Endometrial stromal sarcoma mimicking submucosal myoma protruding to the vagina: MRI findings

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Summary

A 46-year-old woman complained of persistent abnormal vaginal bleeding over ten days. Her intrauterine device had been removed two years before. Soon after, she suffered from menorrhagia and metrorrhagia. An incidental finding of severe anemia was also noted. In this admission, our initial T2-weighted magnetic resonance imaging (MRI) revealed a well-demarcated mass predominantly in the uterine cavity. The mass was depicted by an isointense signal relative to the myometrium on T1-weighted images, high signal intensity on T2-weighted images, and slightly heterogeneous enhancement on post-contrast images. The patient refused surgery. After two years, follow-up MRI showed a pedunculated mass protruding into the upper third of the vagina with a stalk connecting to the posterior wall of the uterine cavity, simulating submucosal myoma. Histological diagnosis was compatible with low-grade endometrial stromal sarcoma.

Key words: Endometrium; Magnetic resonance imaging (MRI); Sarcoma; Tumor; Uterus; Vagina.

Introduction

Endometrial stromal sarcoma is a rare uterine mesenchymal tumor, accounting for 0.2% of all uterine malignancies and 15-23% of primary uterine sarcomas [1]. These sarcomas are less common than endometrial carcinomas. The tumor originates from the endometrium and consists of cells that resemble those of endometrial stroma in the proliferative phase, with a spindle-like appearance.

The most common symptom of endometrial stromal sarcoma is abnormal vaginal bleeding, and the diagnosis is made on tissue obtained at diagnostic dilatation and curettage. The predominant mode of treatment is surgical and usually is total abdominal hysterectomy and bilateral salpingo-oophorectomy.

Few cases of endometrial stromal sarcoma with magnetic resonance imaging (MRI) have been reported in the literature [2, 3]. In this report, we present a case with follow-up MRI of an endometrial stromal sarcoma with unusual findings.

Case Report

A 46-year-old woman complained of persistent abnormal vaginal bleeding over ten days. She had had an intrauterine device implanted 20 years before. She had had menorrhagia and metrorrhagia after she had removed the contraceptive device two years before. Soon after, she visited a local clinic and severe anemia was noted. The patient had also suffered from dizziness for two days. In this admission, hemoglobulin was 3.4 g/dl. Owing to hypermenorrhea, ultrasound of the pelvis was performed and revealed a mass in the uterine endometrial cavity (Figures 1A-1B). The patient then received dilatation and curettage due to suspicion of endometrial tumor. Histologic findings revealed stromal cell proliferation and low-grade endometrial stromal sarcoma.

MRI of the pelvis (1.5-T, Horizon LX, General Electric, Milwaukee, WI) was performed with use of a Torso coil. A 3.7 × 4.6 × 3.4 cm submucosal mass filled up the endometrial cavity. The tumor showed low signal intensity on T1-weighted images (TR = 516 ms, TE = 11.3 ms) and high signal intensity on T2-weighted images (TR = 4000 ms, TE = 123 ms), with heterogeneous enhancement on postgadolinium images (relatively lower in the enhancement signal than that of the uterine myometrium) (Figure 1C-1E).

The patient refused surgery and received only blood transfusion treatment. Owing to the persistence of anemia and hypermenorrhea, repeated blood transfusions over the next two years were performed on the patient.

The patient suffered from persistent vaginal bleeding for ten days before the next admission. Follow-up MRI showed a 4.9 × 7.6 × 3.8 cm pedunculated mass in the uterine cavity, with a 3.5 cm stalk connected to the posterior wall near the left cornue area and protruding to the upper third of the vagina, mimicking submucosal myoma (Figure 2). No significant regional lymph nodes or extrauterine adenexal invasion were noted. Hysteroscopy examination confirmed the protruding mass at the upper third of the vagina, and then hysteroscopic myomectomy, and dilatation and curettage were performed. Frozen section of the histologic examination proved the diagnosis of low-grade endometrial stromal sarcoma, and then extended transabdominal hysterectomy, bilateral salpingo-oophorectomy and bilateral pelvic lymph node dissection was performed. The final histologic findings from the surgical specimen confirmed the diagnosis of low-grade endometrial stromal sarcoma (Figure 3). The mitoses were up to 2 per 10 HPF. Only minimal myometrial invasion in the connecting stalk area was seen. No evidence of lymphatic permeation was noted. No cancerous involvement was seen in any of 37 dissected lymph nodes.

