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CASE REPORT

Spontaneous Rupture of a Low-Grade Endometrial Stromal Sarcoma: A Case Report and Review of Imaging

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Background: Endometrial stromal sarcoma (ESS) is a malignant tumor that develops from the mesenchymal tissue of the uterus. However, spontaneous rupture of ESS is uncommon. This case emphasizes the rarity of spontaneous rupture and the difficulties in preoperative diagnostic imaging, serving as a foundation for clinical diagnosis and prognosis.

Case Description: A 52-year-old woman presented at the clinic with right lower abdominal pain, persisting for over 5 days, significantly impacting her daily activities. Self-examination revealed a palpable abdominal mass. Ultrasonography identified an inhomogeneous echogenic mass within the uterus, initially suspected to be a uterine fibroid. Subsequent magnetic resonance imaging indicated uterine fibroids with tumor rupture and bleeding. The pathological assessment confirmed low-grade ESS. The patient underwent a total hysterectomy and double adnexectomy and is now 4 months postoperative and surviving well.

Conclusion: This case highlights the significance of diagnosing spontaneous rupture of ESS and emphasizes the need for increased awareness and additional research on this uncommon condition. Our results enhance comprehension of spontaneous ESS rupture and underscore the critical need for further investigation into optimal treatment approaches.

Keywords: endometrial stromal sarcoma, spontaneous rupture, magnetic resonance imaging, pathology

Introduction

Endometrioid stromal sarcoma (ESS) is a rare malignant tumor originating from endometrial stromal cells, representing approximately 1% of all uterine neoplasms and 10%–15% of uterine sarcomas, mainly affecting women aged 42 to 59 years, with an annual incidence of approximately 0.30/100,000 per year.[1–3] Clinical symptoms typically include abnormal uterine bleeding and pelvic pain.[3] Biomarkers, including lactate dehydrogenase and cancer antigen 125 (CA125), exhibit low sensitivity in predicting uterine sarcoma. Imaging diagnosis presents challenges; ultrasonography often shows a heterogeneous echogenic uterine mass with low-resistance blood flow but with limited sensitivity and specificity. MRI, however, displays characteristic features such as moderate-to-high T2WI signal intensity, restricted diffusion, and irregular enhancement. Low-grade ESS (LG-ESS) is identified by "worm-like cystic cavities", while high-grade ESS (HG-ESS) is linked to extensive necrosis and "feathery" enhancement.[1,4] The definitive diagnosis of ESS is established through postoperative histopathological examination, revealing features such as myxoid infiltration and intravascular tumor thrombi. LGESSs have distinct fingerlike patterns of myometrial invasion. Approximately 50% of LG-ESS cases involve gene fusions, including *JAZF1/PHF1*.[5] Additionally, FISH and/or targeted RNA sequencing can confirm the diagnosis of LG-ESS by identifying relevant gene fusions.[5] In contrast, HG-ESS is primarily associated with the t(10;17) (q23,p13) translocation and abnormalities in the *BCL-6* transcriptional corepressor gene. HG-ESS is typically characterized by larger size pronounced necrosis. Histologically, it often exhibits a mixed growth pattern, expansive, infiltrative, or both, and is frequently associated with LVSI, active mitosis, and tumor necrosis.[2,3,5] Treatment involves total hysterectomy; hormonal therapy can complement LG-ESS treatment, while HG-ESS necessitates combined systemic therapy.[1] LG-ESS is a slow-growing tumor with a 5-year overall survival (OS) rate of 80%–100%,

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whereas HG-ESS has a poor prognosis, with a median OS of 11–24 months and unfavorable outcomes in 70% of patients.[3]

In summary, spontaneously ruptured ESS is exceedingly rare in clinical practice, posing a significant challenge for preoperative diagnosis. It is frequently misdiagnosed as uterine fibroids, and rupture can have a detrimental impact on patient prognosis.[6,7] Accurate preoperative diagnosis is crucial for selecting treatment strategies and evaluating patient outcomes. Only two documented cases of spontaneously ruptured ESS exist in the literature.[4,8] Therefore, this study aims to present a case of spontaneously ruptured ESS in a 52-year-old woman. The case integrates imaging, pathological findings, and literature analysis to emphasize the importance of MRI in delineating tumors. The objective is to offer diagnostic insights for clinical practice, enhancing the early detection and prognostic management of such rare cases.

