

MR Findings of Extrauterine Müllerian Adenosarcoma Associated with Deep Pelvic Endometriosis¹

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Extrauterine müllerian adenosarcoma is a very rare tumor and it is characterized by a benign glandular component and a low-grade sarcomatous stromal component. These tumors have been reported to arise from ovarian or extraovarian endometriosis. However, there are scant reports on the MR findings of extrauterine müllerian adenosarcoma arising from deep pelvic endometriosis. We describe here a case of a large infiltrating extrauterine müllerian adenosarcoma arising from recurrent deep pelvic endometriosis and we discuss its MR findings.

Index words : Endometriosis

Mixed tumor, Müllerian

Magnetic resonance (MR)

Adenosarcoma

Uterus

Müllerian adenosarcoma was first described in 1974 as a tumor composed of admixed benign epithelial and malignant nonepithelial components, and this tumor usually develops as a solitary lesion in the uterine corpus (1). These neoplasms can develop in the endometrial tissue as well as in the ectopic foci of endometriosis. Rare extrauterine müllerian adenosarcomas have been reported to arise in the ovary, cervix, vagina, pelvis, bladder and colon (2, 3). However, to the best of our knowledge, there are scarce reports on the MR findings of extrauterine müllerian adenosarcoma arising from deep pelvic endometriosis. We describe here a case of a large infiltrating extrauterine müllerian adenosarcoma arising from recurrent deep pelvic endometriosis and

we discuss the MR findings.

Case Report

A 38-year-old nullipara woman underwent left salpingo-oophorectomy and right cystectomy with adhesiolysis in March 2003, and the pathologic results revealed endometriosis without malignant foci (Fig. 1). In December 2006, she revisited our hospital complaining of pelvic pain. On rectal examination, a hard fixed mass was palpable on the rectal wall. Sigmoidoscopy showed a smooth elevated lesion with hyperemic mucosal changes, and this caused extrinsic compression of the rectum. She underwent pelvic MR imaging, which demonstrated a large, infiltrating solid mass in the pelvis (Fig. 2). The mass was mainly located in the cul-de-sac involving the torus uterinus, the bilateral uterosacral ligaments, vagina, rectovaginal septum, cervix and anterior rectal wall. The lesion demonstrated heterogeneously high signal intensity on the T2-weighted images, low signal intensity on the T1-weighted images and homoge-

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Received October 5, 2007 ; Accepted December 20, 2007

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neously strong enhancement on the postcontrast T1-weighted images. The right ovary was normal in appearance with multiple physiologic cysts. Left hydronephroureterosis was seen, and this was due to left distal ureter invasion from the pelvic mass. There was no evidence of enlarged lymph nodes or an abnormal fluid collection. She had no specific findings on the routine laboratory tests, including tumor markers such as CA-125, CA 19-9 and carcinoembryonic antigen. Recurrent deep pelvic endometriosis was most likely for the differential diagnosis based on the pelvic MR imaging, but malignant transformation of deep pelvic endometriosis could not be excluded because of the large size and aggressive morphology of the mass.

She underwent total abdominal hysterectomy with right salpingo-oophorectomy, but all the infiltrative lesions of the deep pelvic endometriosis could not be removed. On the surgical specimen, irregularly soft, white to brown-tan polypoid masses were found at the vagina and cervix. Histopathologically, the lesions showed spindle cell proliferation with increased cellularity around the endometrial gland, but the mitotic activity was low with the range of 0 to 2 mitotic figures per 10 high power fields, and focal stromal predominance was also noted. Further, there was endometriosis in the vagina and cervix. Therefore, this histologic pattern led to a diagnosis of müllerian adenosarcoma arising from recurrent deep pelvic endometriosis (Fig. 3).

Postoperatively, she underwent 3 cycles of combination chemotherapy with ifosfamide and cisplatin, but

the last cycle was not completed. Concurrent radiation therapy was also begun and this was scheduled at 28 doses for a total dose of 50.4 Gy. In the middle of adjuvant chemotherapy and radiation therapy, we performed follow-up MR scan of the abdomen and pelvis, and this showed remaining ill-defined infiltrative lesions in the pelvis and these involved the bilateral uterosacral ligaments, pelvic wall and rectum.

