EJGO European Journal of Gynaecological Oncology

Undifferentiated pleomorphic sarcoma with focally rhabdomyosarcomatous differentiation of the ovary

E. Kurtoglu[1], H. Celik[1], A. Kokcu[1], M. Kefeli[2], A.Z. Ozdemir[1], M. Danaci[3]

[1] Department of Obstetrics and Gynecology, [2] Department of Pathology, [3] Department of Radiology, Ondokuz Mayis University, Samsun (Turkey)

Summary

Purpose of investigation: Primary sarcomas account for of 2-3% of all female genital tract cancers and ovaries are unusual sites for sarcomas. The authors aimed to report a case of primary ovarian sarcoma. *Materials and Methods:* A 46-year-old woman presented with abdominal pain and distention. Abdominal MRI revealed a 20-cm-sized complex mass in the right adnexa and ascites. Preoperative CA-125 was 289 U/ml. She underwent optimal debulking surgery and diagnosed with undifferentiated pleomorphic sarcoma with focally rhabdomyosarcomatous differentiation. *Results:* The patient presented with severe abdominal pain and distention 32 days after the operation and recurrent disease was diagnosed. First cure of adjuvant chemotherapy was administered, but she died 15 days later. *Conclusion:* Coexistence of undifferentiated pleomorphic sarcoma and rhabdomyosarcoma shows highly aggressive behavior and its prognosis is extremely poor. To the best of the authors' knowledge, this is the second case report of the coexistence of these type tumors in the literature.

Key words: Undifferentiated pleomorphic sarcoma; Rhabdomyosarcoma; Ovary.

Introduction

Primary sarcomas, arising from mesenchymal cells, represent 2-3% of all female genital tract malignancies, 10% of which occurs outside the uterus [1]. Ovary is an unusual site for sarcoma which may exist with different types such as osteosarcoma, chondrosarcoma, and angiosarcoma [2-4]. Here, The authors present the second case of undifferentiated pleomorphic sarcoma with rhabdomyosarcomatous degeneration arising from the right ovary in a 46-year-old woman.

Case Report

A 46-year-old, gravida 1, para 1, premenopausal woman was admitted to emergency department with lower abdominal pain and distention lasting for two months. Her past history was unremarkable except for hypertension and new diagnosed diabetes mellitus. On physical examination, her blood pressure was 120/80 mm Hg, heart rate was 100/min. The abdomen was distended but there was no tenderness. Abdomino-pelvic MRI revealed a 20-cm-sized complex mass in the right adnexa, ascites, omental infiltration, and pelvic-para-aortic lymphadenopathies (Figure 1). Preoperative hemoglobin was 10.3 mg/dl and CA-125 was 289 U/ml. The patient underwent laparotomy and solid mass including necrotic and hemorrhagic areas were found arising from the right adnexa and covered by the infiltrated omentum and dense adhesions. Left adnexa

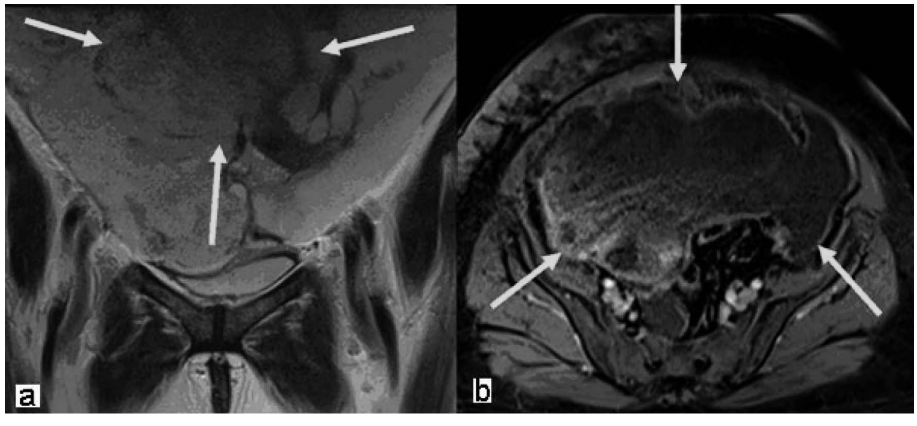

Figure 1. — Abdomino-pelvic MRI, A: T2 weighted, B: T1-weighted dynamic contrast enhanced MRI series. A heterogeneous tumoral mass completely occupying the pelvic cavity in the initial evaluation.