Case Presentation

The patient, a 52-year-old woman, experienced right lower abdominal pain five days ago, impacting her daily activities. She detected a mass without vaginal bleeding, fever, urinary symptoms, or constipation. Despite taking pain medication and anti-inflammatory drugs two days ago, she had inadequate relief. The patient had no history of hormonal medication use or familial tumor predisposition. Menstrual history: Previous regular menstruation, 5/28-30 days, moderate menstrual flow, dysmenorrhea (-), in the last 2 years. The patient presented with scanty menstruation, 5 days/2-3 months, last menstruation 09/2024. Marital history: G5P5, delivered by normal labor, children and spouses in good health. She had been sterilized 20 years ago and had a 1-year history of hypertension that was well-controlled medically. Gynecological examination showed a 10-cm tough mass in the anterior uterus and right adnexa, with unclear borders, poor mobility, and positive tenderness.

Laboratory tests showed abnormalities in blood parameters: white blood cell count (WBC) was elevated at 11.15×10^9/L, with neutrophils accounting for 71.41% (normal range: 50%–70%). Coagulation studies revealed prolonged prothrombinogen time (PT) at 13.6 seconds (reference range: 9.4–12.5), elevated fibrinogen (FIB) levels at 5.24 g/L (reference range: 2.38–4.98), increased D-dimer concentration at 1.13 mg/L (reference range: 0.00–0.24), and elevated fibrin degradation product (FDP) levels at 13.24 μg/mL (reference range: 0.00–2.01). Chorionic gonadotropin β-HCG was measured at 0.4 mIU/mL (normal level: ≤ 5.3). Ultrasound (Figure 1) showed an 8.4 cm × 7.6 cm × 8.1 cm inhomogeneous echogenic mass in the myometrium of the posterior wall of the uterus (Figure 1A), with punctate blood flow observed on CDFI (Figure 1B). The endocardial line was shifted anteriorly, and an inhomogeneous hypoechoic mass was identified in the right hypochondrium, leading to the diagnosis of possible leiomyosarcoma metaplasia and suggesting further investigation. A pelvic MRI plain scan (Figure 2) demonstrated an irregular mass in the myometrium of the posterior uterine wall, which was poorly demarcated from the normal uterine tissues. The lesion showed mixed high signals in T2WI (Figure 2A). Localized diffusion restriction was observed at the edge of the mass

