A sarcomatous-type peritoneal malign mixed mullerian tumor implant in association with ovarian adenocarcinoma: a case report

M. Dede, K. Gezginç, İ. Alanbay, U. Fidan, M. Yenen
[1]*Department of Obstetrics and Gynecology, Gulhane Military Medical Academy, Ankara (Turkey)*

Summary

A rare case of a patient with a histopathological diagnosis of a sarcomatous-type peritoneal malign mixed müllerian tumor implant in association with ovarian adenocarcinoma is reported. A 52-year-old patient was referred to our clinic for an adnexal mass. At pelvic examination, an irregular, fixed, approximately 7-8 cm in size mass was detected in the right adnexal area. At transvaginal ultrasonographic examination, it was observed that there was an 80 x 70 mm sized, irregularly contoured, semisolid mass with hyperechogenous areas inside originating from the ovary in the right adnexal area. At laboratory examination tumor marker CA-125 was 280.4 U/ml (< 35), CA-15-3 was 146.5 U/ml (< 25), whereas other markers were within normal range. The patient was operated on for a right adnexal mass. A staging laparatomy procedure was applied. Postoperative histopathological diagnosis was reported as malignant mixed mullerian tumor of the ovary, with the ovarian component as poorly differentiated adenocarcinoma, and the metastatic foci over serosal surfaces as a sarcomatous component. Postoperatively six courses of adjuvant and consolidation chemotherapy were administered to the patient. Further studies are needed to set a consensus about evaluation of treatment and prognosis for this kind of pathology.

Key words: Ovarian malign mixed mullerian tumor; Peritoneal implant; Treatment.

Introduction

Malign mixed mullerian tumors of ovary are rarely seen [1]. The mean survival in these cases is 7-13 months [2]. For the management of these tumors, similar to malign epithelial ovarian tumors, platinium-based treatment in adjuvant chemotherapy is the accepted treatment modality [3]. Our case of a malign mixed mullerian tumor implant in association with ovarian adenocarcinoma is the first one reported in the literature. There was an aggressive adenocarcinomous component of primary malign mixed mullerian tumor of the ovary, and there was a dominance of the sarcomatous component at the metastatic foci.

Case Report

A 52-year-old patient was referred to our clinic after an adnexal mass was detected in her pelvic ultrasonograph (US) during a routine postmenopausal checkup. At pelvic examination, a 7-8 cm sized, irregularly contoured, fixed mass was detected in the right adnexal area. At transvaginal ultrasonographic (TVS) evaluation, it was observed that there was an 80 x 70 mm sized, irregularly contoured, semisolid mass with hyperechogenous areas inside the right adnexal region. Laboratory results showed tumor marker CA-125 as 280.4 U/ml (< 35) and CA-15-3 as 146.5 U/ml (< 25) while other markers were within normal range. Computerized tomography showed an approximately 10 x 12 x 7 cm sized, lobulated and contoured almost cystic mass with centrally located solid components inside, originating most probably from the right ovary occupying the right middle side of the pelvis. Moreover there were approximately 5 and 3 cm sized solid components located superolaterally to the lesion. There was diffuse dirtiness and thickening on the omental fatty tissue, and omental infiltration (Figure 1). There were no pathological findings at rectosigmoidoscopy and cystoscopy. The patient was operated on with prediagnosis of a 'right adnexal mass'. During operative abdominal exploration, there was an 8 cm sized, fixed, solid, irregularly contoured mass originating from the right ovary, omental infiltration, miliary foci on the serosal surfaces and also diffuse adhesions between the sigmoid colon, omentum, uterus and a lesion located in the right adnexal area. A staging laparatomy procedure, right tumoral oophorectomy, bilateral salpingo-oophorectomy, total abdominal hysterectomy, total omentec-

