Case Report
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Malignant Mixed Mullerian Tumor Arising from the Uterine Cervix: A Case Report[1]

자궁경부에서 발생한 악성혼합뮐러종양: 증례 보고[1]

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Malignant mixed mullerian tumors (MMMTs) are a rare uterine tumor and contribute to approximately 1-3% of all corpus malignant tumors. MMMTs are usually in the uterine corpus, but can also arise from the uterine cervix, vagina, ovaries and fallopian tubes. MMMTs of the uterine cervix are extremely rare. MMMTs are highly malignant and tend to maintain a rapid growth and exhibit a high rate of recurrence. Therefore, the prognosis of patients diagnosed with these types of tumors is extremely poor. We report a rare case of a malignant mixed mullerian tumor arising from the uterine cervix and introduce CT and MRI findings. CT and magnetic resonance findings of the uterine cervical MMMT in our case show highly aggressive features, such as parametrial involvement, pelvic and paraaortic lymphadenopathy, and distant metastasis and high enhancement.

Index terms
Malignant Mixed Mullerian Tumor
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INTRODUCTION

Malignant mixed mullerian tumors (MMMTs) are rare biphasic malignant neoplasm with two components of carcinoma and sarcoma. The most common site of occurrence in the female genital tract is the uterine corpus (1). The clinicopathologic and radiologic characteristics of tumor and treatment are uncertain due to the lack of clinical data. We report one case of a patient diagnosed with a malignant mixed mullerian tumor, arising from the uterine cervix.

CASE REPORT

A 54-year-old woman was presented with intermittent vaginal spotting over a month period of time. Upon pelvic examination, a mass was identified in the cervix. The serum CA-125 level was 5020 U/mL (normal range: 0-35 U/mL).

Pelvis MRI was performed, and a large lobulated mass was identified at the uterine cervix. The mass exhibited low signal intensity on T1 weighted image and heterogenous slight high signal intensity on T2 weighted image (Fig. 1A). On axial T2 weighted image, the margin of cervix was irregular, and the mass invaded the right periureteric parametrium. Further, right hydroureter was also noted (Fig. 1B). Thus, we thought that the mass had invaded parametrium. Following IV contrast enhancement, the mass demonstrated a heterogenous strong enhancement. On an enhanced sagittal image, the epicenter of the mass was located at the uterine cervix and involved the lower uterine body. Thus, it seemed that the mass was arised from the uterine cervix (Fig. 1C). Therefore, the initial was stage IIIB uterine