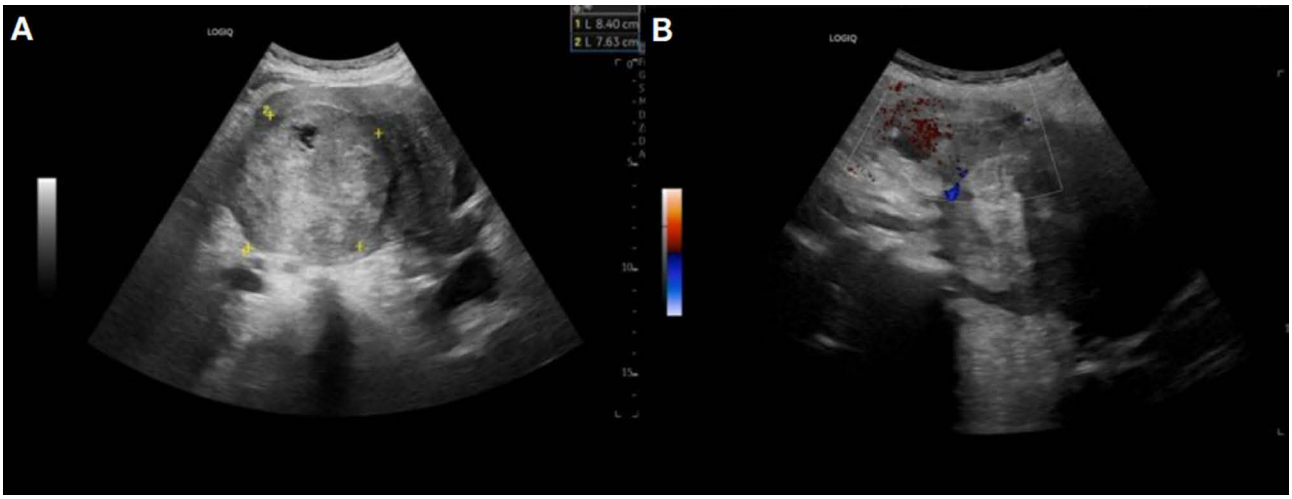

Figure 1 Ultrasound images (**A**) showing an inhomogeneous echogenic mass in the uterus (8.4 cm ×7.6 cm ×8.1 cm) and (**B**) punctate blood flow from the tumor.

