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An Unusual Missed Case of Endometrial Stromal Sarcoma

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Abstract Endometrial stromal sarcomas are rare uterine malignant tumours of mesodermal origin. The diagnosis is usually made post-operatively. The usual presentation is abnormal vaginal bleeding, abdominal lump and mild lower abdominal pain. In this case report, we present a case of low-grade endometrial stromal sarcoma where patient was initially thought to have fibroid uterus with cystic degenerative changes and underwent vaginal myomectomy on two occasions within a span of 4 months. Histopathology on the second occasion showed low-grade endometrial stromal cell sarcoma. Hence she underwent extrafascial total abdominal hysterectomy and bilateral salpingo-oophorectomy followed by continuous high-dose progestogen treatment.

Keywords Endometrial · Stromal sarcoma

Introduction

Sarcomas of the uterus are uncommon, and may arise from connective tissue, smooth muscle or the endometrial stroma. Uterine sarcoma is a rare form of malignancy, occurring in 2–5 % of all patients with uterine malignancy, with an incidence of approximately 1-2 cases per 100,000 women in the general population. Endometrial stromal sarcomas are very rare malignant tumours that make up approximately 10 % of all uterine sarcomas but only around 0.2 % of all uterine malignancies [1]. Pre-operative diagnosis of endometrial stromal sarcoma is usually fibroid uterus. The diagnosis is usually made post-operatively. The usual presentation is abnormal vaginal bleeding, abdominal lump and lower abdominal pain. Endometrial stromal sarcoma is characterised by proliferations composed of cells with endometrial stromal cell differentiation. Low-grade endometrial stromal sarcoma has an infiltrating margin and typically show extensive worm-like vessel invasion.

Case Report

A 36-year-old para 2 was admitted at a local private hospital with history of menorrhagia and dysmenorrhoea for the last 6 months. Her menstrual cycles were at intervals of 25–26 days, and bleeding lasted for 10 to 12 days. An ultrasound scan showed an enlarged uterus of size 10 × 6 × 6 cm, endometrial thickness of 9 mm and well defined cystic lesion in posterior myometrium of 4.6 × 3.8 cm, showing echogenic solid component, indenting the endometrium suggestive of fibroid polyp. As no hysteroscopic surgery facility was available at this hospital, patient underwent vaginal myomectomy. The histopathology report was of necrosed tissue material with no definite pattern. However, she continued to have on and off bleeding episodes over the next 2 months and was started on progesterone therapy with norethisterone 5 mg thrice a day to which she was unresponsive. An ultrasound scan showed an enlarged uterus of size 9.6 × 4.6 × 6.1 cm with

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echogenic lesion of 4.6 × 4.5 cm with its stalk in endometrium suggestive of fibroid polyp. Hence she changed to another private practitioner, and again underwent vaginal myomectomy, again no hysteroscopy facility was available to her. The histopathology showed ulcerated and inflamed round cell tumour infiltrating the myometrium. The tumour was composed of round cells with scanty cytoplasm with fine chromatin. Tumour cells showed 1-2 mitoses per high power field, suggestive of low-grade endometrial stromal sarcoma. She was referred to our tertiary care institute for further management. Her general and systemic examinations were normal. Liver and lungs were normal. Abdominal and pelvic examinations revealed a uniformly enlarged uterus of 10 weeks size, soft in consistency. Ultrasound scan and thorax as well as abdominal and pelvic CT scans were unremarkable except for uniformly enlarged uterus. She underwent extrafascial total abdominal hysterectomy and bilateral salpingo-oophorectomy. The operative findings were uniformly enlarged uterus with normal-looking tubes and ovaries. There were no metastatic deposits elsewhere. The lymph nodes were not enlarged. Cut section of uterus showed thickened walls and no gross pathology was noted on cervix, fallopian tubes and ovaries. Microscopically, no malignant cells could be demonstrated in the peritoneal washings. Examination of uterus revealed low-grade endometrial stromal sarcoma, infiltrating the entire thickness of myometrium. The cervix and ovaries were normal. Post-operative period was uneventful. Immunohistochemistry was not possible as our hospital does not have facilities for the same, and the patient could not afford to have it done in an advanced laboratory. At follow-up, patient was started on high-dose progesterone therapy, medroxyprogesterone acetate 100 mg per day. Patient came for regular follow-up, and is presently fine with no recurrences.

