Extragenital Müllerian Adenosarcoma in the Pouch of Douglas

Ateş Karateke¹, İlker Kahramanoğlu², Remziye Bilgiç³

¹Department of Gynecologic Oncology, Medical Park Hospital, İstanbul, Turkey
²Department of Obstetrics and Gynecology, Süleymanıye Birth and Women Health Training and Research Hospital, İstanbul, Turkey
³Department of Pathology, E-Sıtopatoloji Pathology Laboratory, İstanbul, Turkey

Background: Extragenital Müllerian adenosarcomas are extremely rare tumours characterised by a stromal component of low-grade malignancy and by a benign glandular epithelial component.

Case Report: A 26-year-old woman was admitted to our clinic because of lower abdominal distension and left lower quadrant pain. Clinical and radiological examinations suggested an ovarian malignancy. Laparotomy revealed a cystic mass in the pouch of Douglas, originating from the left sacrouterine ligament. A total excision of the tumour was performed and showed low-grade adenosarcoma without sarcomatous overgrowth. Follow-up at 24 months after the surgery showed no evidence of recurrence.

Conclusion: Müllerian adenosarcoma located in the pouch of Douglas is rare. For treatment, success may be achieved with only excision of the tumour if there is no sarcomatous overgrowth or spread to adjacent tissues. (Balkan Med J 2014;31:100-104).

Key Words: Adenosarcoma, adnexal, Douglas, extragenital, Müllerian, tumour

Müllerian adenosarcoma is a mixed epithelial-mesenchymal neoplasm that originates from the Müllerian duct and is characterised by benign epithelial gland and malignant stromal components as active participants in the neoplastic process (1). The uterine corpus is the most common primary site but Müllerian adenosarcoma has been reported to arise in the ovary, cervix, vagina, pelvic peritoneum, pouch of Douglas, broad ligament, bladder, and colon (2-5). Although adenosarcomas are generally low-grade malignancies, adenosarcoma with sarcomatous overgrowth is a highly aggressive tumour. These tumours are frequently associated with postoperative recurrence or metastases and a fatal outcome, even in early-stage disease (6).

We present a case of Müllerian adenosarcoma located in the pouch of Douglas and report the clinical and pathological findings. This is the second case reported in the literature that was treated by total excision of the tumour, and no recurrence was seen during the subsequent 24-month period.

CASE PRESENTATION

A 26-year-old, gravida 0, para 0 woman with lower abdominal distension and left lower quadrant pain was seen in our hospital. She had no significant medical or family history. Her menstruation cycles were regular and she had used no medica-
completely from the left sacrouterine ligament, the left ureter was identified, intraoperative frozen section was performed, and a low-grade mesenchymal tumour was diagnosed. The surgery included omentectomy and peritoneal washing.

The gross pathological specimen of the mass was described as a multinodular predominantly solid mass with almost ping-pong ball-sized peripheral cystic areas (Figure 2). The solid component broke into the capsule as solid polypoid and papillary excrescences. Microscopically, the diagnosis of low-grade adenosarcoma without sarcomatous overgrowth was made (Figure 3), suggested that the mass arose from surface peritoneal endometriosis. Mitotic activity averaged 4-6 per 10 high-power fields (HPFs; x 400). No heterologous elements were seen. A baseline positron emission tomography (PET)/CT scan showed no FDG-avid intra-abdominal or retroperitoneal nodes or foci. No abnormal FDG-avid foci were noted in the liver, spleen, pancreas, or adrenal glands. Postoperatively, the patient did not receive chemotherapy. Follow-up at 24 months after surgery showed no evidence of recurrence.

**FIG. 1.** Sagittal T2-weighted magnetic resonance image demonstrated a large heterogeneous predominantly solid mass, containing multiple papillary excrescences

**FIG. 2.** Laparotomy revealed the mass in the lower abdomen, which originated from the left sacrouterine ligament and measured 18 cm. The solid mass is covered by a cystic wall, which is seen as a pink tissue

**FIG. 3.** a, b. Coexistence of the epithelial and stromal components. Benign appearing glandular structures form a ‘leaf-like’ pattern. Note the stromal hypercellularity. Haematoxylin and eosin, 100 x magnification (a). The sarcomatous component showed scattered mitotic figures. Periglandular stromal cuffing is seen. Haematoxylin and eosin, 400 x magnification (b)
DISCUSSION

Adenosarcoma was first described in 1974 as a mixed epithelial-mesenchymal tumour of a usually low-grade malignancy (1). Adenosarcoma is a rare tumour that accounts for 8% of malignant stromal tumours. Extrapelvic adenosarcomas occur at a younger age (median age of 49 years) compared with uterine adenosarcomas (7).

