Case Report

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# High-Grade Endometrial Stromal Sarcoma with a Mucoid Grossly Feature: A Case Report of a 69-Year-Old Iranian patient

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**Running Title** Rare Case Report: Endometrial Stromal Sarcoma of an Iranian Patient



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## ABSTRACT

Reporting clinical cases of high-grade endometrial stromal sarcoma tumors is essential for a better understanding of their natural course and for the development of appropriate diagnostic and treatment strategies. To date, only a few cases have been studied in this regard.

In the present study, the authors present the case of a 69-year-old Iranian woman diagnosed with high-grade endometrial stromal sarcoma involving the myometrium and ovaries. The diagnosis was confirmed after pathology, which revealed myometrial and vascular involvement. The patient underwent total abdominal hysterectomy, bilateral salpingo-oophorectomy, and adjuvant chemotherapy with chemotherapy regimens.

## Introduction

Endometrial stromal sarcoma (ESS) is a rare malignancy originating from the endometrial stroma, accounting for 2% of uterine malignancies. According to statistics, the prevalence of this malignancy is one case per 1 million women per year [1-3]. As per the classification by the World Health Organization (WHO) in 2014, ESS tumors are divided into four categories: stromal nodule, low-grade endometrial stromal sarcoma (LESS), high-grade endometrial stromal sarcoma (HESS), and undifferentiated endometrial sarcoma (UES). UES itself is further divided into monomorphic and polymorphic groups [4,5].

Unlike endometrial carcinoma, diagnosing ESS as a mesenchymal tumor is challenging. Many previous investigations using hysteroscopy and uterine endometrial curettage have shown that a definitive diagnosis cannot be reached. The final diagnosis is only possible when the samples obtained from the hysterectomy are examined under the microscope for pathology. Furthermore, no definitive diagnostic method has been reported to diagnose endometrial stromal sarcoma before surgery to date. Even the use of methods such as dilatation, curettage, and

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endometrial biopsy is not a suitable diagnostic method, as the endometrial stromal sarcoma tumor grows towards the myometrium, and only a part of the endometrium is removed during curettage.

To date, very few studies have investigated patients with high-grade endometrial stromal sarcoma tumors. It is crucial to provide more clinical reports to better comprehend its natural course, and to conduct further studies to determine appropriate diagnostic and treatment strategies. Therefore, this article presents a scientific report on a 69-year-old woman from Iran diagnosed with high-grade endometrial stromal sarcoma.

## Case presentation

A 69-year-old woman with a 5-month history of Abnormal Uterine Bleeding (AUB), vaginal bleeding, and a palpable mass in the lower abdomen, with no family history, was referred to the gynecologist. The patient's medical history did not mention smoking and alcohol consumption. The initial examination of the vagina and cervix was normal. Sonographic and Doppler examinations suggested the presence of endometrial polyps and ovarian atrophy. Endometrial aspiration revealed a secretory endometrium with neoplastic cells, and the patient underwent a total abdominal hysterectomy.

During the operation, the uterine opening revealed a necrotic polypoid mass infiltrating irregularly and deeply into the myometrium. The mass appeared pale yellow and soft. The ovaries and right and left iliac cavities were examined macroscopically, and due to the involvement of the ovaries and fallopian tubes, a bilateral salpingo-oophorectomy (BSO) was also performed. Frozen section pathology was used during surgery to investigate lymph node involvement. The samples sent for pathology included four samples as follows:

- A sample of the uterus body and cervix with dimensions of 3 x 8 x 5 cm, along with a fibrotic ovary on the right side with dimensions of 1 x 1.5 x 2.5 cm.
- A sample of the right fallopian tube, 7 cm long and 8 cm wide, along with a 5 cm large para ovarian cyst.
- A sample of the left fallopian tube, 6 cm long and 5 cm wide, along with the fibrotic ovary on the same side, measuring 1 x 2 x 3 cm.
- A section of the endometrial fundus with an endometrial polyp and a yellowish-brown fleshy mass without myometrial involvement, but with foci of necrosis and bleeding measuring 3 x 4 cm.

