Low-Grade Endometrial Stromal Sarcoma Presenting as a Giant Uterine Mass in a Perimenopausal Woman: A Case Report

DR. G.RADHIKA[1], DR. R.GOVINDARAJAN[2], DR. R.REVATHI SHREE[3], DR.P.PRIYANKA[4]

1. Final Year Postgraduate, Department of Pathology, Shri Sathya Sai Medical College & Research Institute, (Sri Balaji Vidyapeeth University (DU), Chennai campus), Ammapettai, Nellikuppam Post, Chengalpattu District, Tamilnadu, 603108, India
2. Professor & Head, Department of Pathology, Shri Sathya Sai Medical College & Research Institute, (Sri Balaji Vidyapeeth University (DU), Chennai campus), Ammapettai, Nellikuppam Post, Chengalpattu District Tamilnadu, 603108, India
3. Professor, Department of Pathology, Shri Sathya Sai Medical College & Research Institute, (Sri Balaji Vidyapeeth University (DU), Chennai campus), Ammapettai, Nellikuppam Post, Chengalpattu District Tamilnadu, 603108, India
4. Professor, Department of Pathology, Shri Sathya Sai Medical College & Research Institute, (Sri Balaji Vidyapeeth University (DU), Chennai campus), Ammapettai, Nellikuppam Post, Chengalpattu District Tamilnadu, 603108, India

Abstract

Low-grade endometrial stromal sarcoma (LG-ESS) is a rare malignant mesenchymal neoplasm of the uterus, accounting for less than 1% of all uterine malignancies and characterized by an indolent clinical course with the potential for late recurrence. The nonspecific clinical presentation frequently leads to misdiagnosis as benign uterine leiomyoma, making histopathological examination essential for definitive diagnosis. We report the case of a 49-year-old perimenopausal woman who presented with progressively increasing abdominal distension and intermittent lower abdominal pain for three months. Clinical evaluation revealed a large uterine mass, and the patient underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy. Gross examination demonstrated a large grey-white tumour measuring 20 × 20 × 18 cm with focal cystic degeneration and necrosis. Histopathological examination revealed diffuse proliferation of uniform endometrial stromal cells with round to oval nuclei, minimal cytological atypia, occasional spindle cell morphology, abundant myxoid stroma, infiltrative myometrial growth, and prominent lymphovascular invasion, confirming the diagnosis of low-grade endometrial stromal sarcoma. The tumour was staged as pT1b pNx pMx. The postoperative period was uneventful, and the patient was advised long-term follow-up because of the known risk of delayed recurrence. This case highlights the importance of considering LG-ESS in the differential diagnosis of large uterine masses in perimenopausal women. Careful gross examination, meticulous histopathological evaluation, and complete surgical excision remain the cornerstones of diagnosis and management. Early-stage disease is associated with an excellent prognosis; however, prolonged surveillance is essential because recurrence may occur many years after initial treatment.

Keywords: Low-grade endometrial stromal sarcoma, uterine sarcoma, endometrial stromal tumour, hysterectomy, lymphovascular invasion, pathology.

How to cite this article: Radhika G, Govindarajan R, Revathi Shree R, Priyanka P. Low-Grade Endometrial Stromal Sarcoma Presenting as a Giant Uterine Mass in a Perimenopausal Woman: A Case Report. Int J Drug Deliv Technol. 2026;16(69s):1040-1045. DOI: 10.25258/ijddt.16.69s.107

Abstract

Low-grade endometrial stromal sarcoma (LG-ESS) is a rare malignant mesenchymal neoplasm accounting for less than 1% of all uterine malignancies and approximately 10–15% of uterine sarcomas. Despite its indolent biological behaviour, LG-ESS possesses significant malignant potential because of its characteristic infiltrative growth pattern, lymphovascular invasion, and propensity for late local or distant recurrence. The diagnosis is often challenging due to its nonspecific clinical presentation and considerable overlap with benign uterine leiomyomas and other mesenchymal tumours. Recent advances in molecular pathology, particularly identification of recurrent chromosomal translocations such as JAZF1-SUZ12 fusion, have substantially improved diagnostic accuracy and understanding of tumour pathogenesis.

Low-Grade Endometrial Stromal Sarcoma Presenting as a Giant Uterine Mass in a Perimenopausal Woman: A Case Report

We report a case of a 49-year-old perimenopausal woman who presented with progressively increasing abdominal distension and intermittent abdominal pain. Radiological evaluation demonstrated a large uterine mass, and the patient subsequently underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy. Histopathological examination revealed a low-grade endometrial stromal sarcoma showing diffuse proliferation of uniform endometrial stromal cells with infiltrative myometrial invasion and prominent lymphovascular invasion. The tumour was staged as pT1b pNx pMx. This case highlights the importance of meticulous histopathological evaluation in differentiating LG-ESS from its histological mimics and emphasizes the role of complete surgical excision for favourable long-term outcomes.

