

A Case of Myxoid Leiomyosarcoma of the Uterus : MR Findings

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Myxoid leiomyosarcoma of the uterus is an extremely rare disorder that has only been documented previously with regard to clinical and pathological aspects. We describe a 50 year-old woman with myxoid leiomyosarcoma of the uterus presenting with polyhypermenorrhea. The serum level of CA-125 (normal equal or less than 35 IU/ml) was elevated at 130 IU/ml. Magnetic resonance imaging revealed multiple, well-defined masses involving the myometrium. The masses showed heterogeneous low signal intensity on T₁-weighted images and marked high signal intensity with septations on T₂-weighted images. On Gd-DTPA contrast enhanced T₁-weighted images, the masses demonstrated minimal enhancement with a signal intensity slightly higher than that of water. Based on these findings, a myxoid-degenerated leiomyoma was suspected. Seven weeks thereafter, however, some of the masses ruptured into the peritoneal cavity requiring emergency laparotomy. Microscopic pathological examination of tissue samples obtained during surgery revealed myxoid leiomyosarcoma of the uterus with a high mitotic count. Differential diagnosis between myxoid leiomyosarcoma and myxoid-degenerated leiomyoma is not possible by means of MR imaging alone. The serum level of CA-125 may be of value for differential diagnosis of this disorder.

INTRODUCTION

The incidence of myxoid degeneration has been reported to be as high as 12% for benign leiomyoma¹⁾ of the uterus. Myxoid change in uterine leiomyosarcoma (myxoid leiomyosarcoma), however, was first described in 1982 by King et al.²⁾ Since then, a few additional cases have been reported, all of which were docu-

mented on the basis of clinical and pathological findings^{3)~5)}. Magnetic resonance (MR) imaging of leiomyosarcoma of the uterus has been previously reported^{6),7)}. To our knowledge, however, MR features of myxoid leiomyosarcoma has not as yet been reported. In this case report, we describe the MR features of myxoid leiomyosarcoma of the uterus.

Keywords myxoid leiomyosarcoma, uterine neoplasm, MRI, CA-125

CASE REPORT

A 50-year-old woman presented with polyhypermenorrhea. Physical examination revealed a distended abdomen and a huge, ill-defined, soft, nontender mass was palpable. The serum level of CA-125 was elevated at 130 IU/ml (normal less than or equal to 35 IU/ml). Microscopic examination of scrapings from the endocervix showed no malignancy. Ultrasonography revealed an enlarged uterus consistent with leiomyoma of the uterus. Magnetic resonance (MR) imaging showed multiple well-defined masses that extended to the anterior myometrium (Fig. 1). The masses showed heterogeneous low signal intensity on

T₁-weighted images (TR/TE=600/15 ms) and marked high signal intensity with septations on T₂-weighted images (TR/TE=2000/70 ms). On Gd-DTPA enhanced T₁-weighted images (TR/TE=600/15 ms), the masses exhibited minimal enhancement with slightly higher signal intensity than water and the septations showed enhancement equal to that of the myometrium. Based on these findings, myxoid-degenerated leiomyoma was suspected and surgery was recommended. Seven weeks thereafter, the patient developed signs of an acute abdomen and lower abdominal fullness. CT showed a large amount of ascitic fluid with heterogeneous high density in part suggesting bloody ascites (Fig. 2a). On contrast-enhanced

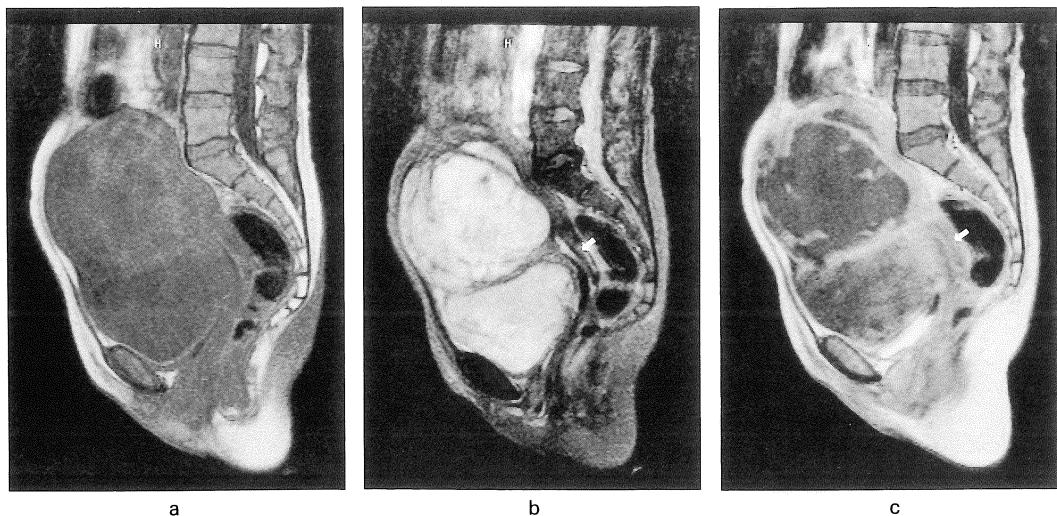


Fig. 1. MR images

Magnetic resonance imaging reveals multiple well-defined masses continuous to the anterior myometrium. The mass shows (a) heterogeneous low signal intensity on T₁-weighted images (TR/TE=600/15 ms) and (b) marked high signal intensity with septations in T₂-weighted images (TR/TE=2000/70 ms). On (c) Gd-DTPA contrast-enhanced T₁-weighted images (TR/TE=600/15 ms), the mass demonstrates minimal enhancement with slightly higher signal intensity than that of water and the septations show enhancement equal to that of the myometrium (arrow : cervical epithelium).

