

Case Report

Malignant Mixed Müllerian Tumor with Small Bowel Metastasis: A Case Report

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Malignant mixed Müllerian tumors (MMMT) are rare aggressive tumors that typically arise from the female genital tract. This malignancy has an extremely poor prognosis due to its rapid growth and the high associated incidence of both local recurrence and distant metastases. Although intraperitoneal metastasis from MMMT is relatively common, no reports exist regarding the radiologic findings of intestinal metastasis from MMMT. Here, we report a case of MMMT with secondary small bowel metastasis and the associated radiologic findings.

Index words : Malignant mixed Müllerian tumor · Small bowel metastasis · Magnetic resonance imaging (MRI) Computed tomography (CT)

INTRODUCTION

Malignant mixed Müllerian tumor (MMMT), also known as carcinosarcoma, is a rare clinically aggressive malignancy that classically arises from the Müllerian system - the embryonic precursor of the female genital tract and peritoneum. These tumors portend a particularly poor prognosis because of the associated rapid tumor growth and high rates of both local recurrence and distant metastases. Five-year survival rates for MMMT are between 30–40%, even when caught in the earliest stages (1). Notably, MMMT can metastasize to any organ by local extension or via lymphatic, peritoneal, hematogenous spread (2, 3).

Although hematogenous metastasis to the lungs, liver and spine has been reported, lymphatic and intraperitoneal spread is more common (1, 4). To the best of our knowledge, no previous reports exist that illustrate the radiologic findings of small bowel metastasis from uterine MMMT. Here, we report a case of MMMT with small bowel metastasis and describe the associated radiologic findings.

CASE REPORT

A 53-year-old woman presented to our hospital with complaints of abdominal pain, changes in bowel habits and vaginal bleeding. In the past, the patient had a history of a large loop excision transformation zone (LLETZ) for cervical intraepithelial neoplasia I, however denied any previous radiation or hormonal replacement therapy. Physical examination was unremarkable. Laboratory examination revealed an elevated serum CA-125 (47.8 U/mL). Subsequent abdominal and pelvic computed tomography (CT) imaging with a 64-MDCT scanner (Somatom

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Sensation 64, Siemens Medical Solutions, Erlangen, Germany) revealed focal circumferential small bowel wall thickening with central low attenuation and adjacent lymph node enlargement (Fig. 1a). This small bowel lesion was initially believed to represent a lymphoma, as there was no evidence of small bowel obstruction in spite of wall thickening and adjacent lymph node enlargement, while a small bowel mucinous adenocarcinoma with lymph node metastasis was also included in the differential. At this time, a huge, infiltrating solid mass was also noted in the uterus (Fig. 1b). The differential diagnosis for this uterine mass included endometrial carcinoma and sarcomatous changes secondary to uterine myoma. Two days later, the patient underwent pelvic magnetic resonance imaging (MRI) using a 1.5 Tesla MR scanner (Achieva, Philips Healthcare, Netherlands), which showed a large solid mass in the uterus with an irregular outer border extending into the endometrium and uterine cervix but not the parametrium. Notably, the lesion demonstrated heterogeneous high signal intensity on T2-weighted images, low signal intensity with internal high signal intensity on the T1-weighted image, and poor enhancement on the post-contrast T1-weighted images (Fig. 2).

The patient underwent exploratory laparotomy with total abdominal hysterectomy, bilateral salpingo-oophorectomy, and segmental small bowel resection. In the resulting surgical specimen, an irregular yellow-colored flesh mass with hemorrhage and necrosis was found in the uterus, involving the endomyometrium,

both ovaries, and cervix (Fig. 3). No parametrial invasion was observed. Microscopic examination of the small bowel specimen showed a nesting pattern growth of cytokeratin-positive tumor cells (poorly differentiated adenocarcinoma) and a diffuse growth pattern of poorly differentiated short spindle cells (sarcoma) without a heterologous component. The final diagnosis of uterine MMMT with small bowel metastasis was made after pathologic examination. The patient received six cycles of taxol and carboplatin chemotherapy, however she developed distant metastasis to cervical lymph nodes, pelvic bone, and liver. Five month after the surgery, the patient expired because of septic shock and neutropenic fever.