Keywords: Low-grade endometrial stromal sarcoma, uterine sarcoma, endometrial stromal tumour, hysterectomy, lymphovascular invasion, pathology

How to cite this article: Radhika G, Govindarajan R, Revathi Shree R, Priyanka P. Low-Grade Endometrial Stromal Sarcoma Presenting as a Giant Uterine Mass in a Perimenopausal Woman: A Case Report. Int J Drug Deliv Technol. 2026;16(69s):1040. DOI: 10.25258/ijddt.16.69s.107

Introduction

Endometrial stromal sarcomas (ESS) are uncommon malignant mesenchymal tumours originating from the specialized endometrial stromal cells of the uterus. They constitute approximately 0.2% of all uterine malignancies and represent nearly 10–15% of uterine sarcomas, making them one of the rarest gynecological malignancies. According to the latest WHO Classification of Female Genital Tumours, endometrial stromal tumours are classified into four major categories: endometrial stromal nodule, low-grade endometrial stromal sarcoma (LG-ESS), high-grade endometrial stromal sarcoma (HG-ESS), and undifferentiated uterine sarcoma.

Among these entities, LG-ESS is characterized by relatively indolent biological behaviour, low mitotic activity, infiltrative myometrial growth, and frequent lymphovascular invasion. Although the tumour demonstrates slow progression, recurrence may occur even decades after initial treatment, necessitating prolonged follow-up. The disease predominantly affects women between 40 and 55 years of age and commonly presents with abnormal uterine bleeding, pelvic pain, dysmenorrhoea, abdominal distension, or a slowly enlarging uterine mass. Occasionally, patients may remain asymptomatic until the tumour reaches considerable size.

The pathogenesis of LG-ESS has been increasingly elucidated through advances in molecular pathology. The majority of tumours harbour recurrent chromosomal rearrangements involving the **JAZF1-SUZ12** gene fusion, while other fusion genes including **JAZF1-PHF1**, **EPC1-PHF1**, and **MEAF6-PHF1** have also been identified. These molecular alterations have become valuable ancillary diagnostic tools, particularly in morphologically challenging cases.

Histologically, LG-ESS closely resembles proliferative-phase endometrial stromal cells and exhibits characteristic worm-like infiltration into the myometrium with extensive lymphovascular invasion. Immunohistochemically, tumour cells usually express CD10, estrogen receptor (ER), progesterone receptor (PR), and WT1, while smooth muscle markers such as SMA and desmin may show only focal positivity, facilitating differentiation from cellular leiomyoma.

Although surgical resection remains the cornerstone of treatment, hormonal therapy has emerged as an important adjuvant modality because of the hormone receptor positivity observed in most cases. Stage at diagnosis remains the strongest prognostic indicator, with reported 5-year survival rates exceeding 90% for Stage I disease but decreasing substantially in advanced stages.

The present case describes a giant low-grade endometrial stromal sarcoma in a perimenopausal woman, highlighting the diagnostic challenges and classical histopathological features that enabled definitive diagnosis.

Case Presentation

A 49-year-old multiparous perimenopausal woman presented to the Department of Obstetrics and Gynaecology with complaints of intermittent lower abdominal pain and gradually progressive abdominal distension for approximately three months. The abdominal swelling had progressively increased in size during this period and was associated with a sensation of heaviness. There was no significant history of abnormal vaginal bleeding, constitutional symptoms, or weight loss. Her last menstrual period had occurred three months before presentation, consistent with the perimenopausal transition.

General physical examination revealed a stable patient with no evidence of pallor, icterus, lymphadenopathy, or pedal oedema. Systemic examination was otherwise unremarkable. Abdominal examination demonstrated a large

abdominopelvic mass corresponding to an enlarged uterus.

Radiological evaluation suggested a large uterine mass, raising the differential diagnoses of degenerating leiomyoma, uterine sarcoma, or other mesenchymal neoplasms. Considering the patient's age, clinical presentation, and imaging findings, surgical management was planned.

The patient subsequently underwent **total abdominal hysterectomy with bilateral salpingo-oophorectomy (TAH-BSO)**. The postoperative period was uneventful, and the surgical specimen was submitted for detailed histopathological examination.

Gross Examination

The hysterectomy specimen consisted of the uterus with bilateral adnexa. Attached to the uterus was a large tumour mass measuring **20 × 20 × 18 cm**.