Discussion

Endometrial stromal sarcomas are very rare malignant tumours, constituting around 0.2 % of all uterine malignancies [1]. Women with low-grade Endometrial Stromal Sarcoma (ESS) are younger than women with other uterine sarcomas, with a median age between 45 and 57 years and, generally do not have the usual risk factors for endometrial cancer [2]. Symptoms at presentation include abnormal vaginal bleeding, progressive menorrhagia and abdominal pain. While often indolent in behaviour, Endometrial Stromal Sarcomas are malignant, and up to 30 % of women with low-grade ESS have extra uterine disease at presentation.

Endometrial stromal sarcoma is characterised by proliferations composed of cells with endometrial stromal cell differentiation. Low-grade endometrial stromal sarcoma has an infiltrating margin and typically show extensive worm-like vessel invasion.

They are of three types depending upon mitotic activity, vascular invasion and observed differences in prognosis into:

1. Endometrial stromal nodule
2. Low-grade endometrial sarcoma and
3. High-grade or undifferentiated endometrial stromal sarcoma [3]

Patients most commonly undergo surgery with presumptive diagnosis of uterine fibroid or pelvic mass. High index of suspicion is required to make pre-operative diagnosis of endometrial stromal sarcoma particularly in fibroids with any abnormal presentation such as rapid enlargement or abnormal ultrasound findings of heterogeneous mass or fibroid with degenerative changes. Also, the physician should have suspicion when the histopathological diagnosis of endometrial sampling yields hyperplastic stroma with few glands [4].

Due to the high recurrence risk even with localised tumours, many clinicians advocate use of adjuvant chemotherapy, radiation therapy, and/or hormone therapy with medroxyprogesterone, tamoxifen, gonadotropin releasing hormone (GnRH) analogues and aromatase inhibitors [5–7]. Our patient received post-operative continuous hormonal therapy with high-dose medroxyprogesterone acetate.

The surgical stage is most significant prognostic regarding recurrence and survival in low-grade endometrial stromal sarcoma. They tend to grow slowly and commonly recur many years after initial diagnosis [8].

Post-operative pelvic radiotherapy reduces local recurrence but has not been consistently shown to prolong the survival.

These tumours typically have an indolent growth with a tendency for late recurrence [9]. Pelvic or abdominal recurrences in stage I disease develop in one-third to one-half of patients [10].

Conclusion

Endometrial stromal sarcomas are very rare tumours of mesodermal origin presenting with abnormal uterine bleeding, mostly in perimenopausal women. The usual pre-operative diagnosis is fibroid and the diagnosis is made after histopathological examination. High index of suspicion of sarcoma in uterine tumours with the features not typical of fibroid can make the pre-operative diagnosis of uterine sarcomas.

Compliance with Ethical Standards

Conflict of interest None.

References

1. Ashraf-Ganjoei T, Behtash N, Shariat M, Mosavi A. Low grade endometrial stromal sarcoma of uterine corpus, a clinico-pathological and survey study in 14 cases. World J Surg Oncol. 2006;4:50.
2. Ganjoei TA, Behtash N, Shariat M, Mosavi A. Low grade endometrial stromal sarcoma of uterine corpus, a clinicopathological and survey study in 14 cases. World J Surg Oncl. 2006;4:50.
3. Berek JS, editor. Novaks gynaecology 14th edn. Philadelphia: Lippincott Williams and Wilkins; 2006. p. 1382–4.
4. Mert G, Aydin C, Cenk MG, Ali B, Seyran Y. A case of malignant low grade endometrial stromal sarcoma and review of the literature. J Turkish Germangynecol Assoc. 2004;5(1):73–5.
5. Husseiny GE, Bareedy NA, Mourad WA, Mohamed G, Shoukri M, Subhi J. Prognostic factors and treatment modalities in uterine sarcoma. Am J Clin Oncol. 2002;25:256–60. doi:10.1097/00000421-200206000-00010.
6. NCCN Clinical practice guidelines in oncology. Uterine neoplasms. 2009; 2.
7. Linder T, Pink D, Kretzschmar A, Mrozek A, Patience PC, Reichardt P. Hormone treatment of endometrial stromal sarcomas: a possible indication for aromatase inhibitors. J Clin Oncol. 2005;23(16S):9057.
8. Mukhopadhyay P, Sharma PP, Muraleedharan PD, Sarkar S. A case of endometrial stromal sarcoma. J Obstet Gynecol India. 2008;58(1):73–4.
9. Shafi M, Luesley DM, Jordan JA. Gynecological oncology. New York: Churchil Livingstone; 2001. p. 120–1.
10. Spano JP, Soria JC, Kambouchner M, Piperno-Neuman S, Morin F, Morere JF, Martin A, Breau JL. Long term survival of patients given hormonal therapy for metastatic endometrial stromal sarcoma. Med Oncol. 2003;20:87–93. doi:10.1385/MO:20:1:87.