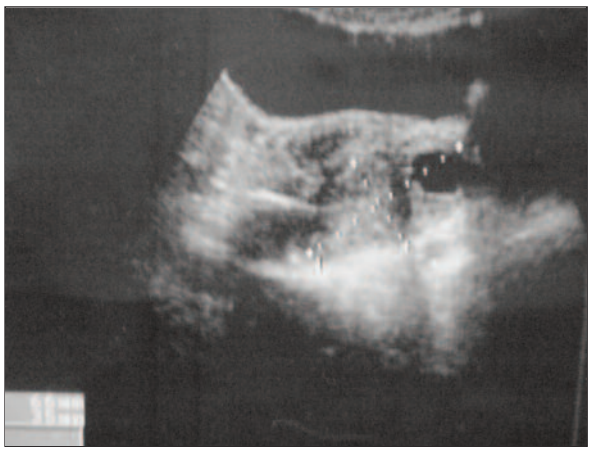


FIGURE 1: Ultrasonographic view of the pelvic mass.

Macroscopically, the mass lesion predominantly in the left fallopian tube and the left ovary was minimally involved. The cut surface showed whitish color solid areas with cystic, hemorrhagic components.

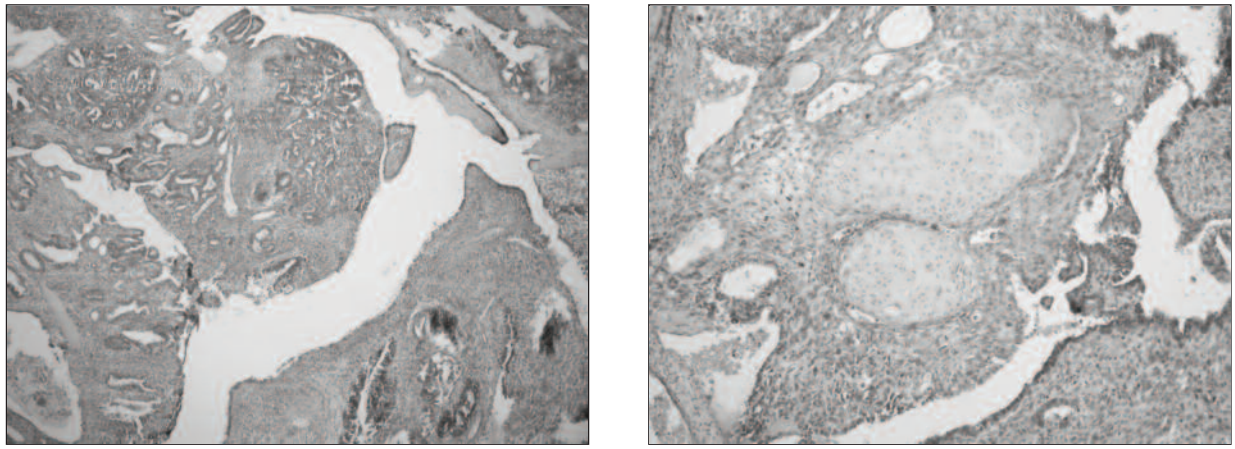
Microscopically, the carcinoma was consisted both adenocarcinoma and dominantly undifferentiated sarcoma. The sarcomatous components of these tumors may contain generally undifferentiated malignant mesenchymal elements with combination of cartilaginous differentiation. Immunohistochemical examinations revealed that the undifferentiated sarcoma cells were positive for vimentin but they were negative for actin, desmin, pan keratin and myoglobin.

Final histopathological examination of the mass was the primary MMMT with heterologous component of the fallopian tube (Figure 2). Also left ovarian and serosa of uterin invasion were determined and two tumor involving pelvic lymph nodes were positive in the fourteen left pelvic lymph nodes. In addition, nine right pelvic nodes, six right paraaortic and seven left paraaortic lymph nodes were reactive. Cytological examination of peritoneal fluid sampled at the time of laparotomy showed atypical malignant cells.

In our patient, the stage of malignancy was Stage IIIc according to FIGO. After the surgical treatment, external radiotherapy and cyclophosphamide, doxorubicin and cis-platinum should be tried for this patient with Stage IIIc. In her first postoperative visit performed three months after her discharge she had no symptoms and the CA-125 level was found to be in normal limits.

DISCUSSION

Primary malignant neoplasms of the fallopian tube are uncommon. The most common of these tumors, carcinoma of the fallopian tube, accounts for less than 1% of all primary neoplasms of the female reproductive tract.[3] They occur mainly in the sixth decade and are typically advanced at the time of diagnosis. MMMTs arising in the fallopian tube are



a b

FIGURE 2: Microscopic observations of malignant mixed Mullerian tumor of the fallopian tube. **a.** Residual nonneoplastic fallopian tube plicae (thick arrow) were seen adjacent to the neoplasm and epithelial component of adenocarcinoma were recognized. **b.** Areas of cartilaginous differentiation (thick arrow) were showed.

exceedingly unusual and frequently seen in postmenopausal women with the symptoms of abdominal pain and abdominal distention such as ovarian neoplasms.[4] In histologic features and behavior, fallopian tube carcinoma is similar to ovarian cancer; thus, the evaluation and treatment are also essentially the same.[5]