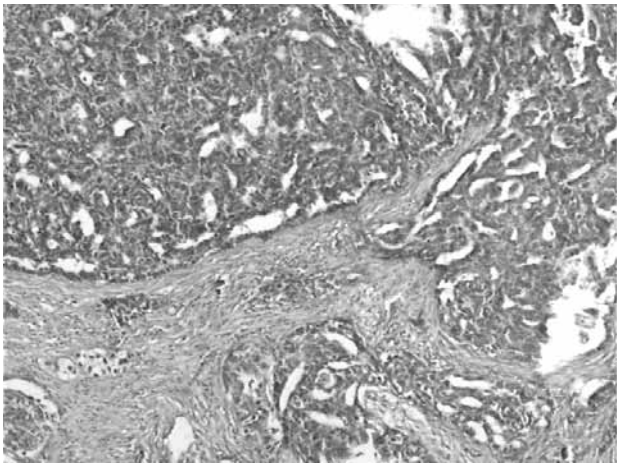
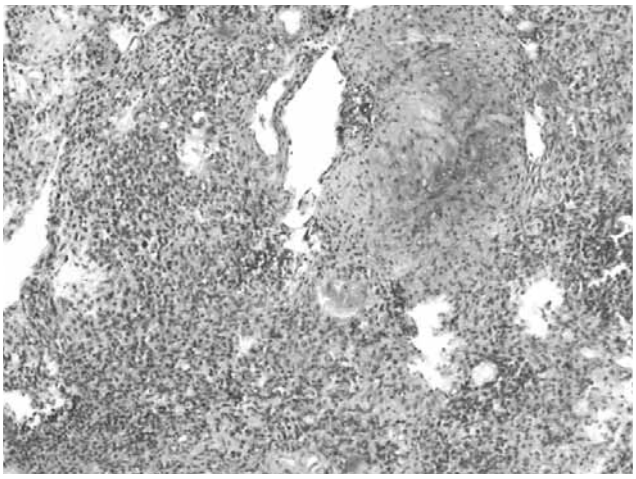
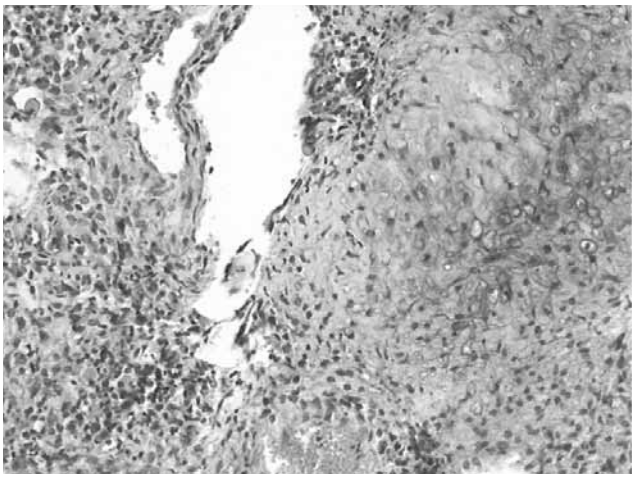


Figure 1. — The mass-like lesion in the ovary - "adenocarcinoma".

Revised manuscript accepted for publication June 28, 2007

Fig. 2a   Fig. 2b

Figures 2a, 2b. — Peritoneal serosal malign mixed mullerian implants (sarcomatous component).

tomy, pelvic-paraaortic lymphadenectomy, and appendectomy were performed. Appendectomy is applied to all ovarian cancer patients according to our clinical approach. The postoperative histopathological diagnosis was reported as malign mixed mullerian tumor of ovarian origin, with an ovarian component as a poorly differentiated adenocarcinoma, and metastatic foci on the serosal surfaces as a sarcomatous (muscle, cartilage) component (Figure 2a, Figure 2b). The patient received six courses of adjuvant chemotherapy consisting of a carboplatin and paclitaxel combination after surgery. Following adjuvant chemotherapy, the patient received six courses of consolidation chemotherapy with paclitaxel (175 mg/m^2), and the last CA-125 level was detected as 9.4 U/ml. The patient died due to a recurrent ileus one year after the operation.

Discussion

Malign mixed mullerian tumors of ovarian origin are rather rare conditions. Tumor prognosis is poor, and advanced stage at the time of diagnosis is the most important factor affecting prognosis negatively. In 70% of cases diagnosis is at Stage 3 and 4 and after a short period of time cases are lost due to advanced stage poor prognosis [4]. Disease is usually observed in the postmenopausal period, in the 7th decade. The tumor originates from pluripotent mesenchymal cells and differentiates to epithelial and stromal components [5]. Ovarian malignant mixed mullerian tumors (OMMMT) are classified as heterologous and homologous. Homologous tumors may differentiate to natural or foreign cells to the ovary such as 'spindle cells', while heterologous tumors may include tissues such as bone and cartilage that are not present in the ovary.