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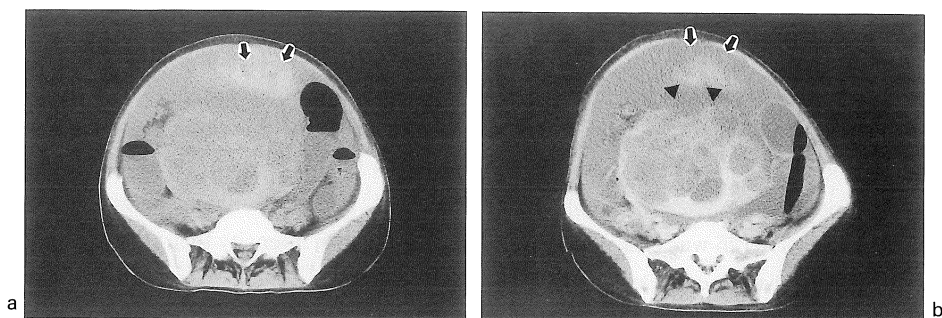


Fig. 2. CT images

CT images ((a) non-contrast (b) contrast enhanced) show a large amount of ascitic fluid with heterogeneous high density (arrow) indicating bloody ascites. Upon contrast-enhanced CT, the anterior contours of the uterine masses (arrowhead) are irregular and ill-defined, for which reason there is suspicion that some of the masses have ruptured into the peritoneal cavity.

CT, the anterior contours of the uterine masses were found to be irregular and ill-defined, upon which basis rupture of some of the masses into the peritoneal cavity was suspected (Fig. 2b). Emergency laparotomy was performed, at which time bloody ascites and rupture of some of the masses into the peritoneal cavity could be appreciated. A total hysterectomy and bilateral salpingo-oophorectomy were performed. Macroscopically, the anterior myometrium was occupied with multiple well-

defined masses, the cut surface of which was yellowish and gelatinous (Fig. 3). Microscopic examination revealed pleomorphic smooth muscle cells with 10 to 20 mitoses per 10 high-power fields with invasion to the surrounding myometrium and endometrium with abundant myxoid matrix distributed diffusely throughout (Fig. 4), upon which basis a diagnosis of myxoid leiomyosarcoma of the uterus was made. The septations seen in MR images correspond-

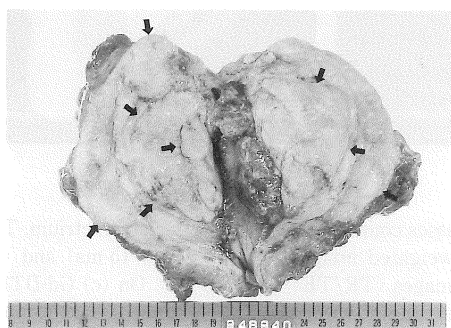


Fig. 3. Macroscopic view of the specimen
The cross section of the resected tumor demonstrates multiple well-defined masses (arrow) occupying the anterior myometrium and the cut surface appears yellowish and gelatinous.

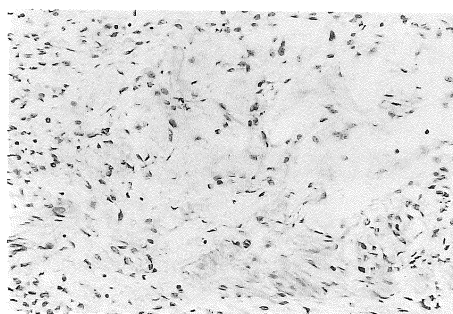


Fig. 4. Microscopic view of the specimen (H & E stain, 200 \times)

Microscopic examination of the mass shows pleomorphic smooth muscle cells with 10 to 20 mitoses per 10 high-power field and abundant myxoid matrix diffusely distributed throughout.

ed to a compressed normal myometrium. Chest and abdominal CT studies showed no distant metastases at the time.

The patient was treated with adjuvant chemotherapy (Cisplatin, Pirarubicin) after which the serum level of CA-125 decreased to 18 IU/ml. Seven months after the surgery, multiple metastases to the lungs, liver and abdominal lymph nodes were detected and the serum level of CA-125 had increased to 290 IU/ml. The patient subsequently died ten months after the initial surgery.