In our case, preoperative diagnosis was ovarian cancer because of the symptoms of the postmenopausal patient, elevated CA-125 levels and ultrasonographic appearance. On the other hand, depending on the size and location, carcinoma of the fallopian tube can mimic ovarian tumors as in the present case so it is so hard to identify tubal mass from ovarian mass on ultrasonographic examination. Serum cancer antigen CA-125 is a secreted glycoprotein present in fetal amniotic and coelomic epithelium and the accepted upper limit of normal post-menopausal women is 35 IU/mL. Elevated levels are detected in approximately 80 percent of ovarian carcinomas at the time of diagnosis; however, elevated serum levels have also been reported in a variety of conditions such as endometriosis and gastrointestinal tumors.[6] Additionally, preoperative CA-125 elevation is common in patients with uterine MMMTs, regarded as high-grade adenocarcinoma of the uterus and that the epithelial component stains immunohistochemically for CA-125.[7] So CA-125 may not be helpful to differentiate the definitive diagnosis.

Standard surgical management for primary malignant neoplasms of the fallopian tube that is extensive staging laparotomy including a total abdominal hysterectomy and bilateral salpingo-oophorectomy with pelvic and paraaortic lymph node dissection is similar to the ovarian neoplasms.[8] So surgical staging is preferred in our case. The treatment of the patients with Stage IIIc MMMTs of the fallopian tube is surgical staging followed by radiation therapy or then chemotherapy, consisting of cyclophosphamide or doxorubicin or cis-platinum or both of them as literature.[9-11] This approach with surgery and combination platinum based chemotherapy should be used successfully in a majority of patients with MMMTs of the fallopian tube. And a report suggested that postoperative external radiation therapy might increase the survival rates of the patients with MMMTs of the fallopian tubes.[12]

If the sarcomatous portions of the tumor show differentiation toward mesenchymal tissues native to the fallopian tube such as smooth muscle, the tumor designated as having homologous elements. Both if differentiation toward mesenchymal elements not normally seen in the fallopian tube such as cartilage or bone is present, the tumor is designated as having heterologous elements.[9] Therefo-

re, our case was diagnosed as the primary MMMT with heterologous component of the fallopian tube. Additionally, in the literature MMMT of the fallopian tube were fimbriated end in five patients and tubal without originating from the fimbriae in approximately 64 patients.[12] Grossly the tumors distend the tube and typically spread to the pelvis, abdomen or both. The lumen of the tube is usually filled with solid neoplastic tissue contains areas of hemorrhage and sarcomatous components resemble those of similar tumors found elsewhere in the female genital tract.

In conclusion, MMMTs with heterologous component of the fallopian tube has various and nonspecific presentations. So the definite diagnosis is usually made postoperatively. Although it is rare, primary MMMT of the fallopian tube should be kept in mind when pelvic mass with high levels of CA-125 are detected in postmenopausal women.

REFERENCES

1. Carlson JA Jr, Ackerman BL, Wheeler JE. Malignant mixed müllerian tumor of the fallopian tube. Cancer 1993;71:187-92.
2. Scully RE, Young RH, Clement PB. Tumors of the ovary, maldeveloped gonads, fallopian tube and broad ligament. In: Rosai JR, Sobin LH, eds. Atlas of Tumor of Pathology: Armed Forces Institute o Pathology. 3rd ed. Washington: DC; 1996. p.474.
3. Benedet JL, Müller DM. Tumors of the fallopian tube: clinical features, staging, and management. Gynecol Oncol 1992;2:853-60.
4. Kuroda N, Inui Y, Ohara M, Hirouchi T, Mizuno K, Kubo A, et al. Hyaline globule-like structures in undifferentiated sarcoma cells of malignant müllerian mixed tumor of the fallopian tube. Med Mol Morphol 2007;40: 46-9.
5. Selvaggi SM. Tumors of the ovary, maldeveloped gonads, fallopian tube, and broad ligament. Arch Pathol Lab Med 2000;124:477.
6. Myers ER, Bastian LA, Havrilesky LJ, Kulasingam SL, Terplan MS, Cline KE, et al. Management of adnexal mass. Evid Rep Technol Assess (Full Rep) 2006;130:1-145.
7. Hoskıns PJ, Le N. Preoperative tumor markers at diagnosis in women with malignant mixed müllerian tumors/carcinosarcoma of the uterus.Int J Gynecol Cancer 2008;6:1200-1.
8. Tütüncü L, Muhcu M, Arslanhan N, Ertekin AA, Yergök YZ. Primary tubal carcinomas: retrospective analysis of 10 cases and review of the literature. Turkiye Klinikleri J Gynecol Obst 2005;15:29-35.
9. Carlson JA Jr, Ackerman BL, Wheeler JE. Malignant mixed müllerian tumor of the fallopian tube. Cancer 1993;71:187-92.
10. De Queiroz AC, Roth LM. Malignant mixed müllerian tumor of the fallopian tube. Report of a case. Obstet Gynecol 1970;36:554-7.
11. Deppe G, Zbella E, Friberg J, Thomas W. Combination chemotherapy for mixed müllerian tumor of the Fallopian tube. Cancer 1984;54:1517-20.
12. Gagner JP, Mittal K. Malignant mixed Mullerian tumor of the fimbriated end of the fallopian tube: origin as an intraepithelial carcinoma. Gynecol Oncol 2005;97:219-22.