DISCUSSION

MMMT is a rare gynecological malignancy arising from the Müllerian system, the precursor to the female genital tract. Previous pelvic irradiation represents the most well characterized epidemiologic risk factor for MMMT, with several studies showing that between 7% and 37% of women who develop MMMT have a history of previous pelvic irradiation (5). Most other risk factors for MMMT are similar (but weaker) to those for routine endometrial adenocarcinoma, including prolonged exposure to estrogen via hormone replacement therapy, obesity, nulliparity, and polycystic ovarian disease (6). While MMMT most commonly arises in the uterus, it can also occur in the

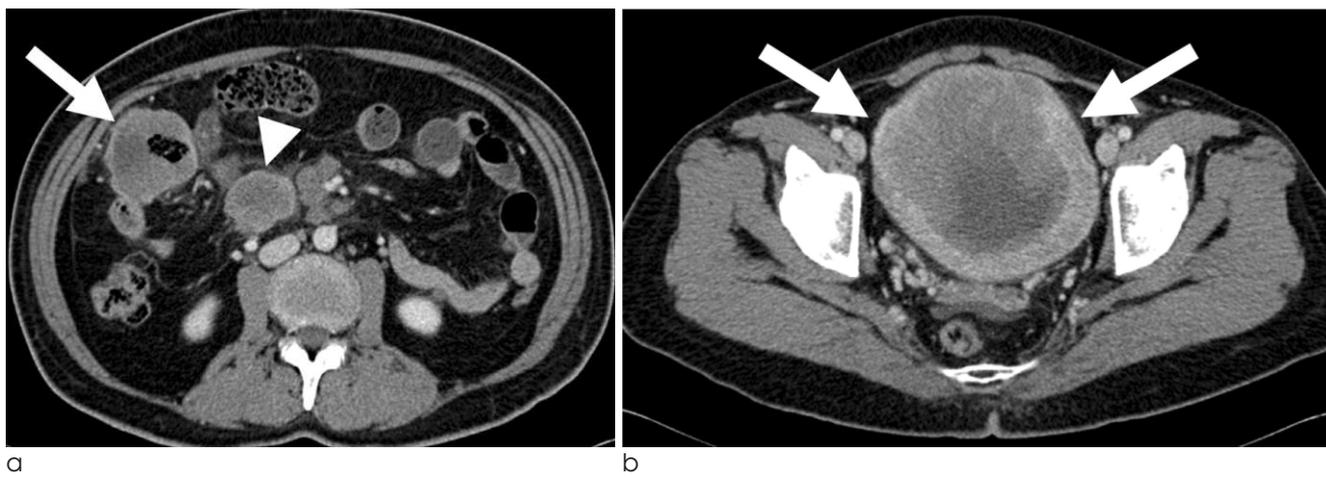


Fig. 1. a. Contrast-enhanced axial CT image shows irregular wall thickening of the small bowel (arrow) with enlarged mesenteric and retroperitoneal lymph nodes (arrowhead).
b. Contrast-enhanced axial CT scan at the pelvic level shows a poorly enhancing mass in the uterus. The mass extends into the endometrial cavity.

ovaries, fallopian tubes, vagina and peritoneum (7). As MMT histologically contains both carcinomatous and sarcomatous elements, the tumor can spread both

via lymphatic channels and blood vessels (3). Reported metastatic sites have included the abdomen, pancreas, liver, lung, thyroid gland, heart, and the