Discussion

Endometriosis is defined as the presence of endometrial tissue outside the uterus. Endometriosis is most commonly located in the ovaries and the pelvic peritoneum, followed by deep lesions of the pelvic subperitoneal space, the intestinal system and the urinary bladder (4). Sampson in 1925 first reported on some cases of malignant tumors that were diagnosed in women with endometriosis (5). Since then, malignant transformation has come to be recognized as a rare complication of endometriosis. The frequency of malignant transformation of endometriosis is unknown, but it is estimated that up to 1% of endometriosis will develop into endometriosis-associated neoplasm. The period between the original diagnosis of endometriosis and extrauterine adenosarcoma was reported to be more than 2 - 5 years (3). Many different types of epithelial and stromal malignancies arising from endometriosis have been reported and well described in the literature (6). These tumors most commonly arise in the ovary and about 20% arise in extraovarian sites. Extraovarian lesions are mostly composed of endometrioid tumors (66%) and sarcomas (25%).

Extrauterine müllerian adenosarcomas occur in younger women and these tumors are more aggressive than uterine tumors (7). The M.D. Anderson Cancer Center experience of 41 cases of müllerian adenosarcomas showed 29% were extrauterine, and the nonvaginal extrauterine adenosarcomas arising from endometriosis are more frequent than vaginal adenosarcomas.

The definition of deep pelvic endometriosis, which is also called deep infiltrating endometriosis, is the infiltration of the implanted endometriosis under the surface of the peritoneum. The MR imaging characteristics of deep pelvic endometriosis have been described as low to intermediate signal intensity with punctate regions of high signal intensity on the T1-weighted images and uniform low signal intensity on the T2-weighted images. The postcontrast images demonstrate enhancement that's due to the abundant fibrous tissue (8). The typical MR

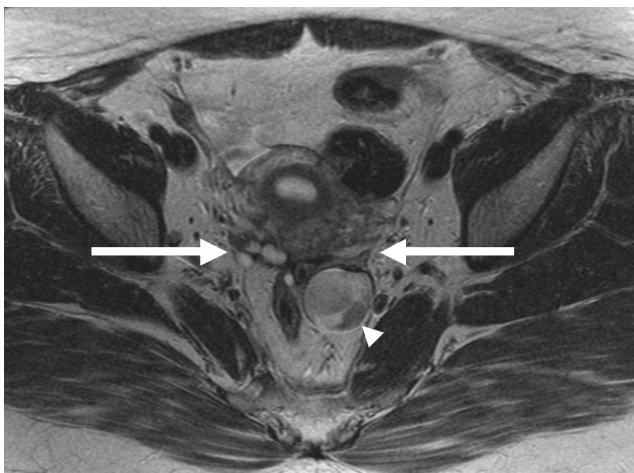


Fig. 1. The axial T2-weighted image (TR/TE, 3716/96) shows multiple small cystic lesions in both adnexae, and this was associated with obliteration and soft tissue lesion of the cul-de-sac (arrows). Note the arrowhead indicating T2 shading of a cystic lesion in the left adnexa.

