

A case of endometrial stromal sarcoma presenting at very young age

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Abstract

Uterine sarcomas are heterogeneous group of malignancies both clinically as well as pathologically. Leiomyosarcoma contributes for majority of uterine sarcomas (30-40%) with the peak incidence in postmenopausal women (50-60 years). Endometrial Stromal Sarcoma accounts for 10% of uterine malignancies, generally an indolent disease with long natural history. It presents in relatively young age. ESS tumors have high incidence of oestrogen and progesterone receptor expression, and are hormonally responsive.

We present a case of a endometrial stromal sarcoma in a 28 years old patient who presented with heavy menstrual bleeding and secondary subfertility. Ultrasound scan revealed a typical appearance of a leiomyoma. However, later diagnosed a ESS and had undergone surgical management by gynaecological oncosurgical team.

Preoperative diagnosis of uterine sarcoma in a young patient is always a challenge. Surgery is the mainstay of treatment. However, due to high expression of hormone receptors peri-operative hormonal suppression plays a vital role in prevention of recurrence and treatment of advanced cancers.

Key words: sarcoma, endometrial stromal sarcoma (MRV)

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Introduction

Endometrial stromal sarcoma (ESS) is a rare type of uterine sarcoma which contributes for 0.2% of uterine malignancies¹. The annual incidence is 1-2 cases per million women. In contrast to other uterine sarcomas, it presents at younger age and the peak incidence is between 42-58 years. Even though the pathogenesis of ESS is unknown, it is evident that unopposed

estrogens, tamoxifen exposure and polycystic ovarian syndrome are associated with this condition. About 55.6% of ESS showed chromosomal on deletion 7 p and might play a role in tumor development². ESS is less virulent compared to other mesenchyme tumors of the uterus and recurrences can occur even after 20 years. We present a rare case of endometrial stromal sarcoma in a women's 3rd decade of life.

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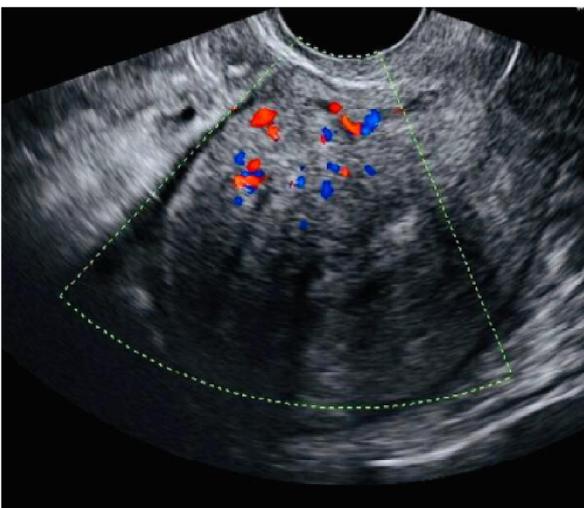
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Case

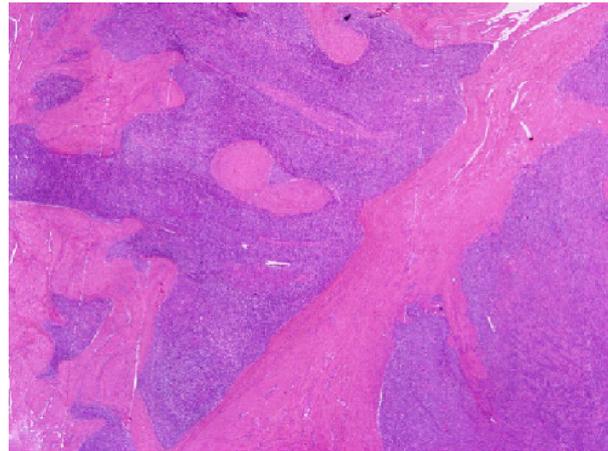
A 28 years old mother of 1 child previously healthy lady presented to us with secondary sub fertility for 3 years duration. She also complains of 3 months history of heavy menstrual bleeding. Her cycles are regular (28 days) but duration bleeding had increased from 3 to 7 days, bleeding significantly affected her quality of life. She was treated with mefenamic and tranexamic acid by a General Practitioner and did not respond to treatment.

