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CASE REPORT

# Low-Grade Endometrial Stromal Sarcoma in Young Age: A Clinicopathological Report

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## Introduction

Endometrial stromal sarcoma (ESS) is a rare tumor of mesodermal origin. It constitutes 0.2 % of all uterine malignancies and represents ~10 % of all uterine sarcomas. It may be mistaken for leiomyoma, as identifying it clinically is difficult and is more often diagnosed postoperatively after a histopathological examination [1]. Classically, ESS appears as a single nodule, multiple solid-cystic masses, or a poorly demarcated lesion with occasional cystic degeneration grossly [1].

Depending on the cellular uniformity, mitotic activity rate (<3 per ten high-power fields vs. >10 per ten high-power fields) and presence of hemorrhage and necrosis, ESS is classified as low-grade (LGESS) or high-grade ESS (HGESS). In recent times, however the division of ESSs into low-grade and high-grade categories has fallen out of favor, and the term ESSs is now considered best restricted to neoplasms that were formally classified as LGESS [2]. High-grade tumors without recognizable evidence of a definite endometrial stromal phenotype are now simply termed as endometrial sarcomas [2]. This is because, although half the LGESS are limited to the endometrium, the other half shows focal, worm-like, or diffuse, multiple, nodular permeations in the myometrium from the endometrial foci. This article presents a LGESS with the nodular mass partially located in the myometrium.

## Case Report

A 29-year-old woman was admitted to the hospital due to excessive bleeding per vaginum for 3 days. She had a history of irregular vaginal bleeding since the last 2 months with menorrhagia and dysmenorrhoea, gradually increasing in intensity. She had been married for 6 years and having no issue, she had undergone an infertility work-up in the past. On examination, the uterus was enlarged to 10–12 weeks size. All laboratory tests including hormonal tests were unremarkable. Ultrasonography confirmed a bulky uterus with a well-defined complex mass of 5.5 × 4.2 cm along the anterior wall. The endometrium was 4.2 mm thick with a small cystic collection. Both adnexa were normal and no free fluid was seen in the pouch of Douglas.

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Endometrial curettage was performed and the results were normal. No tubercular bacilli were present on culture. On laparoscopy, the anterior wall of uterus was visualized as irregular and it appeared like an intramural myoma of 5 × 4 cm size. On chromopertubation, the right fallopian tube looked convoluted but patent, while left fallopian tube had a cornual block. With the provisional diagnosis of leiomyoma, haemostatic dose of Medroxyprogesterone (tab. meprate, 10 mg bid, Serum Institute) was prescribed to control bleeding, but no relief was achieved. Finally, a long-acting gonadotropin (injection Leuprolide; 3.6 mg subcutaneous) was administered once a month for six consecutive months. During this period, she became amenorrheic. Subsequently, hysteroscopic myomectomy was performed and the tissue was sent for histopathological examination. Meanwhile immunohistochemical study of tissue showed the tumor cells were focally positive for CD10 and were negative for smooth muscle actin (SMA) and desmin, leading to the probable diagnosis of ESS. The histopathological report confirmed LGESS with occasional mitosis. Consequently, the patient underwent on MRI scan to assess the extent of vascular involvement. The upper abdomen was normal, but the lower abdomen showed an enlarged uterus with 5 × 5 cm mass in the anterior wall. Anteriorly, the plane in between endometrium and myometrium was lost. Thus, laparotomy was performed and total abdominal hysterectomy with bilateral salpingo-oophorectomy was done. The cut-section of the specimen exhibited a lobulated mass 6 × 6 cm in the anterior wall of uterus with a central gray to pale yellow soft, necrotic area (Fig. 1). The polypoid mass was invading half of the myometrium. There were no associated fibroids. The endometrium and serosal surface appeared normal. Cervix and both adnexa were normal. On histopathological examination, the specimen yielded the diagnosis of a pure LGESS with superficial myometrial invasion (Fig. 2). The patient is in regular follow-up and is doing well; recently she adopted a child also.