On external examination, the uterus was markedly enlarged by a well-defined but infiltrative mass arising from and attached to the myometrium. Cut section revealed a predominantly solid grey-white tumour with multiple cystic spaces, extensive areas of necrosis, and a firm fleshy consistency. The tumour displayed irregular infiltrative margins extending into the surrounding myometrium. No obvious haemorrhagic degeneration was identified grossly. Bilateral ovaries and fallopian tubes appeared grossly unremarkable.

Microscopic Examination

Histopathological examination demonstrated a diffuse proliferation of tumour cells closely resembling proliferative-phase endometrial stromal cells. The tumour was composed of uniform, monotonous small round to oval cells arranged in sheets and short fascicles. Individual cells exhibited scant amphophilic cytoplasm with indistinct cell borders.

The nuclei were round to oval with finely dispersed chromatin and inconspicuous nucleoli. Mild nuclear atypia was present without significant pleomorphism. Occasional spindle cell morphology was also observed.

The tumour was embedded within abundant myxoid stroma. One of the most characteristic microscopic findings was extensive infiltration of the surrounding myometrium accompanied by prominent lymphovascular invasion, producing the classical permeative growth pattern of low-grade endometrial stromal sarcoma.

Mitotic activity was low, consistent with a low-grade neoplasm. No significant tumour necrosis or marked cytological atypia suggestive of high-grade transformation was identified.

Overall histomorphological features were diagnostic of **Low-Grade Endometrial Stromal Sarcoma**.

Differential Diagnosis

The major histopathological differential diagnoses considered included:

- Endometrial stromal nodule
- High-grade endometrial stromal sarcoma
- Cellular leiomyoma

The infiltrative myometrial growth pattern together with definite lymphovascular invasion excluded endometrial stromal nodule. The absence of marked nuclear pleomorphism, high mitotic activity, and significant atypia ruled out high-grade ESS. Lack of characteristic smooth muscle fascicular architecture and the classical endometrial stromal morphology differentiated the lesion from cellular leiomyoma.

Final Histopathological Diagnosis

Low-Grade Endometrial Stromal Sarcoma (LG-ESS)

Pathological Stage: pT1b pNx pMx

Discussion

Low-grade endometrial stromal sarcoma (LG-ESS) is a rare uterine mesenchymal malignancy accounting for less than 1% of all uterine malignancies and approximately 10–15% of uterine sarcomas. It is characterized by an indolent clinical course, infiltrative myometrial growth, and a tendency for late local or distant recurrence despite its relatively favourable prognosis **[1]**. The present case involved a 49-year-old perimenopausal woman, which is comparable to the age distribution reported by Schoolmeester et al. **[1]**, who observed that LG-ESS predominantly affects women in the fourth and fifth decades of life. Thus, the demographic characteristics of our patient are consistent with the existing literature.

Clinically, our patient presented with progressively increasing abdominal distension and intermittent abdominal pain for three months. Unlike the common presentation of abnormal uterine bleeding described by Smith et al. **[2]**, our patient primarily complained of abdominal enlargement due to the massive uterine tumour. Smith et al. also emphasized that many patients are initially

misdiagnosed with leiomyoma because of overlapping clinical and radiological findings. Similarly, the diagnosis in our patient was established only after histopathological examination following hysterectomy, highlighting the diagnostic challenge associated with LG-ESS.

Gross examination demonstrated a large **20 × 20 × 18 cm** solid grey-white tumour with cystic degeneration and focal necrosis arising from the myometrium. Although most LG-ESSs reported by Kikuchi et al. **[3]** measured considerably smaller, their multicentre study showed that tumour stage rather than tumour size is the principal determinant of prognosis. Likewise, despite the unusually large tumour in our patient, the disease remained confined to the uterus (pT1b), indicating a favourable prognostic outlook following complete surgical excision.

Microscopically, the tumour exhibited diffuse proliferation of uniform endometrial stromal cells with round to oval nuclei, minimal cytological atypia, occasional spindle cell morphology, abundant myxoid stroma, infiltrative myometrial invasion, and prominent lymphovascular invasion. These findings closely resemble the characteristic histopathological features described by Kolin et al. **[4]**, who demonstrated that infiltrative growth and vascular permeation remain the most reliable diagnostic features of LG-ESS. Similar to their observations, our case showed low mitotic activity and lacked marked pleomorphism or extensive tumour necrosis, thereby excluding high-grade endometrial stromal sarcoma.