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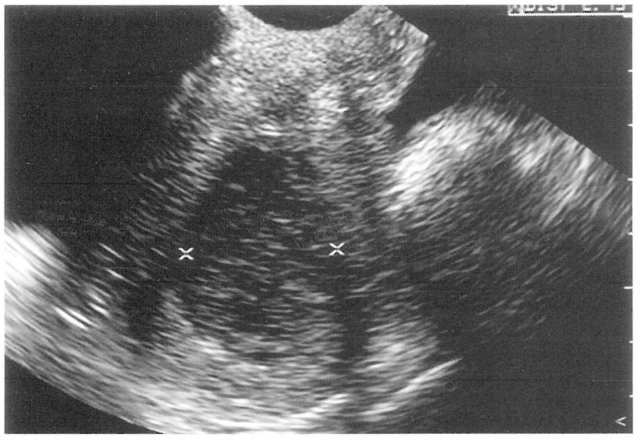
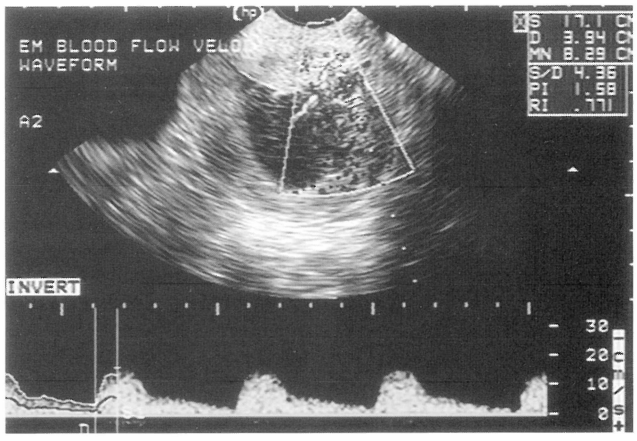
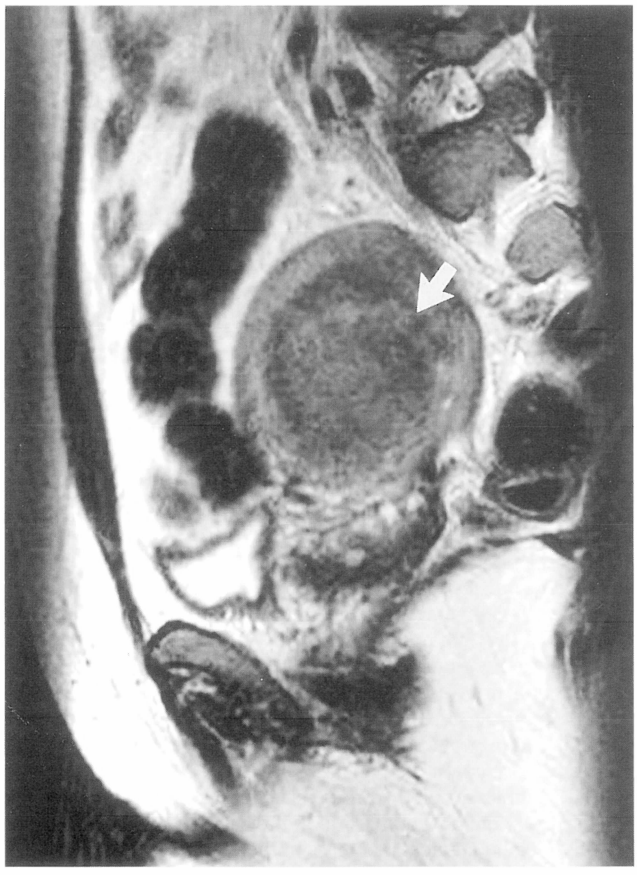
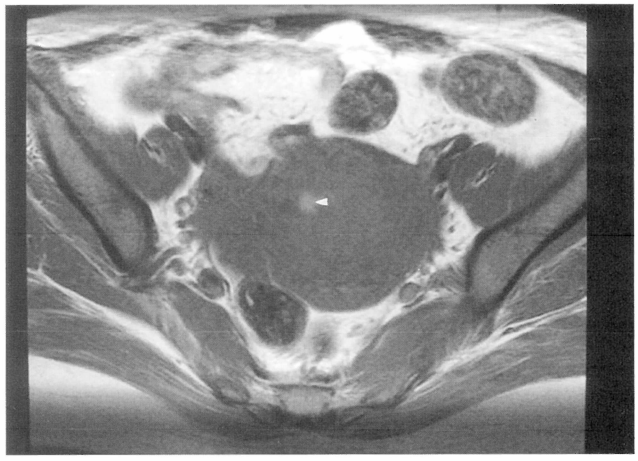