DISCUSSION

Uterine leiomyosarcoma has been diagnosed microscopically based on the number of mitoses per high-power field (hpf) by pathological examination, with 5 or more mitoses per 10 hpf being the criterion most widely used^{1,2}. A tumor with fewer than 5 mitoses per 10 hpf is generally categorized as cellular myoma which is composed of densely packed cellular fascicles of smooth muscle with little intervening collagen. The 6 cases described by King et al., however, were grossly typified by gelatinous appearance and microscopically by large amounts of myxoid material surrounding neoplastic cells and a low mitotic count (0 to 2 per 10 hpf). Each of the 6 patients demonstrated recurrence within 6 months to 10 years despite treatment with total abdominal hysterectomy and bilateral salpingo-oophorectomy or radiation therapy². These tumors behaved in a similar manner to high-grade sarcoma despite the low mitotic counts. Recently, some studies have documented myxoid leiomyosarcoma of the uterus with a high mitotic count as in the case we now report^{4,5}. Myxoid leiomyosarcoma of the uterus can metastasize to the liver, lungs or brain.

No previous patients exhibited rupture into the peritoneal cavity such as occurred in the present case. Laboratory findings were non-specific. Kunzel et al. reported that the serum level of CA-125 can be correlated with tumor growth⁴. The present case demonstrated similar results.

The optimal treatment for this type of tumor is still controversial. As recurrences are frequently observed, complete surgical excision is not sufficient by itself. In addition, the abundance of intercellular myxomatous tissue suggests that this type of tumor may not respond to chemotherapy or radiation therapy.

Magnetic resonance (MR) imaging of leiomyosarcoma of the uterus was employed in some of the reported cases, showing irregular contours or hemorrhagic foci of leiomyosarcoma^{7,8}. To our knowledge, MR features of myxoid leiomyosarcoma of the uterus have not yet been reported. In our case, MR imaging demonstrated multiple well-defined masses with heterogeneous low signal intensity on T₁-weighted images and marked high signal intensity with septations on T₂-weighted images. Upon contrast-enhanced T₁-weighted imaging, the masses demonstrated minimal enhancement with a slightly higher signal intensity than that of water and the septations showed enhancement equal to that of the myometrium. The septations were found to correspond to compressed normal myometrium upon microscopic examination. A mass consisting of myometrium that demonstrates hyperintensity on T₂-weighted images usually represents myxoid-degenerated leiomyoma and cellular myoma^{8)~10}. Yamashita et al. reported that cellular myoma exhibits a homogeneous enhancement on contrast-enhanced T₁-weighted images while myxoid-degenerated leiomyoma has an ir-

regular periphery or minimal enhancement⁸⁾. The findings on contrast-enhanced T₁-weighted images in our case were similar to those of myxoid-degenerated leiomyoma. For this reason, the differential diagnosis between myxoid leiomyosarcoma and myxoid-degenerated leiomyoma was not possible with MR imaging alone. A myometrial mass with high signal intensity on T₂-weighted images and minimal enhancement on contrast-enhanced T₁-weighted images suggests both myxoid leiomyosarcoma and myxoid-degenerated leiomyoma. The serum level of CA-125 may be a marker useful for making the differential diagnosis.

REFERENCES

- 1) Persaud V, Arjoon PD : Uterine leiomyoma. *Obstet Gynecol* 1970 ; 35 : 432-426
- 2) King ME, Dickersin GR, Scully RE : Myxoid leiomyosarcoma of the uterus : a report of six cases. *Am J Surg Pathol* 1982 ; 6 : 589-598
- 3) Peacock G, Archer S : Myxoid leiomyosarcoma of the uterus : case report and review of the literature. *Am J Obstet Gynecol* 1989 ; 160 : 1515-1519
- 4) Kunzel KE, Millis NZ, Muderspach LI, d'Ab-laing G 3d. : Myxoid leiomyosarcoma of the uterus. *Gynecol Oncol* 1993 ; 48 : 277-280
- 5) Schneider D, Halperin R, Segal M, Maymon R, Bukovsky I : Myxoid leiomyosarcoma of the uterus with unusual malignant histologic pattern : a case report. *Gynecol Oncol* 1995 ; 59 : 156-158
- 6) Takemori M, Nishimura R, Sugimura K : Magnetic resonance imaging of uterine leiomyosarcoma. *Arch Gynecol Obstet* 1992 ; 251 : 215-218
- 7) Pattani SJ, Kier R, Deal R, Luchansky E : MRI of uterine leiomyosarcoma. *Magn Reson Imaging* 1995 ; 13 : 331-333
- 8) Okizuka H, Sugimura K, Takemori M, Obayashi C, Kitao M, Ishida T : MR detection of degenerating uterine leiomyomas. *JCAT* 1993 ; 17 : 760-766
- 9) Yamashita Y, Torashima M, Takahashi M, Tanaka N, Katabuchi H, Miyazaki K, Ito M, Okamura H : Hyperintense uterine leiomyoma at T₂-weighted MR imaging : differentiation with dynamic enhanced MR imaging and clinical implications. *Radiology* 1993 ; 189 : 721-725
- 10) Togashi K, Leiomyoma. In : Togashi K, ed. *MRI of the Female Pelvis*. Tokyo, Japan : IGAKU-SHOIN, 1993 ; 81-104