Adenosarcoma should be histologically differentiated from its uterine counterpart, adenofibroma. Such histologies are similar to the presence of large areas of low cellularity and infrequent mitoses. Increased nuclear pleomorphism with a mitotic rate of two per HPF, distinctive periglandular cuffs of cellular stroma with or without intraglandular protrusions of stromal element, and invasion have been suggested as criteria for differentiating between these tumours (7). Both extrapelvic and uterine adenosarcomas may have heterologous elements.

The major factor affecting the outcome is tumour location. The clinical behaviour of adenosarcoma differs based on the site of origin of the tumour. Extraglandular tumours behave more aggressively, with recurrence in 50% compared with 25% in uterine primaries. Haematogenous metastases occur in 33% of extrapelvic cases compared with 2% of uterine primaries. Mortality rates are 40% and 10% in extrapelvic and

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Wks: weeks; Chemo: chemotherapy; Sx: surgery; Rx: radiation therapy
uterine adenosarcomas, respectively (8-10). The reasons for the poorer prognosis of extragenital adenosarcoma have not been explored yet. Possible reasons are increased risk of local complications at extragenital sites such as bowel obstruction, increased size of tumours at presentation, and difficulty of providing complete surgical clearance in comparison with uterine primaries (8).

In addition, the degree of myometral invasion and sarcomatous overgrowth (as defined by pure sarcoma without epithelial elements occupying at least 25% of the tumour) are the other negative prognostic factors (6, 8).

By contrast, associated endometriosis is a favourable prognostic factor in adenosarcoma patients (6). In their review, Huang et al. (10) suggested that the presence of endometriosis might confer a better prognosis in patients with extragenital adenosarcoma. Extragenital adenosarcoma may develop from malignant transformation of endometriosis or originate from multipotential pelvic mesothelial cells (7). In our case, even though foci of endometriosis were not identified before and during the surgery, areas of endometriosis were present on the pathology specimen. While endometrioid adenosarcoma is the second most common histotype among endometriotic lesions, it may not be a coincidence of endometriosis and the tumour in our case.

Some suggest that the therapeutic cornerstone is still radical surgery. A surgical approach similar to that used for the corresponding disease stages of endometrial carcinoma is recommended (14). However, there is no clear consensus about the type of surgery. In English literature, there are only four cases of extragenital adenosarcoma with a pouch of Douglas location. Local excision of tumour was performed in one case. After the 24-month follow-up of this pouch of Douglas-located case, local multiple recurrences were detected and radical surgery performed. Surgical treatment was supplemented by radiation therapy (6) (Table 1). In our case, because the patient was 26, wished to preserve her fertility, and intraoperative frozen section diagnosis was low-grade adenosarcoma, we performed simple excision of the mass.

Some cases with uterine sarcoma respond to doxorubicin-based chemotherapy. Also, Huang et al. (10) observed a clinical response to doxorubicin chemotherapy in their patient and recommend the use of doxorubicin in all patients, especially those with extragenital adenosarcoma with sarcomatous overgrowth, due to the high risk of recurrence and death. Because of the diagnosis of low-grade adenosarcoma without sarcomatous overgrowth, we did not use chemotherapy for our patient. The interesting feature of this case is the fact that extragenital adenosarcomas may be confused with ovarian malignancy on clinical and radiological examination.

There is no clear recommendation for treatment of extragenital adenosarcomas. Although disease recurrence occurs in over half of patients, no recurrence was seen in our patient. The presence of endometriosis and the absence of sarcomatous overgrowth might influence prognosis as favourable factors. However, long-term follow-up is needed. Because adenosarcoma may be seen in patients who wish to preserve their fertility, fertility-sparing surgery can be considered, especially if there is no sarcomatous overgrowth or spread to adjacent tissues.

Ethics Committee Approval: Ethics committee approval was received for this report.

Informed Consent: Written informed consent was obtained from patients who participated in this study.

Conflict of Interest: No conflict of interest was declared by the authors.

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