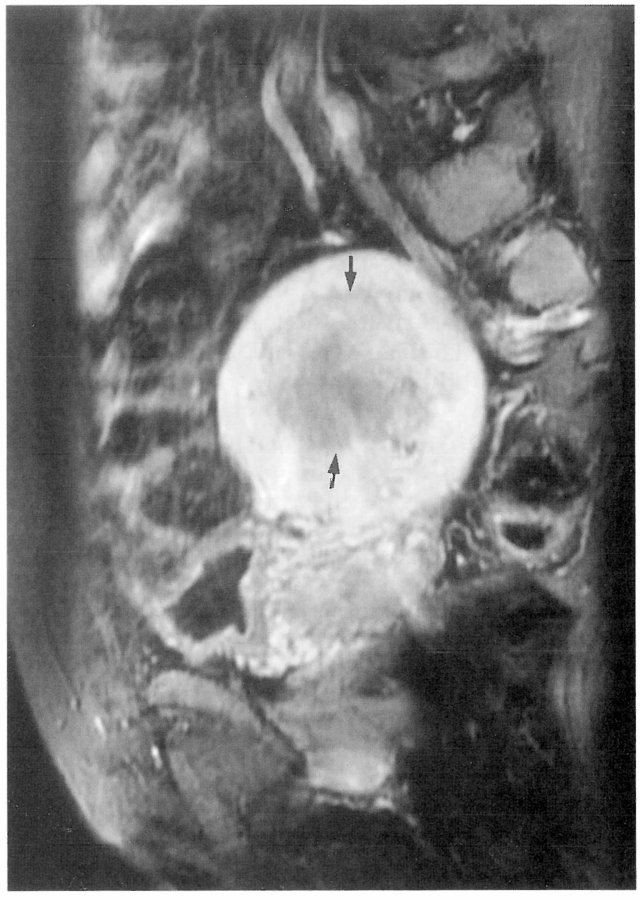
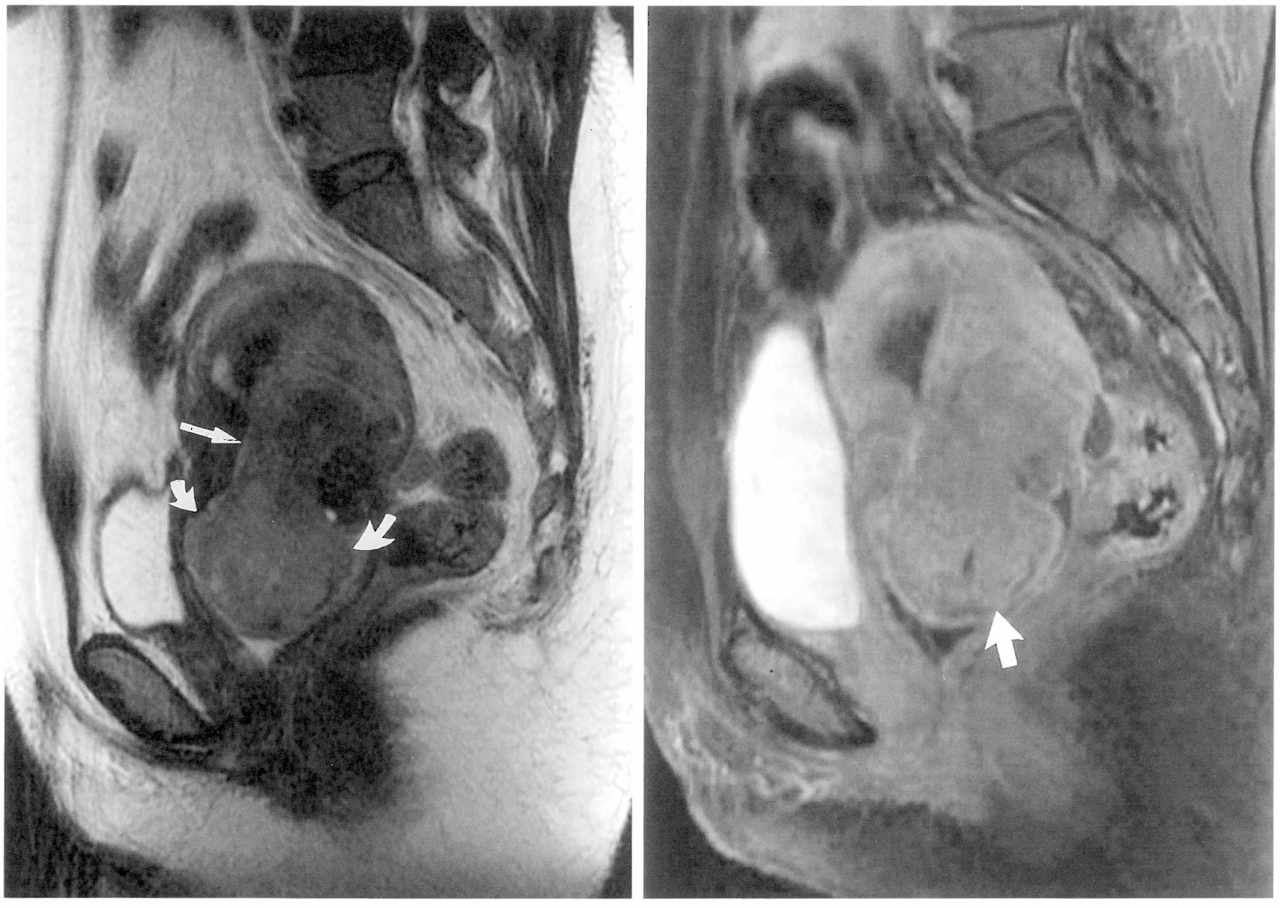


