

TUBANIN MALİGN MİKST MÜLLERİAN TÜMÖRÜ: MORFOLOJİK AÇIDAN FARKLI İKİ OLGU

MALIGNANT MIXED MÜLLERIAN TUMOR OF THE FALLOPIAN TUBE: MORPHOLOGICALLY DIFFERENT TWO CASES

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ÖZET

Amaç: Malign mikst müllerian tümörler (MMMT) epitelyal ve mezenkimal komponentler içeren çok agresif bifazik neoplazmlardır. Kadın genital sisteminde tubada MMMT'ler nadiren görülürler.

Olgu Sunumu: Bu bildiriye tuba fimbrial uç yerleşimli iki MMMT olgusu sunulmaktadır. Her iki hastada FIGO evresi IIIC (T3c N0 M0) idi. Tümörlerden biri homolog, diğeri heterolog tipteydi.

Tartışma: Bu nadir tümörlerin prognozu kötüdür. Hastalarımızdan biri tanı konduktan 6 ay sonra yaşamını yitirmiştir. Bu tümörler için sağkalımı artırmada tam cerrahi, kemoterapi ve eksternal radyoterapi zorunlu görünmektedir.

Anahtar sözcükler: Karsinosarkom, malign mikst müllerian tumor, tuba.

ABSTRACT

Objective: Malignant mixed müllerian tumors (MMMT) are highly aggressive biphasic neoplasms consisting of both epithelial and mesenchymal components. In female genital tract, MMMTs are rarely seen in fallopian tube.

Case Report: We will present two cases of MMMT of fallopian tube located at the fimbrial end. Both patients were FIGO stage IIIC (T3c N0 M0). One of the tumor was homologous and the other one heterologous variety.

Conclusion: These rare tumors have poor prognosis, as one of the patients died only in 6 months after the diagnosis. For these tumors complete surgery, chemotherapy and external radiation seemed to be necessary in order to improve survival.

Key Words: Carcinosarcoma, malignant mixed müllerian tumor, Fallopian tube.

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CASE REPORTS

Case 1

Fifty-six year-old female patient presented with post-menopausal pelvic pain. Bimanual pelvic examination revealed left adnexal mass. Tumor markers were within normal limits. Transvaginal ultrasonography and magnetic resonance imaging confirmed the solid mass lesion. Laparotomy was planned. During surgery, a mass lesion protruding from fimbrial end of the left fallopian tube and a second one located at the serosal surface of intestines were seen and resected. Intraoperative frozen consultation revealed that both lesions were malignant. Total abdominal hysterectomy, bilateral salpingo-oophorectomy, bilateral pelvic and paraaortic lymphadenectomy, omentectomy and appendectomy were performed. Macroscopically, tumoral mass of left tube filling the lumina was grayish-white in color. Tumor had a maximum diameter of 9,5 cm. Intestinal mass was 4,5 cm in maximum diameter and had the same macroscopic features with the mass located at fallopian tube. Microscopically both tumors had similar features. Tumor was composed of epithelial (moderately differentiated adenocarcinoma) and mesenchymal (high grade sarcoma) components (Figure 1). Sarcomatous component was homologous. Heterologous sarcomatous component was not detected. Mitotic figures were numerous. Tumor was extending beyond the wall of fallopian tube. Immunohistochemically both epithelial and mesenchymal components were positive for p53. Epithelial component was positive for CK7 and mesenchymal component was positive for vimentin. Smooth muscle actin (SMA) and desmin were negative in both components. Uterus, left ovary, right ovary, right fallopian tube, omentum, appendix and lymph nodes were free from tumor. The pa-

