CASE REPORT

Spontaneous rupture of uterine smooth muscle tumour presenting acute abdominal pain and haemoperitoneum

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SUMMARY

Uterine smooth muscle tumours are histologically categorised into benign leiomyoma, malignant leiomyosarcoma or smooth muscle tumours of uncertain malignant potentials (STUMPs).¹ Common symptoms of uterine tumours are hypermenorrhea, dysmenorrhea, lumbago or irregular genital bleeding. We experienced a case of uterine tumour with atypical clinical behaviour. A 40-year-old woman who had been diagnosed with leiomyoma presented with severe abdominal pain and intraperitoneal haemorrhage. By emergent surgery, we found that the uterine tumour had ruptured spontaneously. The pathological diagnosis was STUMPs. 14 months later, she underwent a second surgery for a tumour recurrence. Pathological diagnosis was leiomyosarcoma. 20 months later, she underwent a third surgery for a re-recurrent tumour. After the third surgery, massive fluid containing haemorrhage accumulated inside the tumour. Percutaneous drainage of intratumour fluid was successfully performed. Chemotherapy was also taken, but it ended without significant efficacy. 3 years after the first surgery, she died because of intestinal perforation and peritonitis.

BACKGROUND

Leiomyosarcoma is a rare soft tissue neoplasm. The collapse of vessels on the surface of uterine tumour occasionally happens. Besides, spontaneous rupture of leiomyoma is rare with only about 10 cases reported in the last decade.² Spontaneous rupture of leiomyosarcoma is even more unusual. Our case is characteristic for the following points of view: first, the tumour seemed to be leiomyoma by MRI at first, then it was pathologically diagnosed as smooth muscle tumours of uncertain malignant potentials (STUMPs) after the first surgery, and the pathological diagnosis turned into leiomyosarcoma after the second surgery; second, the tumour caused recurrent haemorrhage (peritoneal haemorrhage at the first time and intratumour haemorrhage at the second and third time), and she needed emergent surgery three times; third, the tumour pressure which caused severe abdominal pain was effectively reduced by percutaneous drainage of intratumour fluid. This case may add to the limited experience of clinical behaviour of leiomyosarcoma and its management.

CASE PRESENTATION

A 40-year-old Japanese woman noticed abdominal tumour and visited a gynaecological clinic. A big leiomyoma of 9 cm was found, but she did not accept any therapy or surgery because it was not problematic for her. Four months later, she got acute abdominal pain and visited her clinic. Intraperitoneal bleeding was seen on ultrasonography, and she was referred to our hospital for further evaluation.

INVESTIGATIONS

MRI identified peritoneal haemorrhage and pelvic mass of suspected left ovary origin. The haemoglobin decreased to 8.1 g/dL. Oncological serum markers were increased: CA125 was 204.8 and NSE was 32.4. CEA, LDH AFP and CA19-9 were within normal range.

TREATMENT

During surgery, a huge adherent tumour to the uterus and left ovary was found. It was already ruptured and was bleeding into the intraperitoneal space. Abdominal total hysterectomy and bilateral salpingo-oophorectomy were performed.

On gross examination, a whitish mass of size 18×22×10 cm was surrounding the uterus. Pathologically, it was characterised by spindle cell proliferation. Mitotic activity was 5–6 per 10 high-power fields (HPFs) with mild nuclear atypia. There was no coagulative tumour necrosis (CTCN). The pathological diagnosis was STUMPs. Additional immunohistochemistry was performed. P16 and p53 were negative. Ki-67 was 10%. Oestrogen receptor and progesterone receptor (were positive staining. These results were consistent with STUMPs.^{3 4}

OUTCOME AND FOLLOW-UP

STUMPs is of benign nature; however, rare cases of metastasis is noted.³ No adjuvant therapy, neither chemotherapy nor radiotherapy, is proven to be effective for STUMPs.⁵ Clinical evaluation by ultrasonography and CT was performed every 3 months after the surgery.

Eight months later, a small mass and a haemorrhagic lesion appeared in the vaginal stump, which was to be recurrence (intratumour haemorrhage). At first, the appearance of the recurrent tumour was indistinguishable from haemorrhage. The radiologist also diagnosed the CT image as haemorrhage.

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Figure 1 Massive fluid accumulation in the recurrent tumour.

We carefully followed, and it took 6 months to diagnose it as a recurrence.

Fourteen months later, the second surgery took place. A tumour of 9 cm with adhesion to appendix and omentum was removed. In pathology, mitotic activity was 22 per 10 HPFs, with high-grade atypia and CTCN. The pathological diagnosis was leiomyosarcoma. Ki-67 labelling index was elevated at 40%. The initial histological diagnosis was to be taken as inappropriate, mixed tumour or it is also possible that the tumour evolved to malignancy. In this case, the tumour seems to get malignant evolution because the histological grade and the grade of immunohistochemistry were both apparently evolved in second surgery. After the second surgery, we proposed docetaxel and gemcitabine (DG) chemotherapy to the patient,⁶ which was under clinical trial, but the patient refused it.

Twenty months after the second surgery, a huge tumour recurred, and she suffered from pain caused by the aggressive pressure of the tumour. Suboptimal surgery was performed. Complete cytoreduction was impossible because the tumour was deeply invading the abdominal wall and pelvic cavity.