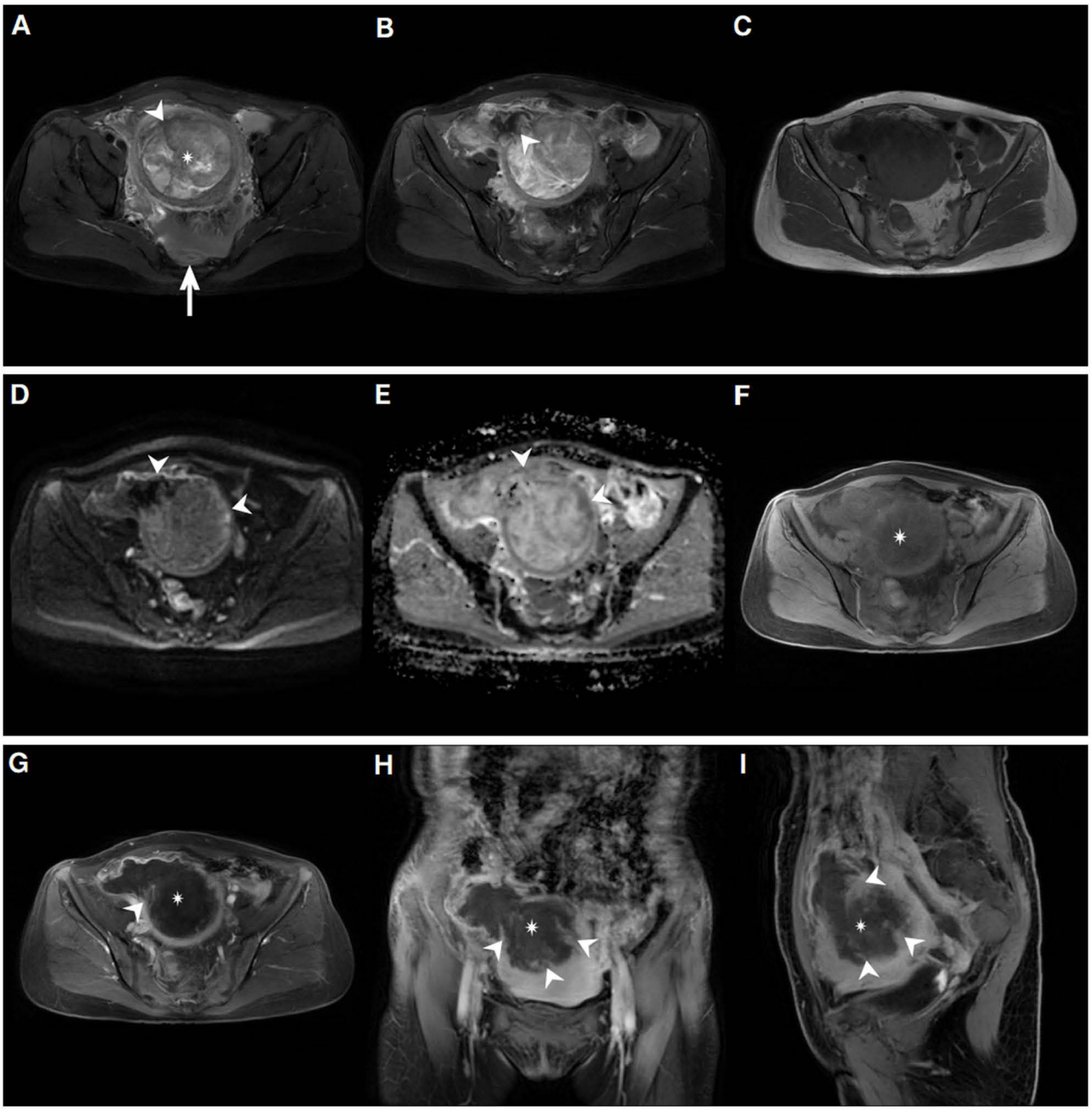

Figure 2 Pelvic MRI findings of uterine mass. (**A**) Axial T2-weighted SPAIR image showing an intermuscular mass with heterogeneous high-low mixed signals (asterisks), containing a hypointense band (white arrowheads) and posterior uterine hemorrhage (T2WI hypointensity, white long arrows). (**B**) Focal mass rupture with adjacent T2WI hypointensity indicating acute hemorrhage (white arrowheads). (**C**) Axial T1WI reveals isointense-to-hypointense signals (asterisk). (**D** and **E**) Diffusion-weighted imaging showing predominant hypointensity with peripherally restricted diffusion (white arrow, ADC 1.1–1.4×10-³mm²/s). (**F**) Fat-suppressed T1WI showing mild hypointensity. (**G–I**) Post-contrast sequences demonstrating central non-enhancing necrosis (asterisks), irregular feather-like peripheral enhancement (white arrows), and ill-defined infiltrative margins with adjacent myometrium.

(Figure 2D and E), with the lowest ADC value of approximately 1.1×10^{-3}mm^2/s. The contrast-enhanced scan revealed a large necrotic non-enhanced area (Figure 2G–I). Evidence of rupture was observed in the right anterior aspect of the lesion (Figure 2B), along with the presence of hemorrhagic effusion in the pelvic cavity (Figure 2A). Imaging findings suggested a diagnosis of a ruptured uterine fibroid with associated pelvic hemorrhagic effusion.

The patient underwent a laparoscopic transabdominal total hysterectomy at our hospital. During surgery, a mucinous nodule, approximately 10 cm in diameter, was found protruding from the posterior wall of the uterus, and the tumor