Clinically, they have a progression similar to epithelial tumors and the initial symptoms mainly include abdominal distension, pain, nausea, vomiting and weight loss. Prognosis in homologous OMMMT appears better compared with heterologous tumors. However there is not any concensus on this topic [6, 7].

Malign mixed mullerian tumors of the uterus have lung metastasis frequently, but ovarian malign mixed mullerian tumor have metastasis primarily on peritoneal and serosal surfaces like primary ovarian cancer [8]. In our case also, metastasis occurred in this way. However in the literature, the histopathological diagnosis of OMMMT of the ovary and peritoneal surfaces is similar. In this case, we detected a carcinomatous component in the ovary, whereas the sarcomatous component was detected in the metastatic foci. Thus, although the case seemed similar to primary ovarian carcinoma, after histopathological investigation it was clear that it was a malign mixed mullerian tumor originating from the ovary.

In OMMMT cases after staging laparatomy, usually platinium-based chemotherapy protocols are preferred [9]. Since the number of cases in the literature is limited, there is not any concencus on an optimal chemotherapeutic regimen. Nonetheless the generally accepted treatment protocol for these tumors is platinum-based adjuvant chemotherapy, similar to that for malign epithelial ovarian tumors. We believe that we would have more information about the efficacy of chemotherapeutic agents used in OMMMT with the help of an increased number of cases and series in older women. Our patient received six courses of adjuvant chemotherapy consisting of a combination of carboplatin and paclitaxel after surgery. Following adjuvant chemotherapy, the patient received six courses of consolidation chemotherapy with paclitaxel (175 mg/m^2), and the last CA-125 level was detected as 9.4 U/ml.

The co-existence of ovarian adenocarcinoma together with a serosal malign mixed mullerian tumor is a rare entity. Thus, to establish a concensus on the treatment and prognosis of these types of pathologies, further studies are needed.

References

[1] Wei L.H., Huang C.Y., Cheng S.P., Chen C.A., Hsieh C.Y.: "Carcinosarcoma of ovary associated with previous radiotherapy". *Int. J. Gynecol. Cancer,* 2001, *11,* 81.

[2] Ariyoshi K., Kawauchi S., Kaku T., Nakano H., Tsuneyoshi M.: "Prognostic factors in ovarian carcinosarcoma: a clinicopathological and immunohistochemical analysis of 23 cases". *Histopathology,* 2000, *37*, 427.

[3] Muntz H.G., Jones M.A., Goff B.A. *et al*.: "Malignant mixed mullerian tumors of the ovary: experience with surgical cytoreduction and combination chemotherapy". *Cancer*, 1995, *76*, 1209.

[4] Bicher A., Levenback C., Silva E.G., Burke T.W., Morris M., Gershenson D.M.: "Ovarian malignant mixed mullerian tumors treated with platinum- based chemotherapy". *Obstet. Gynecol*., 1995, *85,* 735.

[5] Gallardo A., Matias-Guiu X., Lagarda H. *et al*.: "Malignant mullerian mixed tumor arising from ovarian serous carcinoma: a clinicopathologic and molecular study of two cases". *Int. J. Gynecol. Pathol*., 2002, *21*, 268.

[6] Scully R.E., Young R.H., Clement P.B.: "Tumors of the ovary, maldeveloped gonads, fallopian tube, and broad ligament". In: Rosai J. (ed.). Atlas of Tumor Pathology, 3rd series, fascicle 23 Washington, DC: Armed Forces Institute of Pathology, 1998, 128.

[7] Hellstrom A.C., Tegerstedt G., Silfversward C., Pettersson F.: "Malignant mixed mullerian tumors of the ovary: histopathologic and clinical review of 36 cases". *Int. J. Gynecol. Cancer,* 1999, *9,* 312.

[8] Bicher A., Levenback C., Silva EG., Burke T.W., Morris M., Gershenson D.M.: "Ovarian malignant mixed mullerian tumors treated with platinum-based chemotherapy". *Obstet. Gynecol*. 1995, *85*, 735.

[9] Baker TR., Piver MS., Caglar H., Piedmonte M.: "Prospective trial of cisplatin, adriamycin, and dacarbazine in metastatic mixed mesodermal sarcomas of the uterus and ovary". *Am. J. Clin. Oncol*., 1991, *9,* 246.

Address reprint requests to:
K. GEZGINÇ, M.D.
Department of Obstetrics
and Gynecology
Gulhane Military Medical Academy
06018 Etlik, Ankara (Turkey)