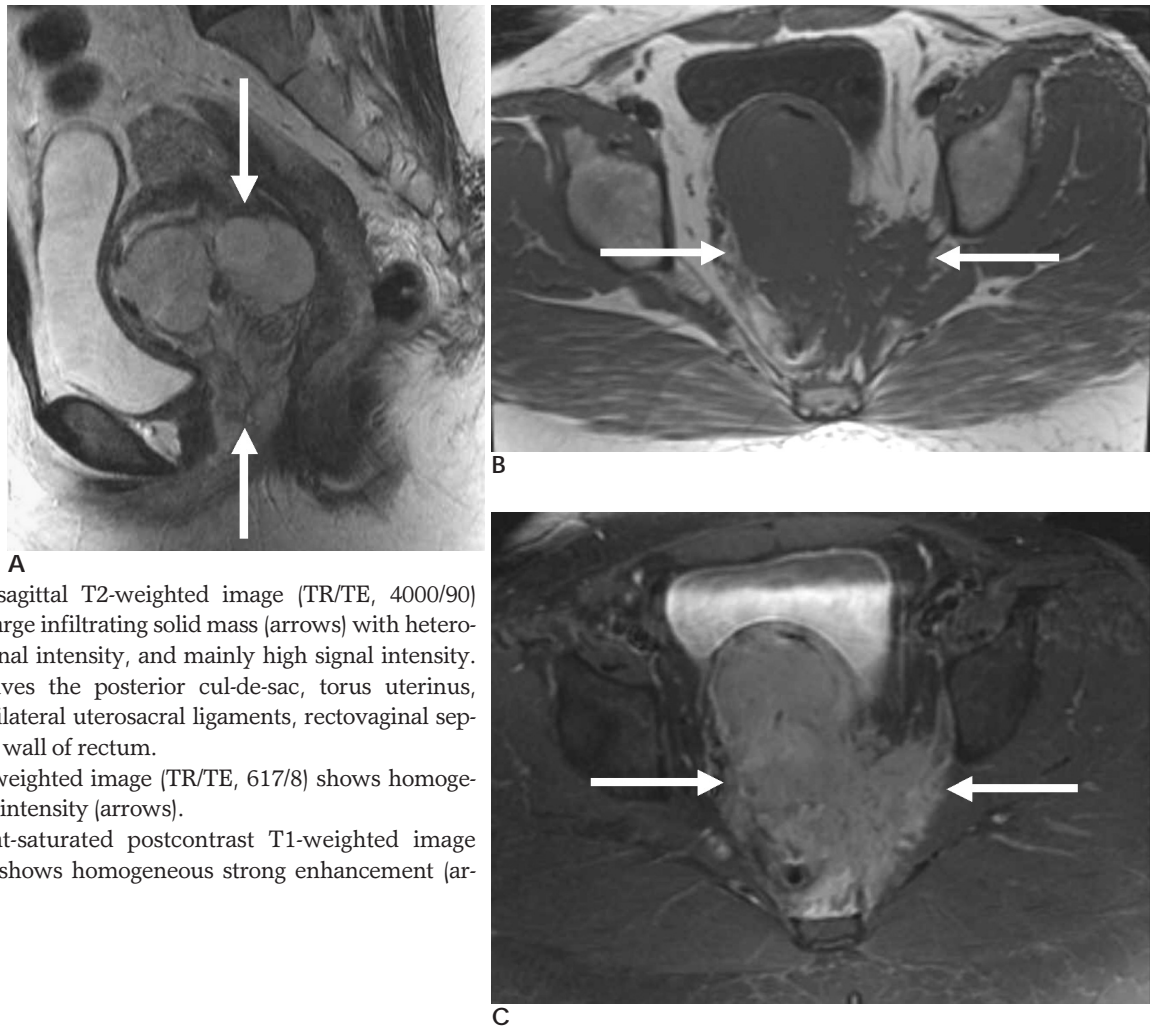


Fig. 2. A. The sagittal T2-weighted image (TR/TE, 4000/90) demonstrates a large infiltrating solid mass (arrows) with heterogeneous high signal intensity, and mainly high signal intensity. This mass involves the posterior cul-de-sac, torus uterinus, cervix, vagina, bilateral uterosacral ligaments, rectovaginal septum and anterior wall of rectum.
B. The axial T1-weighted image (TR/TE, 617/8) shows homogeneous low signal intensity (arrows).
C. The axial fat-saturated postcontrast T1-weighted image (TR/TE, 600/14) shows homogeneous strong enhancement (arrows).

appearance of an ovarian endometriosis-associated carcinoma is that of a unilateral large cystic mass that contains hemorrhagic fluid and mural nodules (9). However, to the best of our knowledge, there are few reports on the MR findings for malignant transformation of deep pelvic endometriosis. Stringfellow (10) et al demonstrated the MRI appearances of extrauterine müllerian adenosarcoma in the right pelvis of a woman who had a history of prior total hysterectomy with bilateral salpingo-oophorectomy due to endometriosis. In their case, the mass showed as a multiloculated pelvic mass with a hemorrhagic component. The MR findings demonstrated heterogeneously high signal intensity on the T2-weighted images and heterogeneously intermediate signal intensity with areas of high signal intensity on the T1-weighted images, and this signal intensity was associated with bony destruction and periosteal reaction.