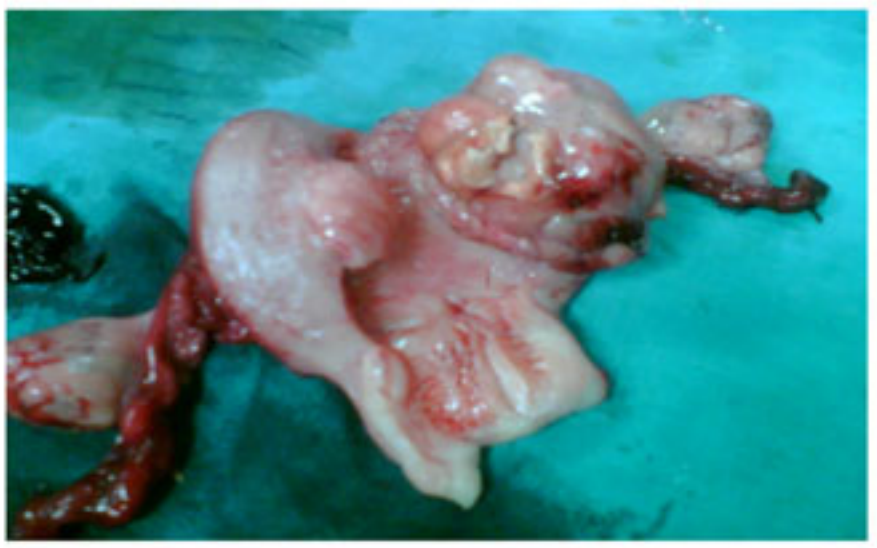

**Fig. 1** Cut section of uterus showing necrotic mass

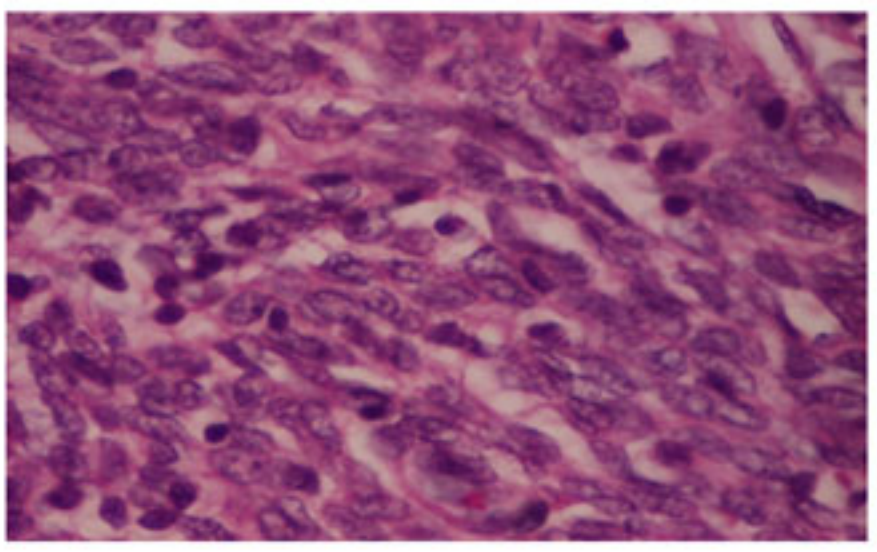

**Fig. 2** Histopathology report shows Low grade ESS. (Courtesy Dr. Kavita Bhardwaj Sr. Consultant Pathologist Saroj Hospital Rohini Delhi)

## Discussion

ESS is a very rare malignant tumor and is characterized by sheets of cells with endometrial stromal cell differentiation. LGESS has an infiltrating margin and commonly exhibits extensive worm-like lymphatic and venous vessel invasion. Although, it is more common in patients between ages 42 and 53, in our case-report, the patient is a 29-year-old woman. Classically, she presented with irregular vaginal bleeding, as this is the most common presenting symptom. Ashraf-Ganjoei et al. [1] also observed vaginal bleeding as the presenting complaint in 86 % of the patients. ESS may also present with pain or pressure symptoms, or it could also be asymptomatic. None of the histologic types of ESS are clearly related to parity. ESS may be confused with leiomyoma, uterine leiomyosarcoma (LMS), or other sarcomas, especially histologically, when associated with myxoid, epithelioid, and fibrous changes. In addition, the immunohistochemical profile of ESS may have similarities with leiomyoma and LMS, with expression of muscle-specific actin (MSA), SMA, and desmin. Diffuse CD10 immunoreactivity has proven to be a useful positive predictive marker for ESS. CD10 is a sensitive and diagnostically useful immunohistochemical marker of normal endometrial stroma and of endometrial stromal neoplasms. Most cellular leiomyomas are completely negative for CD10 [4]. Chu et al. [3] found 100 % positive result for CD10 in their series. LG ESS—have a more protracted clinical course and have <10 MF/10 HPF, Recurrence occurs late. Local recurrence is more (50 % cases), Total abdominal Hysterectomy with bilateral salpingo-oophorectomy is optimum initial therapy, and Radiotherapy is recommended for inadequately excised or locally recurrent pelvic disease.

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