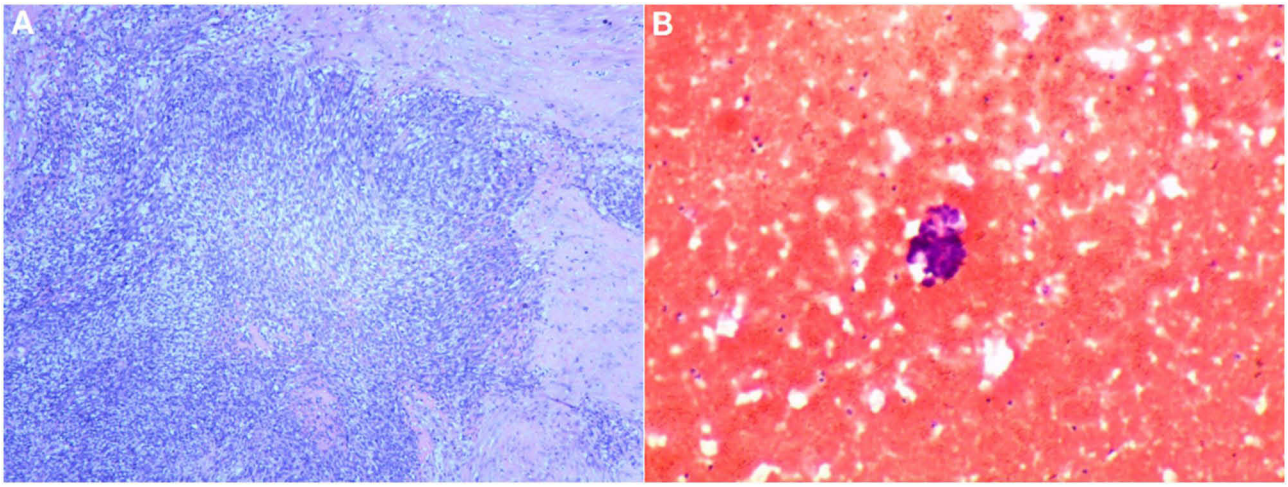

Figure 3 Pathologic findings suggestive of low-grade endometrial mesenchymal sarcoma. (**A**) Hematoxylin-eosin (H&E) staining showing small round cells with active nuclear division (< 5/10 HPF) and spindle-shaped cells infiltrating the myometrium. (×100 magnification). (**B**) Abdominal lavage cytology revealing nuclear heterogeneous cells with irregular nuclear contours and mild nuclear pleomorphism. (×20 magnification).

ruptured. The double accent and uterosacral ligaments were normal. Postoperative visual pathology showed a fragmented uterus measuring 15 cm × 12 cm × 3 cm, with a muscular wall thickness of 2–3 cm and rough endothelium of 3–4 mm. The free cervix was 2.5 cm long, with a lip thickness of 1–2 cm, and the circumference of the external cervical os was 4 cm, with the cervical mucosa still smooth. Multiple pieces of grayish-brown tissue, measuring 1–5 cm in diameter, were observed; the cut surfaces appeared solid and firm. Microscopically (Figure 3A), the uterine tissue was stained by H&E (×100) to show small round and spindle-shaped tumor cells in a sheet-like proliferation, with nuclear schizophrenia < 5 / 10HPF, infiltrating the myometrium. Abdominal lavage smear (Figure 3B), H&E staining ×20: nuclear heterogeneous cells were seen with irregular and mildly heterogeneous nuclei. Immunohistochemistry: CD10 (+); Ki-67 hotspot area: 60%+. The final diagnosis was a low-grade endometrial mesenchymal sarcoma. The patient underwent surgical treatment and was discharged 4 months after the postoperative follow-up. Currently, the patient is in good condition with no discomfort or symptoms.

Discussion

ESS is a rare malignant tumor that originates from endometrial mesenchymal stromal cells, with an annual incidence of about 0.30/100,000, predominantly affecting perimenopausal and postmenopausal women.[3] Its biological behavior is characterized by local infiltration, vascular invasion, and a high recurrence rate. The diagnosis of ESS necessitates a combination of imaging and pathology, with guidelines recommending MRI as the primary modality and pathology for confirming the diagnosis.[5] Ultrasound has limited diagnostic value for ESS, with an accuracy rate of only 5.1%.[9] Ultrasound in our patient also revealed a uterine inhomogeneous echogenic mass highly similar to uterine fibroids.[7] Studies have shown that MRI is significantly more accurate than ultrasound for the preoperative diagnosis of uterine tumors of mesenchymal origin (ESTs) (92.86% vs 67.86%) and may indicate the correct histologic subtype.[1,9]

Clinical reports of combined ESS ruptures are exceedingly rare, with only two cases documented in the global literature. Here, we present a novel case of LG-ESS rupture in a 52-year-old woman. We propose that the rupture stems from two synergistic pathological factors: extensive intratumoral necrosis, which increased intralesional pressure, and an exophytic growth pattern characterized by convex protrusion beyond the uterine serosal surface. Together, these factors contribute to the loss of capsular integrity and subsequent hemorrhagic complications. The exacerbation of abdominal pain may have been due to irritation of the intestinal canal and peritoneum by the ruptured tumor exudate. The patient's leukocyte and neutrophil percentages in laboratory tests were elevated, possibly linked to the inflammatory response triggered by the tumor rupture and peritoneal stimulation.

