Magnetic resonance imaging findings of a myxoid leiomyosarcoma of the uterus: A case report and literature review

Takayoshi Shinya a,*, Kentaro Shibamoto a, Kiyoka Maeba a, Katsuya Kato a, Yasumasa Monobe b, Michihisa Fujiwara c, Atsushi Hongo c

a Department of Diagnostic and Therapeutic Radiology, Kawasaki Medical School General Medical Centre, 2-6-2 Nakasange, Kita-ku, Okayama City, Okayama, 700-8505, Japan
b Department of Pathology, Kawasaki Medical School General Medical Centre, 2-6-2 Nakasange, Kita-ku, Okayama City, Okayama, 700-8505, Japan
c Department of Obstetrics and Gynecology 2, Kawasaki Medical School General Medical Centre, 2-6-2 Nakasange, Kita-ku, Okayama City, Okayama, 700-8505, Japan

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ABSTRACT

Uterine myxoid leiomyosarcomas (MLMSs) are extremely rare. Here, we report a rare case of uterine MLMS with unique and bizarre magnetic resonance imaging (MRI) findings on diffusion-weighted images (DWIs) and dynamic contrast-enhanced (DCE) MRI scans. A 67-year-old woman presented with a uterine MLMS that had a multilocular cystic mass with a septum and solid components. The tumour demonstrated marked hyperintensity on T2-weighted images in a myxoid stroma with gradual partial contrast enhancement and diffusion restriction, which could be a characteristic feature suggestive of a myxoid malignant smooth muscle tumour of the uterus rather than a uterine leiomyoma with myxoid degeneration.

1. Introduction

Myxoid changes in uterus leiomyomas (ULs) are not rare findings and have a reported incidence of 10% [1]. However, uterine myxoid leiomyosarcoma (MLMS) is an aggressive and extremely rare variant of leiomyosarcomas [2]. On magnetic resonance imaging (MRI), ULs with myxoid degeneration has a distinctive appearance with markedly high signal intensity (SI) on T2-weighted images (T2WIs) and enhanced well except for small foci of mucinous lakes or clefts. Delayed and prolonged enhancement can be observed because of the presence of a myxoid stroma [3]. In the literature, to the best of our knowledge, reported MRI features of uterine MLMS have been limited to two cases with T2WIs and contrast-enhanced T1-weighted images (T1WIs) [4,5]. Therefore, MRI features of uterine MLMS have not been completely investigated; in particular, features on diffusion-weighted images (DWIs) and dynamic contrast-enhanced (DCE) MRI scans have not been reported in patients with uterine MLMS. Herein, we report a rare case of uterine MLMS that had unique and bizarre MRI findings on DWIs and DCE MRI. We also present a brief a literature review.

2. Case presentation

A 67-year-old woman was referred to our radiology department for the examination and further assessment of a pelvic mass detected using ultrasonography. The patient had complained of fatigue and nausea since 1 week before presenting to the hospital. Physical examination revealed slight tenderness in the lower abdomen. Laboratory data and tumour markers, including carbohydrate antigen (CA) 125, CA72-4, carcinoembryonic antigen, and CA19-9, were within normal limits. MRI revealed a huge uterine mass measuring 175 mm × 99 mm × 127 mm and involving the uterine corpus almost entirely and the uterine cervix partially, as well as a small myoma in the posterior uterine wall without metastatic lesions (Fig. 1a). The tumour mainly had a markedly heterogeneous hyperintense area with hypointense reticular structures partially on T2WI (Fig. 1a, b). The tumour had a hypointense area and a slightly hyperintense area compared with that of the muscle on T1WIs (Fig. 1c), and no fat component was visible on fat-saturated T1WIs (Fig. 1d). On DCE MRI scans (Fig. 1e) and late contrast-enhanced T1WIs (Fig. 1f), the tumour had gradual and heterogeneous contrast enhancement in the low SI region on T2WIs corresponding to the septum and solid parts within the tumour and in parts of the high SI region on T2WIs corresponding to myxoid degeneration.

* Corresponding author at: 2-6-2 Nakasange, Kita-ku, Okayama City, Okayama, 700-8505, Japan.
E-mail address: midnight-2005@nifty.com (T. Shinya).

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These areas had slightly increased SI region on DWIs (Fig. 1g) and restriction of diffusion on apparent diffusion coefficient (ADC) maps (Fig. 1h) within the myxoid degeneration where contrast enhancement was observed. On the other hand, no contrast enhancement was observed in the central cystic parts of the high SI region on T2WIs with low SI and slightly high SI on T1WIs, which corresponded to myxoid degeneration with partially haemorrhagic necrosis. Based on the MRI findings, MLMS, myxoid leiomyoma (LM), and smooth muscle tumours of uncertain malignant potential were the preoperative differential diagnoses. However, distinguishing these tumours precisely based on MRI findings was challenging.