Our case demonstrated a large, infiltrating extrauterine müllerian adenosarcoma arising from recurrent deep pelvic endometriosis after prior surgery that was

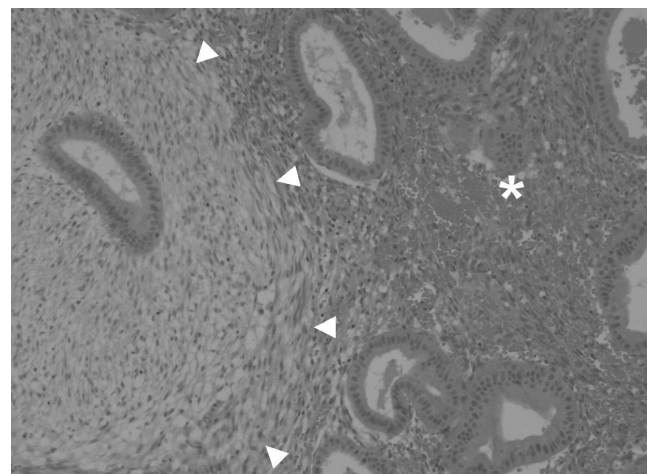


Fig. 3. Photomicrograph of a histopathologic specimen. Endometriosis is identified in the right side of the figure (asterisk), and this shows endometrioid stroma without atypia and a bland looking endometrial gland. In the left side, adenosarcoma (arrowheads) with spindle cell proliferation and increased cellularity, mild cellular atypia and myxoid change is admixed with bland looking endometrial glands.

done to treat pelvic endometriosis. The mass was an ill-defined, large solid mass that was mainly located in the cul-de-sac involving torus uterinus, bilateral uterosacral ligaments, vagina, rectovaginal septum, cervix and anterior rectal wall. MR imaging demonstrated heterogeneously high signal intensity on the T2-weighted images, low signal intensity on the T1-weighted images and homogeneously strong enhancement on the post-contrast T1-weighted images. The signal intensity of the mass on the T2-weighted images in our case was very similar to that of a previous report (10) and the size of the mass in the pelvis was large in the two previously reported cases. The typical MR finding of deep pelvic endometriosis has been uniform low signal intensity on T2-weighted images (8), whereas in our case, the extrauterine müllerian adenosarcoma arising from deep pelvic endometriosis demonstrated heterogeneously high-signal intensity on the T2-weighted images, and mainly a high intensity signal. The reason for this might be suggested that on histopathologic examination, the mass of our case was composed of a large proportion of adenosarcoma with increased cellularity and glandular materials with little fibrotic reaction, which may have resulted in the mainly high signal intensity on the T2-weighted images.

Müllerian adenosarcoma is a tumor with a fair prognosis (2). Surgery is the mainstay of treatment and most tumors can be cured with surgery. When complete removal is impossible, cytoreductive surgery decreases the tumor burden and relieves the symptoms, but further recurrence can occur within a short interval. The rate of recurrence of Müllerian adenosarcoma was reported to be 25 - 30%. A previous study reported that 38% of the patients with müllerian adenosarcoma eventually had recurrent disease (2). Surgery combined with radiation therapy or chemotherapy is not well acknowledged, but this may have efficacy for inoperable or recurrent cases. In our case, complete surgical removal of the mass was impossible and thus, adjuvant chemotherapy and radiation therapy were performed.

In conclusion, the extrauterine müllerian adenosarcoma arising from recurrent deep pelvic endometriosis in our case was a large, infiltrating solid mass that showed heterogeneously high signal intensity on the T2-weighted images, low signal intensity on the T1-weighted images and strong enhancement on the postcontrast-enhanced T1-weighted images. Although it is very rare, if deep pelvic endometriosis shows a large solid mass with aggressive morphology and the MR findings as were mentioned above, then extrauterine müllerian adenosarcoma should be included into the differential diagnosis.

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