Table 1 Comparison of Clinical Characteristics Between the Present Case and Previous Cases of ESS Rupture

	Age (Years)	Tumor Diameter	Rupture Characteristics	Pathological Findings
Case 1	52	10 cm	Surface rupture, extensive necrosis within the tumor, bloody pelvic effusion	Low grade
Case 2	49	6–7 cm	Surface rupture with no active bleeding from the breach and yellowish pelvic fluid	Low grade
Case 3	69	15 cm	Rich blood supply, extensive necrosis, invasion of the sigmoid colon, and intraoperative hemorrhage	High grade

Notes: Case 1 is the case in this study; Cases 2[8] and 3[4] are quoted from the literature.

To systematically compare the characteristics of the ESS rupture cases, the cases in this study were integrated with previous reports in the literature (Table 1).

Table 1 shows that all cases of ESS rupture exhibited large tumor size (>6 cm), extensive intratumoral necrosis, and Location of the subplasma outgrowth. Collectively, these factors constitute a high-risk anatomical basis for rupture.

In this instance, preoperative ultrasonography and MRI indicated the potential presence of uterine fibroids, while the intraoperative frozen section pathology incorrectly diagnosed it as a benign lesion. This case highlighted substantial radiological resemblances among ESS, leiomyomas, and other uterine mesenchymal tumors, emphasizing the critical requirement for improved diagnostic discrimination methods.

Our patient exhibited MRI findings, including an indistinct mass border, rupture, moderate/high T2WI signal, T2WI low signal bands, extensive necrosis, feathery enhancement, limited DWI, and an ADC value of approximately (1.1–1.4) $\times 10^{-3}$ mm^2/s. The T2WI high signal differed significantly from the typical uterine fibroid's T2WI uniform low signal and well-defined boundary. ESS typically presents with T2WI isohigh signal and infiltrative growth characteristics. Imaging findings in this case overlapped with those of atypical smooth muscle tumors, cell-rich smooth muscle tumors, and cystic or mucinous degenerative smooth muscle tumors. Atypical metaplastic leiomyosarcomas may exhibit T2WI high signal and necrosis, while rich cell leiomyosarcomas may show restricted diffusion on DWI. Benign tumors generally have well-defined borders, unlike the infiltrative growth seen in ESS. Notably, ADC values can be diagnostically significant. Abdel Wahab C[10] et al suggested that ADC values $\leq 0.905 \times 10^{-3}$ mm^2/s could serve as a threshold for diagnosing uterine malignancy. Although this patient's ADC value did not meet the threshold, with a minimum value of approximately 1.1×10^{-3} mm^2/s, it could serve as a reference for diagnosing similar cases in the future. Considering the aforementioned imaging features, uterine and atypical fibroids can be excluded, leading to a preliminary diagnosis of malignant uterine tumors. Additional imaging features support differential diagnosis: a T2 low-signal band suggests LG-ESS, while feather-like enhancement and significant necrosis tend to indicate HG-ESS. The combined consideration of the diagnosis of ESS is highly likely and cannot be ruled out as HG-ESS.