After the third surgery, she received two cycles of DG chemotherapy (docetaxel 75 mg/kg, gemcitabine 900 mg/m²)⁶ but the tumour was resistant to the chemotherapy and it enlarged rapidly. The patient refused to continue the chemotherapy because of the severe side effects of nausea, fatigue and headache. On abdominal CT, this rapidly growing tumour seemed to be composed of tumour substance and massive fluid accumulation inside (figure 1). The patient suffered from severe abdominal fullness and pain caused by the tension of the tumour. We performed percutaneous catheterisation and drainage of intratumour fluid. The fluid was serous, brownish-red and seemed to contain haemorrhage. The haemoglobin decreased from 9.0 g/dL to 7.4 g/dL. Three litres of intratumour fluid were drained which relieved her symptoms temporarily but not for long. The fluid accumulated again in the tumour, and her symptoms reappeared. She needed drainage every 1 or 2 weeks. Also, she received intraperitoneal administration of paclitaxel 90 mg/body.^{7,8} This intraperitoneal chemotherapy did not cause severe side effects, but it did not show significant efficacy either.

Six months after the third surgery, that is 3 years and 4 months after the first surgery, she died because of intestinal perforation and peritonitis.

DISCUSSION

STUMPs are uterine smooth muscle tumours which cannot be diagnosed as unequivocally benign or malignant. The prognosis of STUMPs is generally good, and no fatal case is reported ever but sometimes, even histologically, the differentiation of STUMPs and leiomyosarcomas is difficult, and it happens to be underdiagnosed or overdiagnosed. Malignant evolution of STUMPs is also reported. In the case of STUMPs, we strongly suggest to follow-up tightly by ultrasonography and CT.

Haemorrhagic degeneration is one of the suspicious findings of leiomyosarcoma, but as far as we searched, there is no literature which report that massive fluid and haemorrhage emerge inside the tumour and that the tumour size can be reduced by drainage. In similar cases, drainage of intratumour fluid is possibly effective to relieve the patient's suffering caused by the tumour pressure.

Intraperitoneal administration of paclitaxel was not effective in our case but its efficacy is not completely deniable. Further exploration of a potential role for intraperitoneal paclitaxel in the management of leiomyosarcoma is required. Several chemotherapy protocols for leiomyosarcoma, like DG or cyclophosphamide, vincristine, doxorubicin and dacarbazine, are now under clinical trials, and we are longing for the verification of effectiveness. These chemotherapies are sometimes difficult to pursue because of the severe side effects. In these cases, intraperitoneal chemotherapy may be considered as an option.

Patient's perspective

- ▶ My first doctor told me that I had a simple leiomyoma. My mother had also been diagnosed as leiomyoma, and she is safe and sound without any treatment. So we had not paid much attention to my disease. Then suddenly I got abdominal pain and hemorrhage and needed emergent surgery. The diagnosis was STUMPs, which was borderline malignancy, then it turned to malignant leiomyosarcoma. I was confused, but I could gradually accept and face to my disease.
- ▶ My doctor told me that there is not yet standard chemotherapy to leiomyosarcoma. The first intravenous chemotherapy suffered much, and I was not motivated to continue it. Intratumor drainage helps me much. It relieves my suffering and improves the quality of life. Intratumor chemotherapy does not stress me, and I hope that this chemotherapy improves my prognosis.

Learning points

- ▶ Intratumour fluid and haemorrhage sometimes emerge in leiomyosarcoma.
- ▶ If the patient suffers from symptoms caused by the pressure of a huge tumour and if the tumour contains massive fluid inside it, percutaneous drainage is possibly effective to relieve the symptoms.
- ▶ For the patients who can hardly pursue intravenous chemotherapy, intraperitoneal chemotherapy may be considered as an option. Further exploration is required for this chemotherapy.

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REFERENCES

- Hendrickson MR, Tavassoli FA. *Mesenchymal tumors and related lesions. Tumors of the Breast and Female Genital Organs*. Lyon: IARC PRESS, 2003:233–44.
- Danikas D, Theodorou SJ, Kotrotsios J, *et al*. Hemoperitoneum from spontaneous bleeding of a uterine leiomyoma: a case report. *Am Surg* 1999;65:1180–2.
- Bell SW, Kempson RL, Hendrickson MR. Problematic uterine smooth muscle neoplasms. A clinicopathologic study of 213 cases. *Am J Surg Pathol* 1994;18:535–58.
- Amant F, Moerman P, Vergote I. Report of an unusual problematic uterine smooth muscle neoplasm, emphasizing the prognostic importance of coagulative tumor cell necrosis. *Int J Gynecol Cancer* 2005;15:1210–2.
- Guntupalli SR, Ramirez PT, Anderson ML, *et al*. Uterine smooth muscle tumor of uncertain malignant potential: a retrospective analysis. *Gynecol Oncol* 2009;113:324–6.
- Hensley ML, Ishill N, Soslow R, *et al*. Adjuvant gemcitabine plus docetaxel for completely resected stages I–IV high grade uterine leiomyosarcoma: results of a prospective study. *Gynecol Oncol* 2009;112:563–7.
- Markman M, Francis P, Rowinsky E, *et al*. Intraperitoneal taxol (paclitaxel) in the management of ovarian cancer. *Ann Oncol* 1994;5(suppl 6):S55–8.
- Nagao S, Iwasa N, Kurosaki A, *et al*. Intravenous/intraperitoneal paclitaxel and intraperitoneal carboplatin in patients with epithelial ovarian, fallopian tube, or peritoneal carcinoma: a feasibility study. *Int J Gynecol Cancer* 2012;22:70–5.

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