Revised manuscript accepted for publication February 10, 2015

Eur. J. Gynaecol. Oncol. - ISSN: 0392-2936
XXXVII, n. 3, 2016
doi: 10.12892/ejgo2844.2016

7847050 Canada Inc.
www.irog.net

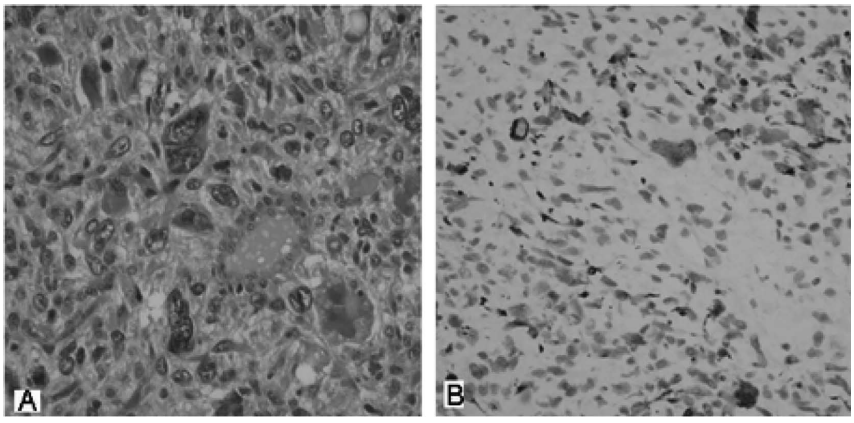

Figure 2. — A: Atypical mesenchymal cells and multinucleated giant tumor cells in undifferentiated pleomorphic sarcoma (H&E, ×100 and ×400). B: Focally desmin positivity for tumor cells (DAB, ×200).

was infiltrated by the tumor but there were no implants on the peritoneal surface or the entire organs including bowels, liver, and spleen. For optimal debulking, total hysterectomy, bilateral salpingo-oophorectomy, and total omentectomy were performed. There was no postoperative complication and the patient was discharged after nine days.

Macroscopically, the left ovarian mass was enlarged and measured 18×11×6 cm. The mass in gross form had mostly solid components and outer surface of mass was irregular. Section surface was brown and had some small cysts. There was extensive necrosis and hemorrhage on section surface. The right ovarian mass measured 12×10×9 cm, and had same features of left ovarian mass. There was capsule invasion in the both ovaries.

Microscopically, the both ovarian mass were a high-grade sarcoma. It consisting of spindle- to oval-shaped pleomorphic cells with hyperchromatic nuclei and scanty cytoplasm. There were multinucleated giant tumor cells. Rhabdomyoblast-like cells were polygonal shaped with abundant bright eosinophilic cytoplasm. Mitotic figures were common and counted 30-40 mitosis in per ten high power fields. Tumor had extensive necrosis and hemorrhages (Figures 2A, B). Immunohistochemically, these cells were diffusely positive for vimentin (clone V9), but negative for cytokeratin (AE1/AE3), S-100 (clone 4C4.9), CD-34 (clone QBEnd/10), CD-31 (clone JC/70A) and Caldesmon (Clone h-CALD). Desmin (Catalog No RP011), and SMA (Clone 1A4) were positive for scattered cells. Sections from the right parametrial tissue revealed tumoral invasion. The uterus, fallopian tubes, and omental specimens were free of the tumor. There were no Müllerian epithelial components and teratomatous elements in multiple samples. The final histological diagnosis was undifferentiated pleomorphic sarcoma with focally rhabdomyosarcomatous differentiation of both ovaries.

