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High-Grade Endometrial Stromal Sarcoma Mimicking an Ovarian Malignancy: A Case Report

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Case Report

Authors

Mohammad Reza Nateghi[1,2*], Sara Nejateslami[1]

1- Sarem Gynecology, Obstetrics and Infertility Research Center, Sarem Women's Hospital, Iran University of Medical Science (IUMS), Tehran, Iran.
2- Sarem Cell Research Center (SCRC), Sarem Women's Hospital, Tehran, Iran.

Corresponding Authors:
Mohammad Reza Nateghi; Sarem Fertility & Infertility Research Center (SAFIR), Sarem Women's Hospital, Iran University of Medical Sciences (IUMS), Tehran, Iran.
Address: Sarem Women Hospital, Basij Square, Phase 3, Ekbatan Town, Tehran, Iran. Postal code: 1396956111, Phone: +98 (21) 44670888, Fax: +98 (21) 44670432.

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ABSTRACT

Introduction: Endometrial Stromal Sarcoma (ESS) is a rare malignant uterine tumor originating from endometrial stromal cells. High-grade ESS is of particular clinical significance due to its aggressive biological behavior and potential for extrauterine spread. Diagnosing these tumors can be challenging, as their clinical manifestations are often non-specific, and their imaging features may resemble those of other pelvic masses. Reporting cases with atypical presentations can enhance clinical awareness and improve the diagnostic process.

Case Presentation: A 42-year-old woman with a long history of abnormal uterine bleeding, and a prior history of myomectomy and cystectomy, underwent evaluation at Sarem Subspecialty Hospital in Tehran due to worsening bleeding and the detection of a suspicious right ovarian mass (O-RADS 5). Following a laparotomy, which included a total abdominal hysterectomy, bilateral salpingo-oophorectomy, and lymphadenectomy, the patient was diagnosed with high-grade endometrial stromal sarcoma based on pathological and immunohistochemical findings. Pathological examination revealed myometrial invasion, lymphovascular invasion, and involvement of an internal iliac lymph node. The patient was referred to an oncologist for further treatment.

Conclusion: This case highlights the importance of considering a uterine origin for pelvic masses in patients presenting with a suspicious ovarian mass. High-grade endometrial stromal sarcoma can present with imaging features similar to those of ovarian malignancies; a definitive diagnosis requires careful histological and immunohistochemical evaluation. Therefore, when assessing pelvic masses accompanied by abnormal uterine bleeding, it is crucial to precisely determine the origin of the lesion and conduct comprehensive pathological studies.

Keywords: Endometrial Stromal Sarcoma; Pelvic Mass; Ovarian Malignancy; Abnormal Uterine Bleeding; Case Report.

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Introduction

Uterine sarcomas are a rare and heterogeneous group of mesenchymal malignancies that originate from muscle tissue, endometrial stroma, or uterine connective tissue. These tumors are among the most challenging malignancies in the field of gynecological oncology due to their low prevalence, morphological diversity and different biological behavior [1]. Endometrial stromal sarcoma (ESS) is one of the important subgroups of these malignancies, which originates from endometrial stromal cells and is classified according to the classification of the World Health Organization (WHO), in the group of endometrial stromal tumors with low grade, high grade (HG-ESS) and undifferentiated uterine sarcoma. This classification is of clinical importance; Because different subtypes have significant differences in terms of histological characteristics, molecular changes, aggressive behavior and prognosis [2, 16].

Among the subtypes of endometrial stromal sarcoma, the HG-ESS type is of particular importance due to its more aggressive biological behavior and the possibility of local and extrauterine spread. Pathologically, these tumors are usually characterized by round or spindle-shaped cells with significant nuclear atypia, high mitotic activity, infiltrative and destructive growth patterns, and sometimes tumor necrosis [4, 5]. Lymphovascular invasion is also a common finding in this group. The advancement of knowledge in the field of molecular biology has led to the identification of specific genetic changes in some subtypes of HG-ESS, including YWHAE::NUTM2 gene fusions and BCOR-related changes. These changes, along with the morphological characteristics and immunohistochemical profile, play an important role in the accurate classification and differentiation of these tumors from other uterine sarcomas [6, 7].