On examination she was pale, abdominal and pelvic examination revealed a 12 weeks size firm tender uterus. Transvaginal examination showed anterior wall well circumscribed 5 cm size hypoechogenic lesion with increased vascularity. The endometrial cavity was distorted but morphology of endometrium was normal. A radiological diagnosis of sub mucosal fibroid was made and proceeded with myomectomy. However, during surgery highly vascular uterus was found and myometrium was very fragile without a tissue plane, and wedge biopsy was taken for histology from anterior wall of uterus. Bilateral ovaries, pelvic side wall and omentum were normal.

Histology revealed and a low grade endometrial stromal sarcoma. Patient was referred to national cancer unit. After multidisciplinary team discussion she had undergone total abdominal hysterectomy, bilateral salphingo oopho-rectomy and pelvic node dissection and planned for post-operative chemo therapy.



- Pelvic ultrasound scan with doppler studies are inconclusive, mostly diagnosed as adenomyosis or leiomyoma. Endometrial biopsy may be helpful if tumor involved the endometrium.



- Histologically, ESS shows well-differentiated endometrial stromal cells with mild nuclear atypia and typically invade the lymphovascular spaces of the myometrium. Necrosis of tumour cell is rare.

Discussion

The classic presentation of ESS is with abnormal uterine bleeding and can present with pelvic pain or dysmenorrhea. Preoperative diagnosis of endometrial stromal sarcoma is still being a challenge to gynaecologist. Pelvic ultrasound scan with doppler studies are inconclusive, mostly diagnosed as adenomyosis or leiomyoma. Endometrial biopsy may be helpful if tumor involved the endometrium.

MRI is helpful to differentiate between the uterine masses. MRI features include bands of low-signal intensive areas within the myometrium or extension of lesion into adjacent structures such as fallopian tubes, ovaries or along the blood vessels³. However, the definitive diagnosis of ESS can only be made by histological assessment.

According to WHO 2003 classification endometrial stromal tumor is classified in to main 3 types based on nuclear pleomorphism and necrosis. The subtypes are a) endometrial stromal nodule, (b) low-grade endometrial stromal sarcoma, (c) undifferentiated endometrial or uterine sarcoma.

Surgery is the mainstay of treatment as for any other sarcoma, which includes total abdominal hysterectomy and bilateral salphingo oophorectomy⁴. Role of lymphadenectomy is controversial. As immunohistochemical studies reveal rich expression of hormone receptors (oestrogen and progesterone) post-operative hormone replacement therapy is contraindicated.

However, for stage 1 disease (tumour limited to uterus with <5cm) retention of ovarian function can be considered in a very young patient.

Post-operative hormonal suppression with GnRH analogues and aromatase inhibitors plays an important role. Effective duration of therapy is still undetermined. Radiotherapy in the form of brachytherapy can be considered in advance stage or recurrent tumor, but not as routine therapy in early stage disease⁵.

Prognosis of the disease is mainly depends on the stage of the disease, presence of hormone receptors and general well-being of the patient. Presence of lymph node metastasis is an indicator of poor outcome. Five-year survival between 54-100% in stage 1 and 2 diseases, and for advance disease it is only 11%. Indolent nature of these tumors with late recurrence needs frequent follow up in first 4 years followed by lifelong annual follow up.

Conclusion

ESS is a rare type of uterine malignancy, 10-25% of cases present in premenopausal women¹. Pre-operative diagnosis is always a challenge and MRI is helpful

in suspected cases. Surgery is the main mode of management. Post-operative hormonal suppression is promising mode of management due to rich expression of estrogen and progesterone receptors.

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