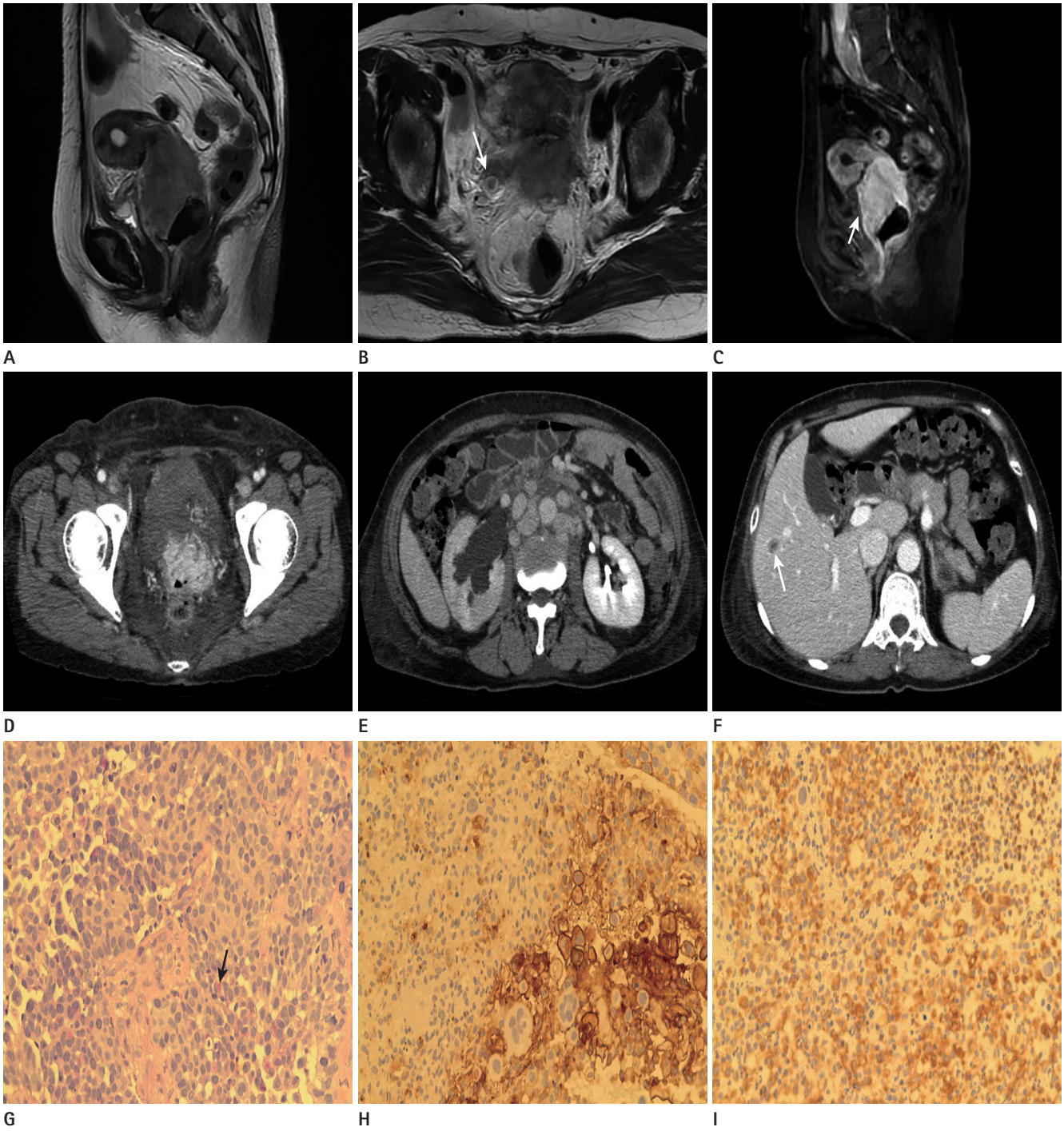

Fig. 1. Imaging and pathologic findings of 54-year-old woman with uterine cervical MMMT.
A. Sagittal T2 weighted image demonstrates large lobulated mass with Heterogenous slightly high signal intensity at the uterine cervix.
B. On axial T2 weighted image, low signal intensity cervical stoma is not visible. Right periureteric parametrial invasion and right hydroureter are also noted (arrow).
C. Gadolinium enhanced T1 weighted image shows strong enhancement of the mass and the epicenter of mass is located at the uterine cervix (arrow).
D. Contrast enhanced axial CT scan demonstrates highly enhanced uterine cervical mass.
E. On delayed scan, right hydronephrosis and metastatic paraaortic lymph nodes are identified.
F. Low attenuation metastatic nodule in liver is also noted (arrow).
G. On PAS staining (× 20), several cytoplasmic staining are identified (arrow).
H, I. The specimen shows high positivity on CEA (× 20) **(H)**, and vimentin (× 20) **(I)** staining.
Note.—CEA = carcinoembryonic antigen, MMMT = malignant mixed mullerian tumor, PAS = Periodic Acid-Shiff

cervical cancer.

For the evaluation of distant metastasis, abdominopelvic CT was performed. On an enhanced scan, highly enhanced uterine cervical mass was revealed (Fig. 1D). And right hydronephrosis and multiple metastatic paraaortic lymph nodes were also identified (Fig. 1E). Furthermore, several multiple hepatic metastatic nodules were also identified (Fig. 1F). Based on these results, the diagnosis was changed to uterine cervical cancer stage IVB.

Next, a colposcopic punch biopsy was performed. On a Periodic Acid-Shiff staining (Fig. 1G), several cytoplasmic stainings were identified. It means the presence of mucin, adenocarcinoma component, was revealed. Immunohistochemical tests were also performed and the pathologic specimen showed high positivity on carcinoembryonic antigen (carcinoma marker) and vimentin (sarcoma marker) staining (Fig. 1H, I). Thus, foci of squamous carcinomatous, adenocarcinomatous and sarcomatous components were all identified. So, the pathologic diagnosis was malignant mixed mullerian tumor, originated from the uterine cervix.

DISCUSSION

Malignancies of the cervix tend to almost always be carcinomas. Approximately 80-90% of these are squamous cell carcinomas, while 10-20% are adenocarcinomas, and the remaining are adenosquamous carcinoma, sarcoma, melanoma, lymphoma, and metastatic tumors. MMMTs of the uterine cervix represent a rare form of all cervical cancers (2).