Clinically, the manifestations of uterine sarcomas are non-specific and may include abnormal uterine bleeding, pelvic pain or a sensation of pelvic pressure, and the presence of an abdominal or pelvic mass. These symptoms may overlap with more common benign conditions, particularly uterine leiomyomas, making preoperative diagnosis challenging [8]. Ultrasound and magnetic resonance imaging (MRI) are the primary modalities for the initial evaluation of uterine masses; however, imaging findings alone are insufficient to definitively confirm or rule out a sarcoma. Furthermore, in cases where imaging suggests a pelvic mass of ovarian origin, precisely determining its location and origin becomes particularly important, as certain uterine tumors can be clinically and radiologically mistaken for ovarian masses. In such scenarios, a comprehensive examination of the surgical specimen—incorporating morphological assessment, immunohistochemistry, and, when necessary, molecular studies—forms the basis for a definitive diagnosis [9, 10].

Given the rarity of high-grade endometrial stromal sarcoma (HG-ESS), its non-specific clinical presentation, and the difficulty of distinguishing it from other tumors of the female genital tract, detailed clinical case reports can help enhance physicians' awareness regarding the presentation patterns and diagnostic challenges associated with this condition. In this context, the present study describes a case of HG-ESS in a 42-year-old woman with a long-standing history of abnormal uterine bleeding. Initial imaging evaluations had revealed a mass in the right adnexa, classified as O-RADS 5 and suspected to be an ovarian malignancy. Intraoperative examination and final pathological assessment confirmed the uterine origin of the lesion. The presence of myometrial invasion, lymphovascular invasion, and involvement of an internal iliac lymph node, alongside normal-appearing ovaries, underscores the importance of a meticulous investigation into the origin of the mass and an accurate histological diagnosis. Consequently, this case report aims to highlight the challenges in the differential diagnosis of high-grade endometrial stromal sarcoma (HG-ESS) versus ovarian malignancies, emphasize the critical role of pathological and immunohistochemical evaluation, and draw greater clinical attention to the possibility of a uterine origin for pelvic masses otherwise suspected to be of ovarian malignant origin.

Case Presentation

The patient was a 42-year-old single woman who presented to Sarem Subspecialty Hospital in Tehran with a long-standing history of abnormal uterine bleeding (AUB). According to her medical history, she had experienced intermenstrual spotting since the age of 21, for which she had been treated with oral contraceptive pills (OCPs) for an extended period. Over time, she developed heavy uterine bleeding that did not respond adequately to hormonal therapy, including high-dose (HD) hormonal pills. In 2018 (1397 SH), she underwent a myomectomy and cystectomy due to the presence of multiple uterine fibroids and an ovarian cyst. The pathology report from that time indicated a submucosal fibroid and adenomyosis. Although her symptoms improved relatively for about a year following the surgery, the spotting recurred and persisted despite the use of hormonal medications. In the year leading up to her most recent presentation, the severity and duration of her uterine bleeding had increased. In May 2026, the patient presented complaining of abnormal uterine bleeding, specifically polymenorrhea and hypermenorrhea, and an ovarian mass detected via ultrasound. An MRI scan revealed adenomyosis and a uterine fibroid measuring approximately 50×50 mm (classified as FIGO type 2). Additionally, a mass measuring approximately 50×75 mm was observed in

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the right ovary; imaging assessment suggested potential malignancy, placing it in the O-RADS 5 category. No evidence of extrauterine involvement was found during the evaluation of other organs. Laboratory investigations revealed a hemoglobin level of 8.5, an FSH level of 2.7, and a CA-125 level of 58.89. An ultrasound confirmed the presence of a solid, hypoechoic lesion in the right ovary, measuring approximately 70×40 mm, with internal vascularization. Based on clinical and imaging findings, the initial differential diagnoses included dysgerminoma and sex cord-stromal tumors; the patient underwent a laparotomy with a suspicion of ovarian malignancy and uterine fibroids. On June 6, 2026, the patient underwent surgery. During the procedure, a lesion was observed lateral to the right side of the uterus, which was determined to originate from the uterus rather than the ovarian tissue. Given the sarcomatous appearance of the lesion, a sample was sent for intraoperative pathological assessment (frozen section). Additionally, a sample from the endometrium was submitted for pathological examination; the intraoperative results suggested an endometrial malignancy. Subsequently, a total abdominal hysterectomy (TAH), bilateral salpingo-oophorectomy (BSO), and lymphadenectomy were performed. Peritoneal fluid cytology was negative for malignant cells. Based on the final pathological examination and immunohistochemical (IHC) studies, a diagnosis of high-grade endometrial stromal sarcoma (HG-ESS) was confirmed. Myometrial involvement and lymphovascular space invasion (LVSI) were also noted. Both ovaries were pathologically normal; however, examination of the lymph nodes revealed involvement of one of the internal iliac lymph nodes by tumor cells. Following the completion of the surgical procedures and confirmation of the final diagnosis, the patient was referred to an oncologist for further evaluation and adjuvant treatment.