When the patient's results were prepared to be discussed at the gynecologic oncology council of the university, she presented with severe right lower abdominal pain, fatigue, nausea, and vomiting 32 days after the operation. Abdomino-pelvic CT demonstrated a complex mass consisted of two main parts, each of which was ten cm in diameter, ascites and multiple vaginal, peritoneal, and intestinal mesenteric implants. (Figure 3) The CA-125 level was 29 U/ml. She was diagnosed with recurrent disease and adjuvant chemotherapy including cisplatinum and doxorubicin was planned by medical oncology department. However, the therapy was delayed due to upper gastrointestinal bleeding and intensive care. As soon as the patient became hemodynamically stabilized, she received the first cycle of combined chemotherapy. However, 15 days later she died due to hypotension unresponsive to the treatment.

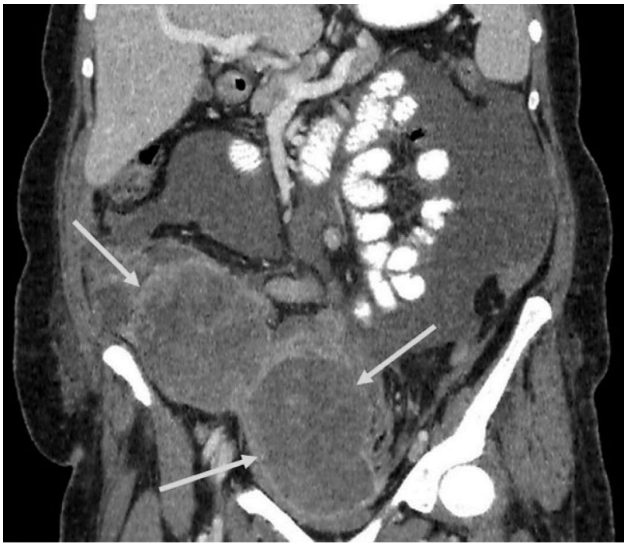

Figure 3. — Coronal contrast enhanced CT image. Appearance of recurrent tumoral mass consisting of two parts, each of which is in size of ten cm, 32 days after the primary operation.

Discussion

Female reproductive tract, particularly the ovary, is an exceedingly rare site for primary mesenchymal tumors [5]. There have been only a few reports of primary sarcomas in the ovary including only one rhabdomyosarcomatous degeneration in undifferentiated pleomorphic sarcoma, consequently this case is the second report of this variant [6].

Since mesenchymal tumors are highly malignant and tend to be locally and hematogenously aggressive, the main reason for admission of the patients with primary ovarian sarcoma was abdominal pain due to the compression of the pelvic mass to other organs and ascites [5-9]; however, particularly in inflammatory sarcoma, common cold-like symptoms and fever was reported secondary to

overproduction of interleukin-6 [10]. In the present case, in both first and second presentations, the patient had abdominal pain and distention as main complaints.

The management of primary ovarian sarcoma is controversial because of the heterogenous nature of the disease and the small number of the reported cases. The primary standard treatment for sarcoma is surgical excision with a tumor-free resection margin. Adjuvant chemotherapy including cisplatinum plus doxorubicin/cyclophosphamide or gemcitabine plus docetaxel, and radiation are also considered for non-myxoid tumors since they would not influence the prognosis in myxoid ones. However, the optimal adjuvant therapy is still unclear due to the rare occurrence of the disease [8, 11, 12]. The present authors performed complete surgical excision with tumor-free margin, but since the patient had a rapid recurrent disease, which was only 32 days after the primary surgery, adjuvant chemotherapy including cisplatinum and doxorubicin was not able to have any effect on the gross tumor mass.