MMMTs are biphasic tumors that consist of an admixture of malignant epithelial and mesenchymal components. The epithelial component represents a variety of different histologic sub-types, alone or in combination, which include squamous cell carcinoma, basaloid squamous carcinoma, adenocarcinoma, adeno-squamous carcinoma, adenoid-basal carcinoma, adenoid-cystic carcinoma and undifferentiated carcinoma. The sarcomatous component may be homologous (fibroblasts and smooth muscle) or heterologous (cartilage, striated muscle, bone etc.). On an immunohistochemical examination, both epithelial and sarcomatous components of MMMT may show positivity for broad spectrum cytokeratins, high molecular weight cytokeratin, low molecular weight cytokeratin and epithelial membrane antigen. Sarcomatous components may be positive for vimentin, desmin, muscle specific actin and smooth muscle-specific actin (3).

MMMT is usually in the fundus. But MMMTs can arise anywhere along the mullerian axis and have a high incidence of lymphatic spread, peritoneal seeding, and higher rate of pulmonary metastases than other uterine malignancies (4).

The largest single study of 9 cases of uterine cervical MMMTs revealed several key features: the age at presentation varied widely (range, 23-87 years; mean, 65 years), abnormal vaginal bleeding was the most common presenting symptom, the tumors frequently displayed non-glandular epithelial components, and may be associated with better outcome than MMMTs of the uterine corpus (5).

Although it is difficult to determine the optimal therapy for patients with cervical MMMT, overall patients with low stage disease (IB1 and IB2) had a good outcome with therapy, traditionally used for patients with squamous cell carcinoma of the cervix (2).

Magnetic resonance (MR) imaging findings of MMMT from the uterine cervix have not been reported to our knowledge. Bharwani et al. (4), described MRI appearance of the uterine MMMTs. In their study, 88% of MMMTs, the tumor epicenter was endometrial; whereas, 4% of tumors had a myometrial epicenter and 8% of tumors had a cervical epicenter. On T1-weighted images, the majority of MMMTs were isointense to myometrium (76%) and endometrium (71%). On T2-weighted images, 92% of MMMTs were hyperintense to myometrium and either hypointense (55%) or isointense (41%) to endometrium. MMMT texture on T2-weighted images was heterogenous in 82%. In the study by Teo et al. (6), two MMMTs showed intense, but heterogenous enhancement after gadolinium administration; whereas, 80% of patients in the study by Tanaka et al. (7) showed area of avid enhancement.

In our case, the mass shows slightly heterogenous high signal intensity on a T2-weighted image and is well enhanced on a T1-weighted image after injection of gadolinium contrast material. So the mass is similar to MMMT of the uterine corpus, except the mass is arised from the uterine cervix.

On T1-weighted images, cervical carcinomas are usually isointense to the normal cervix and may not be visible. On T2-weighted images, cervical cancer appears as a relatively hyperintense mass and is easily distinguishable from that of low sig-

nal-intensity cervical stroma. On dynamic contrast-enhanced MRI, small tumors enhance homogeneously and earlier than the normal cervical stroma. Large tumors are frequently necrotic and may or may not enhance, but are often surrounded by an enhancing rim that facilitates tumor definition (8, 9). On a CT, the cervical cancer can be hypoattenuating or isoattenuating to normal cervical stroma after administration of intravenous contrast material (10). In our case, however, the mass showed heterogenous high enhancement after IV contrast enhancement on an MRI and CT.

CT and MR findings of uterine cervical MMMT, in our case, show highly aggressive features, such as parametrial involvement, pelvic and paraaortic lymphadenopathy, and distant metastasis and high enhancement. However, these findings are not particularly different from the other types of cervical carcinoma. Therefore, it could not be differentiated from other types of cervical carcinoma.

In conclusion, the imaging findings of uterine cervical MMMT of our case are not pathognomic. But, imaging findings of MMMT, arising from the uterine cervix, have not been reported in our knowledge; As such, we introduce CT and MR imaging findings of MMMT arising from the uterine cervix.

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자궁경부에서 발생한 악성혼합뮐러종양: 증례 보고[1]

심종준[1] · 심재찬[1] · 이혜경[2] · 이경은[1] · 이기재[1] · 김호균[1] · 서정호[1]

악성혼합뮐러종양은 악성자궁종양의 1~3%로 발생빈도가 낮다. 주로 자궁체부에서 발생하며 자궁경부, 질, 난소 및 난관에서도 드물게 발생한다. 악성혼합뮐러종양이 자궁경부에서 발생하는 경우는 매우 드물다. 악성혼합뮐러종양은 매우 악성도가 높은 종양으로 빠른 성장을 하고 전이 및 재발이 잘되어 예후가 불량하다. 저자들은 자궁경부에서 발생한 악성혼합뮐러종양의 증례에 대해 보고하고 CT 및 MRI 소견을 소개하고자 한다. CT와 MRI에서 이 종괴는 자궁주위 조직과 골반 및 대동맥 주위 림프절을 침범하는 등 침습적인 양상을 보였으며 강한 조영증강 및 원격전이를 보였다.

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