Discussion

High-grade endometrial stromal sarcoma (HG-ESS) is a rare and aggressive uterine malignancy that can pose diagnostic challenges due to its nonspecific clinical manifestations and imaging features that resemble other pelvic masses. The significance of this report lies in the disease's presentation as a mass suspicious for ovarian malignancy (O-RADS 5) and the discrepancy between the initial imaging-based impression and the final pathological diagnosis. In this case, a 42-year-old woman with a long-standing history of abnormal uterine bleeding and a suspicious right ovarian mass was evaluated; following surgery, pathological examination, and immunohistochemical analysis, a diagnosis of HG-ESS was confirmed. Final findings revealed myometrial invasion, lymphovascular invasion, and involvement of an internal iliac lymph node, while the ovaries were reported as normal. This case underscores the importance of carefully investigating the origin of pelvic masses and highlights the decisive role of pathology in establishing an accurate diagnosis.

In 2026, Chakalian et al. reported a case of high-grade endometrial stromal sarcoma with metastasis in a 42-year-old woman who presented with menorrhagia, pulmonary embolism, and recurrent pneumothoraces [11]. The authors emphasized the nonspecific nature of the initial symptoms, such as abnormal uterine bleeding, anemia, and pelvic pain, and the potential for diagnostic delays in this malignancy. The findings in the present case align with their report regarding the patient's age and the history of prolonged uterine bleeding. However, unlike the case reported by Chakalian et al., the pelvic mass in our patient was initially considered a potential ovarian malignancy, and the final diagnosis of HG-ESS was confirmed, with evidence of myometrial invasion, lymphovascular invasion, and lymph node involvement. This discrepancy may be attributed to the variability in the clinical presentations and patterns of spread of this tumor [11].

In 2021, Lee et al. reported a rare case of high-grade endometrial stromal sarcoma (HG-ESS) of primary ovarian origin in a 49-year-old woman who presented with tumor rupture, disseminated intravascular coagulation (DIC), and multiple peritoneal metastases [12]. Following initial surgery and chemotherapy, the patient underwent cytoreductive surgery combined with hyperthermic intraperitoneal chemotherapy (HIPEC). The present case is comparable to their report regarding the initial presentation of an ovarian mass and the final diagnosis of HG-ESS; however, unlike the case described by Lee et al., the tumor in our patient originated in the uterus, with the ovaries appearing normal upon pathological examination. This distinction underscores the importance of precisely determining the primary site of the tumor to differentiate uterine endometrial stromal sarcoma from a primary ovarian variant [12].

In 2015, Torres-de la Roche et al. reported a case of high-grade endometrial stromal sarcoma, specifically a myxoid variant, in a 36-year-old woman who presented with complaints of rapid abdominal enlargement and abdominal pain [13]. Initial evaluations suggested a possible uterine leiomyoma; however, histological, immunohistochemical, and genetic analyses confirmed a diagnosis of HG-ESS, characterized by high expression of *Cyclin D1* and a *YWHAE* gene rearrangement. Clinically, the present case aligns with their report regarding the difficulty of achieving a preoperative diagnosis and the necessity for supplementary pathological investigations. However, unlike the patient described in their report, who presented with a very large mass in the interligamentary region, our patient's pelvic mass was initially interpreted on imaging studies as a probable ovarian malignancy, whereas the final evaluation

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revealed its uterine origin. This discrepancy in clinical presentation and the initial localization of the mass may be attributed to differences in the tumor's site of growth, size, and pattern of spread. Overall, both reports underscore the non-specific nature of the clinical manifestations of HG-ESS and the importance of establishing a definitive diagnosis through histological and immunohistochemical analysis [13].