Figure 1. — Initial ultrasound and MRI of the pelvis. **A.** Transabdominal ultrasound shows an isoechoic solid mass filling the uterine cavity. **B.** Color Doppler ultrasound shows high flow resistance within intralesional arteries (P.I. = 1.58, R.I. = 0.771). **C.** Fast spin-echo sagittal T2-weighted MRI (TR 4200/TE 123) shows a slightly hyperintense signal mass filling the uterine cavity, with a focal breakthrough into the myometrium (arrow). **D.** Axial T1-weighted MRI shows that the mass has similar signal intensity to the myometrium, and the focal high-signal-intensity area on T1-weighted image (arrowhead) also remains intense on the T2-weighted image, correlating with focal hemorrhage. **E.** Sagittal gadolinium-enhanced T1-weighted MRI with fat suppression shows heterogeneous enhancement of the tumor (arrows), but the enhancement is less than that of the myometrium.



2a Fig. 2b

Figure 2. — Follow-up MRI of the pelvis two years later. **A.** Sagittal T2-weighted image (TR 4000/TE 122) of the uterus reveals only a mild increase in size of the main mass, but the main mass (curved arrow) "drops" to the upper third of the vagina and with a stalk (arrow) connects to the posterior wall of the uterus near the cornue, mimicking a submucosal myoma. **B.** Sagittal gadolinium-enhanced MRI with fat suppression shows a focal discontinuous surface (arrow) due to ulceration. This MRI finding was later proved by hysteroscopy.