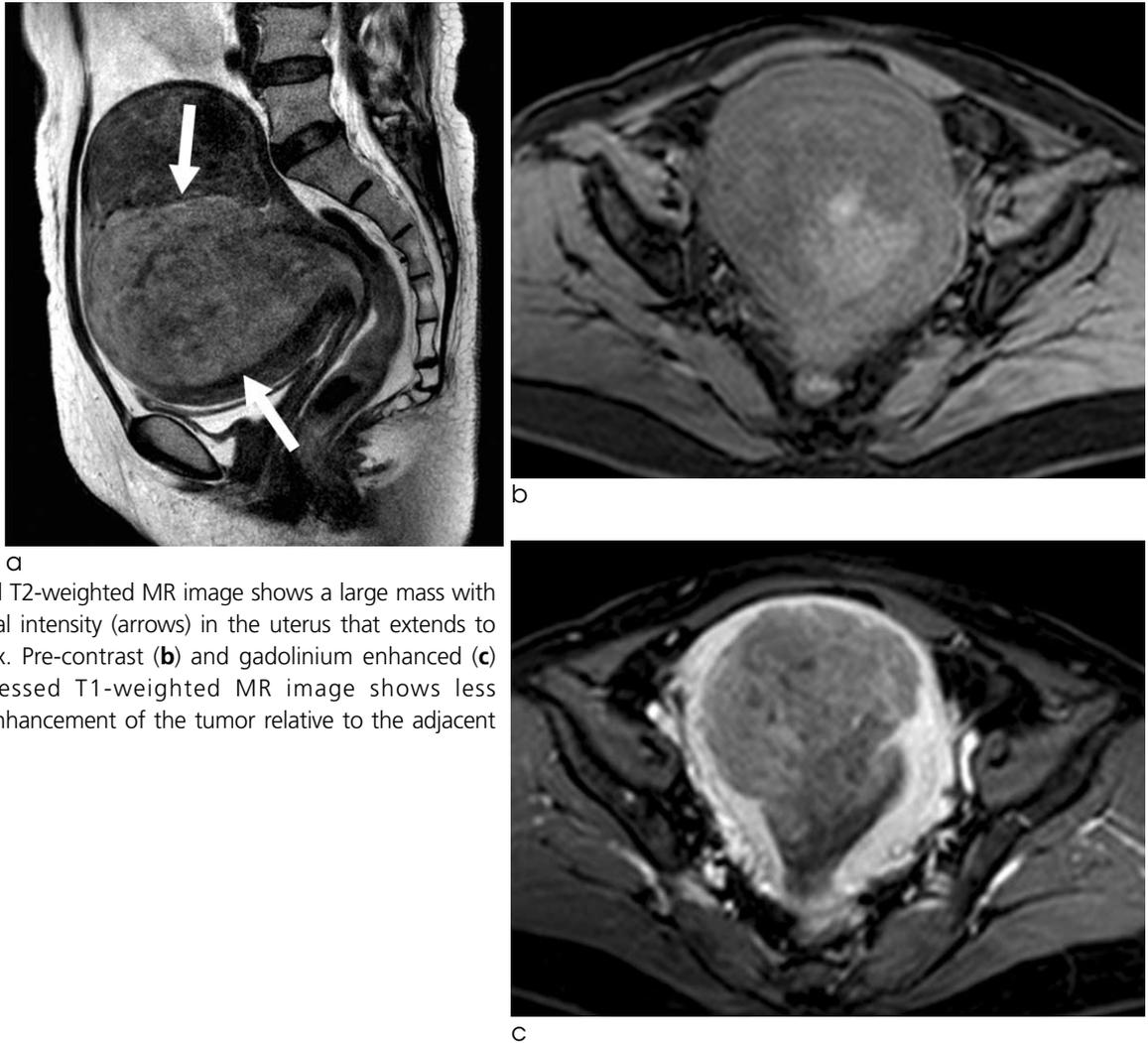


Fig. 2. (a) Sagittal T2-weighted MR image shows a large mass with slightly high signal intensity (arrows) in the uterus that extends to the uterine cervix. Pre-contrast (b) and gadolinium enhanced (c) axial fat-suppressed T1-weighted MR image shows less heterogeneous enhancement of the tumor relative to the adjacent myometrium.