This case was initially misdiagnosed as uterine leiomyosarcoma through intraoperative freezing but was later identified as LG-ESS based on gross pathology and immunohistochemistry findings. The tumor exhibited an infiltrative growth of myometrial mass invading the myometrium, with no vascular invasion observed microscopically. The tumor cells displayed infiltrative growth without vascular invasion, low nuclear schizophrenia of < 5/10HPF, and mild cellular anisotropy. Additionally, immunohistochemistry showed a CD10-positive. These manifestations are consistent with LG-ESS.[11] However, the abnormally elevated Ki-67 index (up to 60%) deviated significantly from the low proliferative activity usually observed in LG-ESS and even exceeded the commonly reported range for HG-ESS (10%–25%). This finding strongly suggests the possibility of HG-ESS.[11] Notably, studies have identified high Ki-67 expression, particularly levels ≥ 35%, as the threshold for predicting recurrence. This elevated expression has also been strongly associated with high aggressiveness, risk of recurrence, and poor prognosis of uterine sarcomas.[12] In the differential diagnosis, uterine smooth muscle tumors (exhibiting expansive growth and positive myogenic markers) and endometrial mesenchymal nodules (well-defined with no heterogeneity) were ruled out.[1,2] While the low nuclear atypia count favors LG-ESS, the CD10 marker lacks specificity for mesenchymal uterine tumors. Imaging revealed extensive necrosis and a characteristic "feather-like" enhancement within the lesion, features typical of HG-ESS.[1,11] Unfortunately, due to the lack of a pathologic specimen from the necrotic area for microscopic evaluation, a definitive diagnosis of HG-ESS could

not be confirmed. The combination of the extremely high Ki-67 proliferation index, imaging evidence of necrosis, and limitations of CD10 labeling raised doubts about the LG-ESS diagnosis and strongly suggested HG-ESS. Final confirmation necessitates molecular genetic testing, considering the potential bias from limitations in biopsy specimen analysis.[3]

The surgical approach must align with the recommended criteria for patients undergoing laparoscopic total hysterectomy with bilateral salpingo-oophorectomy. Postoperative adjuvant therapy demands personalized protocols; LG-ESS mainly needs antiestrogen therapy, while HG-ESS usually requires combined chemotherapy and radiotherapy.[5,9] However, the patient did not start adjuvant therapy post-surgery. Therefore, enhancing molecular pathology testing is advised to elucidate tumor grading and develop adjuvant therapy strategies according to the grading results for precise management.

The prognosis of ESS is closely related to the histological grading. The LG-ESS reported by Zeng et al[8] received postoperative external pelvic irradiation along with megestrol, resulting in no recurrence at a six-month follow-up. Conversely, Chia et al[4] reported a case of HG-ESS where the patient succumbed to disease progression three months post-surgery despite undergoing total hysterectomy and bilateral adnexectomy, attributed to tumor rupture with sigmoid colon invasion. This control highlights the decisive impact of histologic grading on survival outcomes.

Compared with the ESS cases reported in the literature, the present case benefits from multimodal imaging data and prompt intraoperative pathologic verification. However, it is constrained by a limited number of pathologic images and a lack of long-term follow-up results. These differences suggest that the present case achieved a high level of diagnostic thoroughness. However, enhancing follow-up procedures and supplementing pathological data are necessary to comprehensively evaluate tumor biological behavior and clinical outcomes.

The definitive diagnosis of ESS depends on pathological examination, emphasizing the significance of precise preoperative assessment in clinical practice. MRI serves as a crucial diagnostic tool, aiding in surgical planning and offering essential guidance for postoperative adjunct therapy.

Conclusion

This study presents a rare case of spontaneous rupture of endometrial mesenchymal sarcoma, along with postoperative pathological findings, to provide case data for research on endometrial mesenchymal sarcoma. The aim is to reduce the misdiagnosis rate and offer clinical experience for subsequent treatment and prognostic management of patients. The precise management of ESS relies on a multimodal assessment that integrates imaging, pathology, and molecular markers, in addition to standardized procedure selection and individualized adjuvant therapy. Clinicians must fully recognize the diagnostic complexity and therapeutic variability of ESS to enhance patient outcomes, as emphasized by the insights from this case.

Ethics Approval and Informed Consent

The present case study received ethical approval from the Institutional Review Board at Jincheng People's Hospital (Ethics Approval JCPH.NO20250314001). The approval covers collection, analysis of case data, and publication of the report, ensuring compliance with ethical standards for patient privacy protection and information disclosure.

Acknowledgment

The case report follows the CARE guidelines. Written informed consent was obtained from the patient for the publication of this case report, including any accompanying clinical data and images.

Disclosure

The authors confirm the absence of any competing financial or nonfinancial interests related to the conduct and reporting of this study.

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