Tumour resection with total hysterectomy and bilateral oophorectomy was performed. On the cut section, the uterine tumour was an 18-cm-sized well-circumscribed tumour with a gelatinous surface. The tumour was composed of yellowish, partly dark red foci of haemorrhage, and gelatinous texture deposits separated by fibrous septum (Fig. 2).

Microscopically, the spindle-shaped cells with ovoid or elongated nuclei and eosinophilic cytoplasm were arranged into irregular bundles or separated by myxoid stroma. The tumour invaded the neighbouring myometrium via vascular invasion. Mitotic figures were found at a rate of 2–5 per 10 high-power fields (Fig. 3a, b). Immunohistochemical analysis revealed tumour cells to be positive for calponin, α-smooth muscle actin, oestrogen receptor, p16, and CD10, and focally positive for desmin, S100 protein, and p53. Tumour cells were not immunoreactive for progesterone receptor, CD34, and ALK. MIB-1 immunostaining showed positive nuclear staining in approximately 26% of cells. These pathological findings resulted in the diagnosis of a myxoid leiomyosarcoma of the uterus.

3. Discussion

Uterine MLMS is a rare malignant counterpart of myxoid LM, which
has been rarely reported in the literature. Despite its deceptively benign histologic appearance and a low mitotic count, uterine MLMS can metastasise, and unexpected malignant transformations of pre-existing ULs have been reported [4,6]. Additionally, MLMSs are more aggressive and have worse 5-year overall and disease-free survival rates than conventional leiomyosarcomas [2,7,8].

In a previous report, areas of myxoid degeneration within a UL showed heterogeneous and marked hyperintensity on T2WIs, with progressive enhancement developing after contrast administration except for small foci of mucinous lakes or clefts [3]. Regarding uterine MLMS, Holzmann et al. demonstrated that main parts of the large tumour had hyperintense cystic areas on T2WIs and the solid part with contrast enhancement was within the central part of the tumour [4]. In contrast, Shintaku et al. demonstrated that the large uterine MLMS had contrast enhancement in the most of the hyperintense regions on T2WIs [5]. In the present case, the uterine MLMS had the hyperintense regions on T2WIs, which corresponded to myxoid degeneration, with or without gradual and prolonged contrast enhancement, and the septum and solid parts showed hypointense signals on T2WIs with gradual contrast enhancement on DCE MRI scans. We suspected that the extent of the vascular network within myxoid degeneration in the UL and uterine MLMS was responsible for the difference in enhancement patterns in hyperintense regions on T2WIs as extraperitoneal soft tissue tumours with myxoid stroma [9]. However, there have been no published reports on uterine MLMS with DCE MRI findings, and typical and detailed enhancement patterns of uterine MLMS remain unclear.

DWsIs shows tissue characteristics based on the diffusion motion of water molecules. In general, DWIs can delineate malignant lesions as a hyperintense area with excellent tissue contrast, and cases with hypointense signals on DWIs may be regarded as benign lesions in smooth muscle tumours of the uterus [10,11]. In contrast, previous studies have reported that there were overlaps in ADC values between a UL, such as cellular LM and bizarre LM, and uterine sarcoma, suggesting that DWIs alone are not sufficient to make a definite diagnosis [11–15]. Regarding uterine smooth muscle tumours with myxoid degeneration, UL with myxoid degeneration has no restriction on DWIs and ADC maps [16]. A previous study reported a uterine LMS with an intermediate SI on DWI associated with mucous degeneration without any imaging feature of the case and revealed that uterine smooth muscle tumours with intermediate-to-high SI may indicate uterine leiomyosarcoma [11]. The present case revealed high SI on DWIs for myxoid stroma with contrast enhancement. The MRI findings in the present case suggest that DWIs and ADC values can be potentially useful for differentiating MLMS from myxoid LM, and this needs to be assessed in further research in the future.

In conclusion, typical imaging findings of uterine MLMS include a multilocular cystic appearance and tumour with markedly high SI on T2WIs with gradual enhancement on DCE MRI scans. High SI on DWIs may be a potential marker in differentiating myxoid LM from MLMS. Therefore, it is critical to consider the possibility of uterine MLMS when examining uterine cystic tumours with high SI on T2WIs with gradual enhancement on DCE MRI scans and high SI on DWIs.
4. Informed consent and patient details

This study does not require institutional review board approval. Informed consent was obtained for the case report to be published.

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Declaration of Competing Interest

None of the authors have any relevant conflict of interest or industry support related to this report.

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Appendix A. Supplementary data

Supplementary material related to this article can be found, in the online version, at doi: https://doi.org/10.1016/j.ejro.2021.100328.

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