Primary ovarian sarcoma is expected to have poor prognosis when the factors including high tumor grade, tumor size > ten cm, and tissue necrosis on histologic examination are present [13]. In this case, the patient had a recurrent high grade 20-cm-sized tumor mass and a large tissue necrosis. In previously reported cases, recurrence occurred as early as fourth month after the primary surgery, whereas there was a rapid recurrence on 32nd day of the first operation with a large-sized tumor mass in the present case. Hence, the patient had a very poor prognosis and was not able to receive planned chemotherapy cure and succumbed to the disease. The present case demonstrated that coexistence of undifferentiated pleomorphic sarcoma and rhabdomyosarcoma shows highly aggressive behavior and its prognosis is extremely poor. Fortunately, the coexistence of these tumors is very rare. To the best of the authors' knowledge, this is the second case report of the coexistence of these type tumors in the literature.

References

[1] Leppien G.: "Non-uterine gynecological sarcomas." *Arch. Gynecol. Obstet*., 1987, *241*, 25.

[2] Talerman A., Auerbach W.M., van Meurs A.J.: "Primary chondrosarcoma of the ovary." *Histopathology*, 1981, *5*, 319.

[3] Yeasmin S., Nakayama K., Ishibashi M., Katagiri A., Iida K., Manabe A., *et al.*: "Primary osteosarcoma of ovary". *Int. J. Clin. Oncol*., 2009, *14*, 163.

[4] Quesenberry C.D., Li C., Chen A.H., Zweizig S.L., Ball H.G. 3rd., *et al.*: "Primary angiosarcoma of the ovary: a case report of Stage I disease." *Gynecol Oncol.*, 2005, *99*, 218.

[5] Kransdorf M.J., Murphey M.D.: "Malignant fibrous and fibrohistiocytic tumors". *In:* Kransdorf M.J., Murphey M.D., (eds). *Imaging of soft tissue tumors.* Philadelphia: Lippincott Williams&Wilkins, 2006, 257.

[6] Mukherjee S., Sen S., Biswas P., Choudhuri M.: "Primary pleomorphic sarcoma of the ovary with rhabdomyosarcomatous differentiation." *Indian J. Pathol. Microbiol.,* 2009, *52*, 217.

[7] Choi SY, Kim TH, Park HS, Sung CO, Lee JW, Song SY, *et al.*: "Myxoid malignant fibrous histiocytoma of the ovary: a case report." *Int J Clin Oncol.,* 2011, 16, 601-4.

[8] Dilek T.U., Dilek S., Pata O., Tataroglu C., Tok E.: "Malignant fibrous histiocytoma of the ovary: a case report." *Int. J. Gynecol. Cancer*, 2006, *16*, 352.

[9] Stefanovic A., Stojnic J., Jeremic K., Jeftovic M., Arsenijevic L., Zecevic N., *et al.*: "Malignant fibrous histiocytoma of the ovary: a case report." *Eur. J. Gynaecol. Oncol*., 2012, *33*, 236.

[10] Yamakawa Y., Fujimura M., Hidaka T., Yasoshima K., Saito S.: "Inflammatory malignant fibrous histiocytoma of the ovary producing interleukin-6: A case report." *Gynecol Oncol.*, 1999, *75*, 484.

[11] Roque D.M., Jones D.F., Carter G., Kelley J.L.: "Primary giant cell malignant fibrous histiocytoma of the ovary: case report and review of the literature". *Gynecol. Oncol*., 2010, *119*, 397.

[12] Zagars G.K., Mullen J.R., Pollack A.: "Malignant fibrous histiocytoma: outcome and prognostic factors following conservation surgery and radiotherapy". *Int. J. Radiat. Oncol. Biol. Phys*., 1996, *34*, 983.

[13] Pezzi C.M., Rawlings M.S. Jr., Esgro J.J., Pollock R.E., Romsdahl M.M.: "Prognostic factors in 227 patients with malignant fibrous histiocytoma". *Cancer*, 1992, *69*, 2098.

Address reprint requests to:
E. KURTOGLU, M.D.
Ondokuz Mayis University
Department of Obstetrics and Gynecology
Kurupelit, Samsun 55200 (Turkey)
e-mail: emel0022@mynet.com