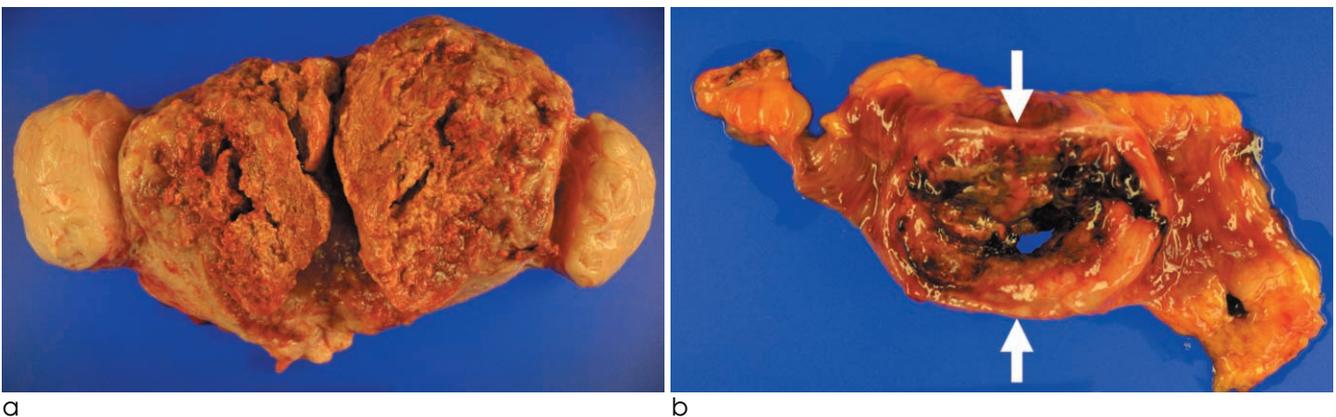


Fig. 3. a. A photograph of the gross uterine specimen shows a poorly circumscribed infiltrative yellow, solid mass in the endomyometrium with invasion of more than one half of the myometrium. b. The gross small bowel specimen shows an irregular mass (arrows) that is adherent to the serosal surface of the ileum.

orbit, with more than half of patients presenting with metastases at the time of diagnosis (4). Interestingly, intestinal metastasis is relatively rare in MMMT, and only a few cases of metastasis from uterine MMMT to the gastrointestinal tract have been documented in the literature (2). In the case presented here, the lesions in the small bowel and uterus were initially diagnosed separately (as small bowel lymphoma and either endometrial carcinoma or sarcomatous changes secondary to uterine myoma) for two reasons: the patient had no prior history of radiation or hormonal replacement therapy, and small bowel MMMT metastasis is rare.

The CT and MRI appearances of MMMT and any associated metastases can be varied, and occasionally are indistinguishable from routine endometrial adenocarcinoma (8). Per the previously published literature, MMMT most commonly appears on contrast enhanced CT scans as heterogeneous masses with peripheral rim enhancement and central low attenuation, thus indicating central necrosis, focal lack of blood supply, and rapid tumor growth (2). On T2-weighted MR imaging, MMMT classically presents as a heterogeneous hyperintense mass, while T1-weighted MR images often reveal a slightly hypointense mass with scattered areas of high signal intensity (indicating subacute hemorrhages). On gadolinium enhanced T1-weighted images, such masses often are heterogeneous but show marked enhancement (9). Such imaging findings are comparable with the case presented here, with the exception of the enhancement pattern on gadolinium enhanced T1-weighted imaging, where a poorly enhancing mass was seen.

On contrast enhanced CT, the small bowel metastasis showed irregular circumferential bowel wall thickening with internal homogenous low attenuation, corresponding to the necrotic and hemorrhagic content seen in the surgical specimen. Because no evidence of small bowel obstruction was observed, despite the marked bowel wall thickening, such findings may mimic small bowel lymphoma. As such, mucinous adenocarcinoma should also be included in the differential diagnosis since these lesions would show homogenous hypoattenuation of the thickened bowel wall (10).

The diagnosis of MMMT is confirmed by pathologic review of biopsy or surgical specimen. Immunohistochemistry may be helpful in providing the origin for

the sarcomatous and carcinomatous components of these neoplasm (11).

The primary treatment for uterine MMMT is total hysterectomy and bilateral salpingo-oophorectomy, with or without lymph node dissection. However, the high rates of both local and distant recurrences after this surgery have highlighted the need for effective adjuvant treatment with chemotherapy and radiotherapy. Even with a multidisciplinary therapeutic approach including surgery, radiotherapy, and various chemotherapy regimens, outcomes are uniformly poor because of the rapidly fatal course and high rate of distant recurrence associated with this malignancy (1).

In conclusion, we report a case of MMMT with small bowel metastasis in a patient with no prior history of radiation or hormonal replacement therapy. Despite the rare incidence, small bowel metastases should be considered in the differential for any case of suspected uterine MMMT where irregular small bowel wall thickening and internal low attenuation is seen on CT.

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소장전이를 보이는 악성 물러리안 종양: 증례 보고

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악성 혼합 물러리안 종양은 여성생식기에서 발생하는 매우 드문 종양으로 생물학적으로 공격적인 성향을 보인다. 이들은 대개 빠르게 성장하며 국소적 재발 및 원격 전이의 빈도가 높아 예후가 극도로 불량하다. 악성 혼합 물러리안 종양은 복강 내 전이가 상대적으로 흔하지만 장관 전이의 영상 소견에 관한 보고는 아직 없다. 저자들은 소장으로 전이한 악성 혼합 물러리안 종양 1예와 그 영상 소견에 대하여 보고하고자 한다.

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