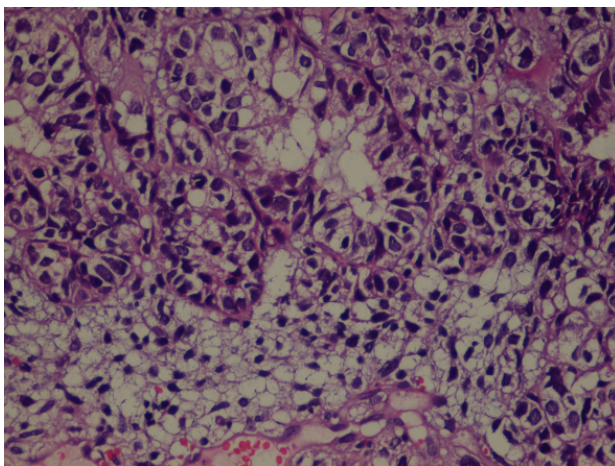


Figure 1 • Moderately differentiated adenocarcinoma (in the upper portion of the figure) and high grade sarcoma (at the bottom) components in Case 1 (H-E, x200).

tient was in FIGO stage IIIC (T3c N0 M0). She was well after surgery and 6 courses of paclitaxel and carboplatin chemotherapy within 3 weeks interval was scheduled. However before therapy regimen has been started, the patient was lost to follow up.

Case 2

Sixty-six year-old female patient presented with post-menopausal vaginal bleeding. By ultrasound, left adnexal mass was detected. Laparotomy was planned. During surgery, mass lesion located at the fimbrial end of the left fallopian tube was observed. Tumor was measuring 6,5x5x2,5 cm. Surgical exploration revealed two more mass lesions located at omentum and Douglas' pouch. Total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy, bilateral pelvic and paraaortic lymphadenectomy were performed. Microscopically tumoral lesions located at left fallopian tube, omentum and Douglas' pouch were composed of both epithelial (poorly differentiated carcinoma) and mesenchymal (high grade sarcoma) components. In fallopian tube, in situ carcinoma regions were detected (Figure 2). Sarcomatous areas were consisting of homologous and heterologous components. Heterologous component was chondrosarcoma (Figure 3). Mitotic figures were numerous. Immunohistochemically sarcomatous areas were positive for vimentin. CK5/6 and desmin were negative. Uterus, left ovary, right ovary, right fallopian tube and lymph nodes were free from tumor. The patient was in FIGO stage IIIC (T3c N0 M0). She was well after surgery. Paclitaxel and carboplatin chemotherapy was given for six courses. One month after completion of her chemotherapy regimen, recurrent MMT occurred in vaginal cuff. During her second chemotherapy the patient died of disease.

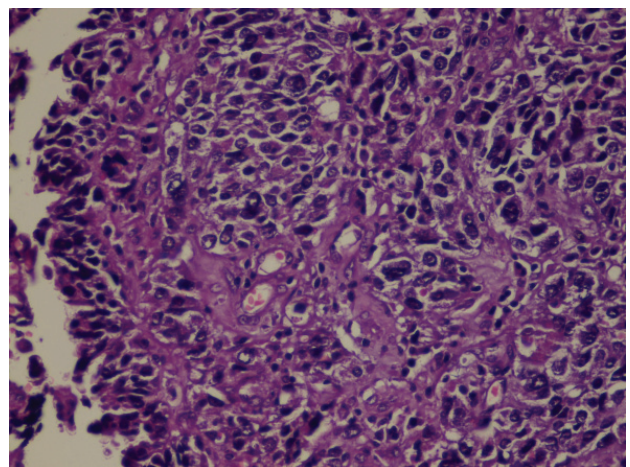


Figure 2 • Poorly differentiated carcinoma having an in situ carcinoma component at the surface in Case 2 (H-E, x200).