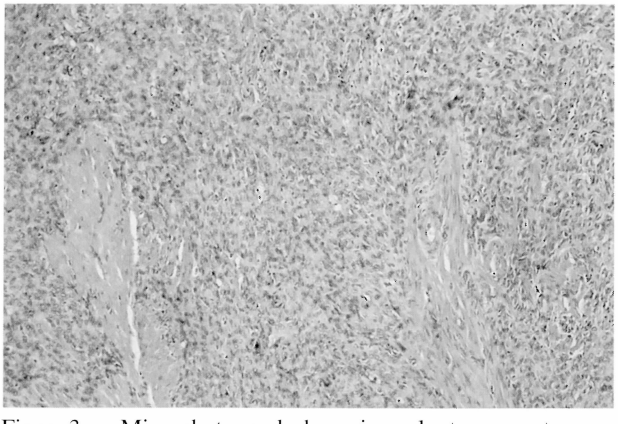


Figure 3. — Microphotograph shows irregular tumor nests composed of oval-nucleated tumor cell infiltrate within the regional myometrium (H & E × 200).

Discussion

Endometrial stromal sarcoma is a rare lesion and the origin is from endometrial stromal cells. The mean age of these patients is between 42 and 58 years, and about 10% to 25% of patients are premenopausal. No risk factors for endometrial carcinoma have been proposed. The most common symptom is unusual vaginal bleeding. No specific clinical symptoms or laboratory tests suggest the diagnosis of endometrial stromal sarcoma.

MRI is the best imaging modality to evaluate uterine neoplasms because of its excellent soft tissue contrast to extrauterine invasion. Several papers have attempted to describe MRI appearance of uterine sarcomas and their mimickers [4, 5] and concluded some overlapping between endometrial stromal sarcoma, leiomyoma and leiomyosarcoma. The typical gross patterns of endometrial stromal sarcoma are variable.

Three main pictures have been reported in the literature. The first pattern is a sharply demarcated polypoid tumor predominantly within the endometrial cavity [2, 6], similar findings were noted in our case. The MRI signal intensity of this type tumor on T1-weighted images, T2-weighted images and post-enhancement T1-weighted images could be homogeneous or heterogeneous. The second pattern is extensive myometrial involvement with either irregular margins, multiple nodular mass formations [7] or bands or low SI within the areas of involved myometrium on T2-weighted images [2, 8]. The two

findings of the second pattern are considered the characteristic features for endometrial stromal sarcoma. The third pattern is multiple cystic mass in the myometrium as presented by Ueda *et al.* [9], which is difficult to distinguish from leiomyoma with cystic changes and leiomyosarcoma.

The MRI signal intensity of endometrial stromal sarcoma is commonly low on T1-weighted images, similar to that of myometrium, and the tumor on T2-weighted images has higher intensity than that of the myometrium. Necrosis and hemorrhage may be observed. Takashi *et al.* [2] reported that the most specific finding of such a tumor was the low-signal-intensity bands in the myometrium on T2-weighted images, although this finding was not seen in every case. As in our case, the initial MRI showed a well-defined mass within the uterine cavity with an obscured endometrial-myometrial border. The signal intensity of the mass was low on T1-weighted images and high on T2-weighted images.

Follow-up MRI images after two years revealed the mass had slightly increased in size and the most salient feature was the main mass protruding to the upper third of the vagina with a stalk connected to the uterus at the posterior wall near the left corner, simulating a submucosal leiomyoma. There were no obvious low-signal-intensity bands in the myometrium on T2-weighted imaging study.

Although leiomyomas typically exhibit low signal intensity on T2-weighted images, these tumors can show variable signal intensities depending on the extent and nature of degeneration. Another point for the differential diagnosis is that leiomyomas almost always are sharply demarcated in contrast to endometrial stromal sarcoma. In our case, most of the tumor was sharply demarcated, mimicking uterine leiomyoma, and the connection site of the stalk with an equivocal infiltrative margin was the only clue to the correct diagnosis.

In summary, MRI is the imaging modality of choice in detection and diagnosis of uterine neoplasms. Radiologists should be familiar with the variations in MRI appearance of endometrial stromal sarcoma, and careful evaluation of the endometrial-myometrial border, the tumor margin and signal intensity change can help to address a correct diagnosis. However, attention should be given to the border of the tumor, which still can mimic a submucosal myoma on MRI. Biopsy should be used to obtain a correct diagnosis in those equivocal cases.

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