Low grade endometrial stromal sarcoma: A case report

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Case Report

Abstract

Low grade endometrial stromal sarcoma is a rare uterine mesenchymal tumor comprising 0.2% of all uterine malignancies. We present here a case of low grade endometrial stromal sarcoma presenting clinically as fibroid uterus.

Keywords: Endometrial Stromal Sarcoma; Fibroid; Menorrhagia

Introduction

Low grade Endometrial stromal sarcomas (ESS) are rare malignant tumors that make up less than 10% of all uterine sarcomas and only around 0.2% of all uterine malignancies. Its clinical recognition may be difficult and most often a preoperative diagnosis of a uterine fibroid is made. Low grade ESS resembles endometrial stromal cells in the proliferative phase. It affects younger women and the mean age is 42 to 58 years. ESS is an indolent tumor with local recurrences and distant metastasis and can occur even many years after initial diagnosis.

Case presentation

A 47-Year-old woman P3 presented with menorrhagia of 6 months duration. Her previous menstrual history was normal. On examination she was severely anemic Hb was 6 gm%, her general physical examination was normal. On per abdominal examination a lump was palpable corresponding to 16 weeks gestation. On per vaginal examination uterus was uniformly enlarged to 16 weeks size. USG showed a isoechoic and hypoechoic mass of 80 mm by 70 mm size in the uterus suggestive of fibroid uterus with cystic degeneration. Bilateral ovaries and fallopian tube were normal, so clinical diagnosis of fibroid uterus with cystic degeneration was made. Since the patient had intractable menorrhagia and the mass was quite big, so a total abdominal hysterectomy with cystectomy was performed. She was transfused 4 units of blood prior to surgery. Intraoperative findings were those of an enlarged uterus of 16 weeks with smooth surface.

Her postoperative period was uneventful and the patient was discharged 5 days after surgery. On

histopathological examination uterus with cervix measured 10 × 7 × 5 cms. On cutting open large multinodular growth measuring 8 × 7 cms involving whole of the myometrium was identified. The growth was extending up to the serosa. C/S through growth was tan/yellow (Figure 1).

Figure 1: Low-grade endometrial stromal sarcoma showing diffuse permeation of the myometrium in the form of small nodules bulging on the cut surface.

Microscopic examination showed highly cellular tumor composed of uniform cells with oblong/spindle shaped nuclei and scant cytoplasm arranged in sheets. The tumor cells showed characteristic infiltration in the form of irregularly shaped tongues/islands placed randomly between bundles of smooth muscle cells. Vascular invasion was also seen at places. Many mitosis (6/10 hpf) were seen An arborising vascular pattern was also seen at places. Final diagnosis of low grade endometrial stromal sarcoma was made ((Figure 2) and (Figure 3)).

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Discussion

Endometrial stromal tumors are among the least common neoplasms of the uterine corpus, with an annual incidence of about 2 per million women. They comprise 0.2% of all female genital tract malignancies. They usually occur at an earlier age (42-58 yrs) in comparison to other uterine malignancies with about 10-25% patients occurring in the premenopausal age group. The usual clinical presentation of ESS is abnormal uterine bleeding, uterine enlargement or pelvic pain.

Most tumors grow through the intramural sections of the uterus rather than intracavitary, hence making it difficult for preoperative histopathology diagnosis. These tumors have an indolent growth with a tendency for late recurrence. Metastasis are rarely detected before the diagnosis of the primary lesion. The latest 2014 WHO classification scheme now incorporates recent molecular findings into the classification, dividing ESTs into endometrial stromal nodule (ESN), low-grade endometrial stromal sarcoma (LGESS), high-grade endometrial stromal sarcoma (HGESS) and undifferentiated uterine sarcoma (UUS).

Diagnosis of low grade endometrial stromal sarcoma is often made post operatively in most instances by histopathological examination since in majority of cases a preoperative clinical diagnosis of fibroid uterus is made. Immunohistochemistry may be needed when there is a problem in distinguishing these tumors from histologic mimics like cellular leioymoma cellular endometrial polyp and adenomyosis. A panel of immunostains that include CD10 and two smooth muscle markers desmin caldesmon should be used, as there is no marker specific for ESS.

The treatment of choice is total abdominal hysterectomy with bilateral salpingo oophorectomy and pelvic and periaortic selective lymphadenectomy. Due to the high recurrence risk even with localized tumors adjuvant treatment either with progestins, radiation therapy or even aromatose inhibitors may be given in order to suppress the tumor growth.

Conclusion

LGESS may not be familiar to the gynaecologists because of the rarity of the tumor. The usual preoperative diagnosis is fibroid and diagnosis is usually made post operatively after histopathological examination. In our case also clinical diagnosis of fibroid uterus was made. Final diagnosis of low grade endometrial stromal sarcoma was made on histopathological examination alone.

Conflict of interest

The authors declare that they have no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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