In the microscopic sections of endometrial polyps, a spindle cell neoplasm was observed, characterized by large and polymorphous round endometrial cells, with no regular vascular pattern. More than 10 mitotic patterns were observed in 10 large microscopic fields (High power field, HPF), along with some abnormal cells (Figures 1 and 2).

IHC was performed and showed positive results for CD10 and BCOR, while it was negative for actin, desmin, and caldesmon. Invasion of lymphatic vessels and infiltration at the edges were observed, along with nuclear atypia without vascular branches. Spindle cells with moderate to severe polymorphism and

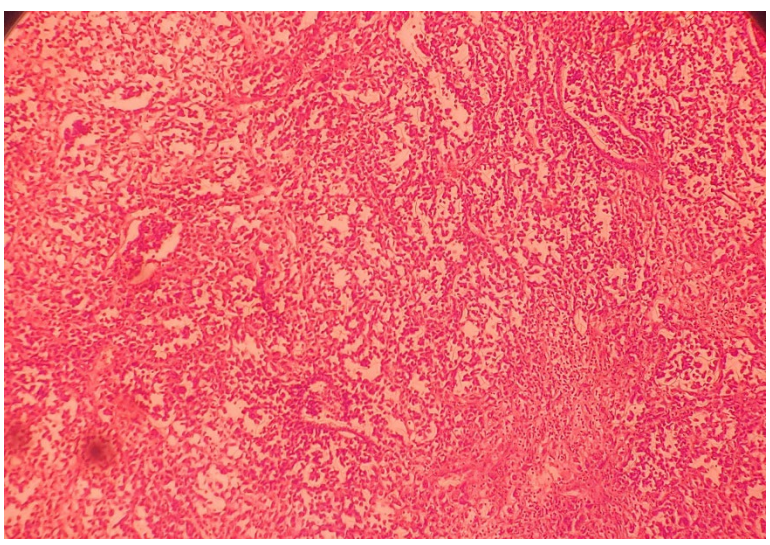

**Fig. 1.** Microscopic findings indicating round endometrial cells were seen as large and polymorphous without a regular vascular pattern (H&E staining; X10).

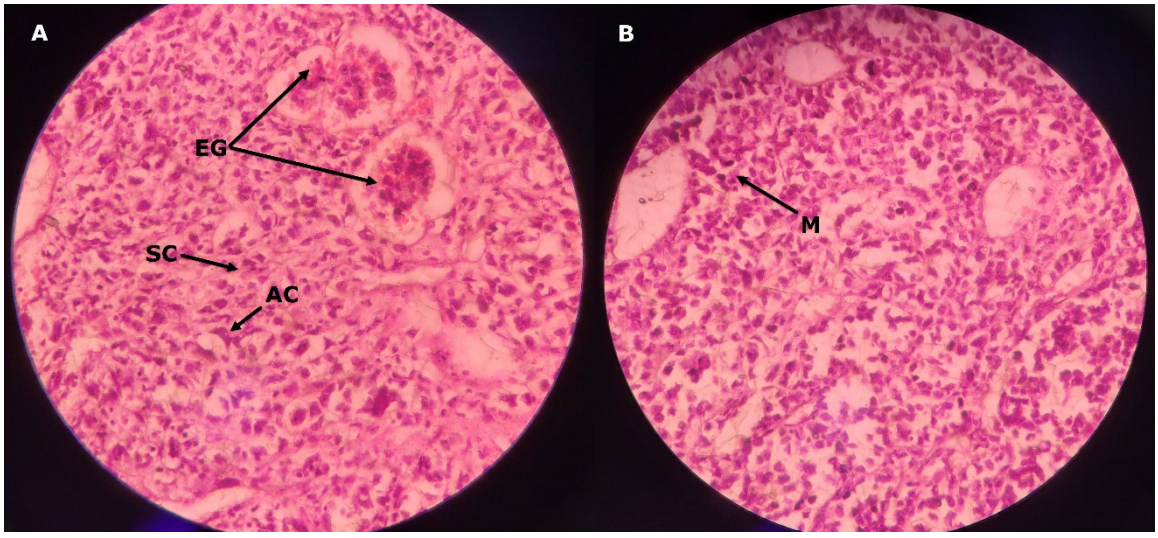


**Fig. 2.** Endometrial glands (EG) with stromal cells (SC) and atypical cells (AC) are seen in A. Mitosis (M) can be seen in B (H&E staining; X40).