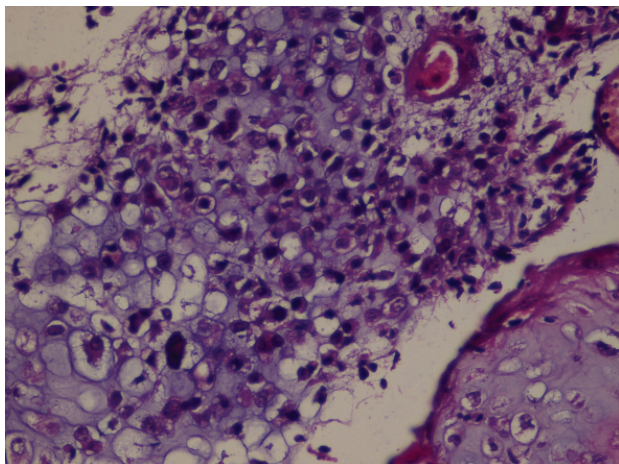


Figure 3 • High grade chondrosarcoma component in Case 2 (H-E, x200).

DISCUSSION

MMMTs of the fallopian tube are very rare neoplasms. In the English literature 82 cases had been reported and with this report authors add two more (1,2). The incidence of these tumors is reported to be 2-5% of all MMMTs (3,4). Although it has been reported in young patients (5), the majority of the patients are postmenopausal. Mean age is reported to be 57 (6). These tumors are highly malignant neoplasms and they have a poor prognosis.

There were different theories regarding the histogenesis of MMMTs. The collision theory suggests both components are different neoplasms. The conversion theory claims that sarcomatous component derives from epithelial component. The composition theory suggests that mesenchymal component is a pseudosarcomatous stromal reactive process to the presence of carcinoma, which is unlikely because of the apparent malignant nature of the mesenchymal component. The combination theory suggests that both components are derived from a single cancer stem cell (7). In a recent report it was claimed that in uterine MMMTs a CD133+ tumor cell subpopulation displayed biphasic properties and showed characteristics like cancer stem cells, supporting the theory of single stem cell undergoing divergent differentiation (8).

A homologous and a heterologous variety of MMMTs occur. In homologous variety sarcomatous component is composed of tissues normally found in the organ. In heterologous variety sarcomatous component consist of tissues nonspecific to the fallopian tube. Although in uterine MMMTs previously it was claimed that homologous variety had better survival than heterologous variety (9), in some recent reports it

was suggested that this separation is not prognostically significant (10). One of the cases presented here was homologous and the other one was heterologous variety. Both cases were FIGO Stage IIIc. Ozguroglu et al. claimed that in ovarian and endometrial MMMTs, patients with predominating carcinomatous component instead of sarcomatous component, responded better to chemotherapy regimen (11). Due to the lack of large case series of MMMT of fallopian tube we cannot predict the outcome in these neoplasms according to the predominant histopathologic feature; however we may speculate that this may be valid for these tumors also. In our both cases sarcomatous component were predominating and they both presented in advanced stage and furthermore behaved aggressively.

Rare cases of peritoneal primary MMMT have been reported (12), however the presented MMMTs of fallopian tube are unlikely to have a peritoneal origin because in case 1 omentum involvement was not present. In case 2 we have detected in situ carcinoma of fallopian tube. In both cases, the suggestion of a uterine origin is not supported due to the lack of involvement by tumor.

Presented tumors were both located at fimbrial end of the fallopian tube. Because fimbrial tumors are exposed to the peritoneal cavity without invasion of the tubal wall, it was suggested that fimbrial tumors might be associated with an increased risk of peritoneal spread and a worse prognosis (13,14).

It was suggested that treatment comprising of complete surgery, chemotherapy and external radiation seemed to be necessary in order to improve survival both in early and advanced stage patients (3,15). Treatment modality is the same as in uterine and ovarian MMMTs'. Combination paclitaxel-platinum chemotherapy was suggested to be effective in MMMT of the ovary (16) and reported to be beneficial also in patients with MMMT of the fallopian tube (2,17). It has been stated that paclitaxel-carboplatinum chemotherapy regimen resulted in shrinkage of the tumor size and combined with debulking surgery complete regression (2,17).

In conclusion, we have presented two morphologically different MMMT cases of fallopian tube origin in advanced stage. These rare tumors have poor prognosis, as one of the patients died only in 6 months after the diagnosis.

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