A case of high-grade endometrial stromal sarcoma (HG-ESS) in a 63-year-old postmenopausal woman was reported by Gregg in 2022; the patient had presented with postmenopausal bleeding and a pelvic mass [14]. During initial evaluations, considering a history of uterine fibroids, ultrasound findings, and an endometrial biopsy, the lesion was initially diagnosed as a uterine fibroid. However, further pathological and immunohistochemical analyses, combined with the confirmation of *YWHAE* gene rearrangement, established the diagnosis of HG-ESS. Clinically, the present case aligns with Gregg's report regarding the presence of abnormal uterine bleeding, a pelvic mass, and the difficulty of reaching an accurate initial diagnosis. Nevertheless, unlike the patient in the aforementioned report, whose mass was misdiagnosed as a uterine fibroid, our patient's imaging findings suggested an ovarian malignancy, whereas intraoperative assessment and pathology confirmed a uterine origin. Furthermore, while the patient in Gregg's report [14] was diagnosed with stage I disease without lymph node involvement, our case exhibited myometrial invasion, lymphovascular invasion, and involvement of an internal iliac lymph node. This discrepancy in the extent of disease spread may be attributed to differences in the stage at diagnosis and the biological characteristics of the tumor. Overall, both reports underscore the non-specific nature of the clinical manifestations of HG-ESS and highlight the necessity of including this malignancy in the differential diagnosis of pelvic masses, particularly in patients presenting with abnormal uterine bleeding.

In 2023, Salehi et al. reported a case of low-grade endometrial stromal sarcoma (LG-ESS) in a 52-year-old woman who presented with abnormal uterine bleeding [17]. Imaging studies revealed a large mass in the left ovary and a suspicious lesion in the uterus; the patient underwent surgery based on an initial diagnosis of an ovarian mass. Final pathological and immunohistochemical evaluations confirmed the presence of a uterine-origin LG-ESS with bilateral ovarian metastases. The present case aligns with their report regarding clinical presentation and the initial suspicion of an ovarian malignancy; however, a significant difference lies in the tumor grade and the pattern of ovarian involvement. In the study by Salehi et al. [17], the tumor was low-grade with bilateral ovarian metastases, whereas our patient was diagnosed with high-grade endometrial stromal sarcoma (HG-ESS), and both ovaries were found to be normal upon pathological examination. This distinction highlights those pelvic masses suspected of being ovarian malignancies may, in some instances, originate from the uterus; therefore, determining the primary site of the tumor and conducting a thorough pathological evaluation are essential for an accurate diagnosis.

Overall, a review of previous case reports indicates that high-grade endometrial stromal sarcoma can present diagnostic challenges due to non-specific clinical manifestations and imaging findings that resemble benign uterine masses or ovarian malignancies. Studies by Torres-de la Roche et al. (2015) and Gregg (2022) have highlighted the difficulty of preoperative diagnosis and the critical importance of histological and immunohistochemical examinations, as well as molecular studies, when necessary. Furthermore, Salehi et al. (2023), in a report of a case of low-grade endometrial stromal sarcoma (LG-ESS) with bilateral ovarian metastases, demonstrated that uterine-derived stromal tumors can manifest as suspicious ovarian masses. The findings of the present study align with these reports; the mass was initially interpreted on imaging as a probable ovarian malignancy, yet intraoperative assessment and final pathological evaluation confirmed its uterine origin. Nevertheless, the presence of myometrial invasion, lymphovascular invasion, and involvement of an internal iliac lymph node in this patient underscores the aggressive nature of the disease. Therefore, in patients presenting with abnormal uterine bleeding and pelvic masses suspected of being ovarian malignancies, it is essential to consider the possibility of a uterine origin and to conduct comprehensive pathological and immunohistochemical evaluations to ensure an accurate diagnosis and the selection of an appropriate therapeutic approach.

Conclusion

High-grade endometrial stromal sarcoma is a rare and aggressive malignancy that may present with nonspecific clinical manifestations and imaging findings, including masses suggestive of ovarian malignancy. This case report highlights that in patients presenting with abnormal uterine bleeding and pelvic masses, the possibility of a uterine origin for the lesion should be considered, even when imaging findings point toward an ovarian malignancy. A definitive diagnosis is established through detailed histological and immunohistochemical examination. Therefore, awareness of the atypical presentations of this tumor and achieving an accurate diagnosis are crucial for determining the extent of the disease and selecting an appropriate therapeutic strategy.

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Ethical Considerations
In conducting this research, all ethical principles in medical and biological research were observed in accordance with the Declaration of Helsinki, and the rights, dignity, and confidentiality of the participants were protected.

Conflict of Interests
There was no conflict of interest in this study.

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