multinucleated cells were not seen in the samples. Sections of the pelvic lymph nodes on both sides and the peritoneal nodule revealed multiple metastatic foci. Focal involvement of the cervix was observed, and it was also noted that the ovaries on both sides were involved. According to the FIGO staging system, the disease was classified as stage IIIC due to its myometrial invasion, cervical invasion, pelvic node involvement, and absence of tumor invasion of the bladder, bowel, or distant metastasis.

Based on the above microscopic findings and the patient's history, the diagnosis of high-grade endometrial stromal sarcoma with myometrium and vascular involvement was confirmed. A CT scan of the abdomen and chest was performed, and no metastasis was detected in other organs.

Following total abdominal hysterectomy and bilateral salpingo-oophorectomy, the patient underwent adjuvant chemotherapy including Adriamycin 250 mg/m$^2$ and Ifosfamide 5 mg/m$^2$ every three weeks. The patient passed away after receiving three cycles of this chemotherapy regimen due to sudden cardiac arrest.

## Discussion

AUB is one of the common reasons for women to visit a gynecologist during the period of menstruation and menopause [7]. AUB can have various causes, one of which, although much less common, is cancer [8]. Endometrial cancer, vaginal rhabdomyosarcoma, and adenocarcinoma of the cervix can each cause abnormal uterine bleeding alone [9,10]. In this report, a case of high-grade endometrial stromal sarcoma was described in a 69-year-old woman who presented with abnormal vaginal bleeding and a pelvic mass, along with diagnostic and therapeutic measures. Considering the characteristics of this disease, it seems that all measures should be considered and initiated at the first visit.

ESS is a rare uterine tumor that mostly affects postmenopausal women. Due to the wide variety of pathological features and lack of patients, there is no comprehensive information about complications and symptoms, its different types in geographical distribution and specific races, and its possible relationship with other characteristics of patients' lives. The prognosis for patients is also not satisfactory, and for this reason, early diagnosis of the disease is necessary because patient survival is directly related to the stage of the tumor [11,12].

According to data from the US National Cancer Database, developed by the American Cancer Society and the US College of Surgeons Commission on Cancer, survival in patients with high-grade endometrial stromal sarcoma is extremely poor. The median overall survival is 19.9 months, and the five-year overall survival is only 32.6%. This statistic underscores the critical importance of relevant investigations. The most significant factors that reduce survival are 1) the age of patients, 2) tumor size, and 3) distal or nodal metastasis [2].

Some of these prognostic factors were observed very weakly in the examined patient. The reasons for this were the age over 65 years, the final stage of the tumor, and metastasis to the peritoneal and pelvic lymph nodes. It should be acknowledged that ESS

is malignant and can spread to the vagina, fallopian tubes, ovaries, bladder, and ureter, and distant metastasis to the lung, heart, and other sites has also been reported [13,14].

## Conclusion

The definitive approach to ESS, both for diagnosis and treatment, is surgery. In surgery, an enlarged uterus with a soft yellow necrotic and hemorrhagic tumor (especially with worm-like extension into pelvic veins) serves as a pointer for diagnosis. Definitive treatment is complete abdominal hysterectomy, bilateral salpingo-oophorectomy, and removal of all detectable masses [15].

## Ethical Considerations

### Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

### Conflict of Interests

The authors have no conflict of interest to declare.

### Ethical Approval

This study is approved under the ethical approval code of IR.ARUMS.REC.1402.046.

### Acknowledgment

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