An important pathological feature in our case was extensive lymphovascular invasion, which helped differentiate LG-ESS from endometrial stromal nodules. Huang and Peng **[5]** also reported that infiltrative myometrial growth and vascular invasion are characteristic pathological findings associated with malignant behaviour and potential recurrence. Although molecular analysis was not performed in our patient, recent studies have demonstrated that recurrent gene fusions such as **JAZF1-SUZ12** play an important role in the pathogenesis and diagnosis of LG-ESS, particularly in histologically challenging cases **[4,5]**.

The patient underwent **total abdominal hysterectomy with bilateral salpingo-oophorectomy**, which remains the recommended treatment for localized LG-ESS. Borella et al. **[6]** reported that complete surgical excision offers excellent long-term disease control, while ovarian preservation may increase the risk of recurrence in selected patients. Therefore, the surgical management adopted in our patient was consistent with current evidence-based recommendations for perimenopausal women with localized disease.

Although no adjuvant hormonal therapy was administered in the present case because the tumour was completely excised and confined to the uterus, hormonal therapy has become an important treatment option for recurrent or advanced disease. Wu et al. **[7]** demonstrated favourable outcomes with endocrine therapy using progestins or aromatase inhibitors in hormone receptor-positive tumours, whereas chemotherapy has shown limited benefit in low-grade disease. Consequently, current management strategies emphasize individualized treatment based on tumour stage, hormone receptor status, and recurrence risk.

The pathological stage in our patient was **pT1b pNx pMx**, indicating disease limited to the uterus. Li et al. **[8]** reported that early-stage LG-ESS is associated with excellent long-term survival following complete surgical resection, although recurrence may occur several years after initial treatment. Similar to their findings, our patient is expected to have a favourable prognosis because of complete tumour excision and localized disease. Nevertheless, prolonged clinical follow-up with periodic imaging remains essential because of the well-recognized potential for late recurrence even after apparently curative surgery.

Conclusion

Low-grade endometrial stromal sarcoma is an uncommon uterine mesenchymal malignancy that often presents with nonspecific clinical features, making preoperative diagnosis difficult. The present case demonstrates that even a giant uterine mass may represent LG-ESS and should be considered in the differential diagnosis of large myometrial tumours in perimenopausal women. Classical histopathological features, including infiltrative myometrial growth and lymphovascular invasion, remain the key diagnostic hallmarks. Complete surgical excision by total abdominal hysterectomy with bilateral salpingo-oophorectomy continues to be the treatment of choice for localized disease and offers an excellent prognosis when diagnosed at an early stage. Nevertheless, because of the potential for late recurrence, prolonged clinical follow-up is mandatory to ensure timely detection and management of recurrent disease.

GROSS:

FIGURE 1

Figure 1. Gross photograph of the total abdominal hysterectomy with bilateral salpingo-oophorectomy specimen showing a large infiltrative grey-white fleshy tumour arising from the uterus with extension into the myometrium.

FIGURE -2

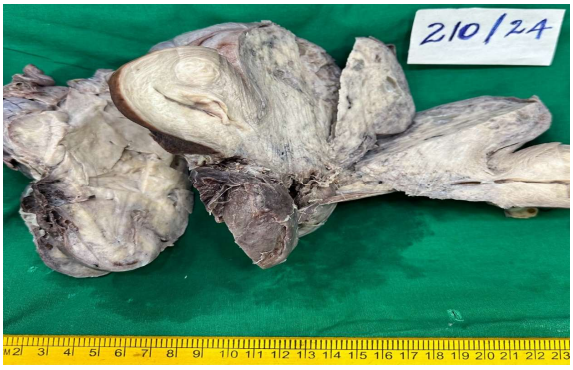


Figure 2. Cut section of the uterine tumour showing a solid grey-white fleshy mass with focal cystic degeneration and infiltrative margins, consistent with low-grade endometrial stromal sarcoma.

MICROSCOPY

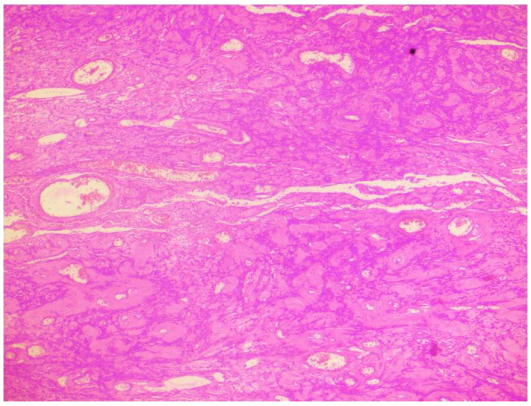


Figure 3. Scanner view (Hematoxylin and Eosin, ×4) showing an infiltrative endometrial stromal neoplasm permeating the myometrium.

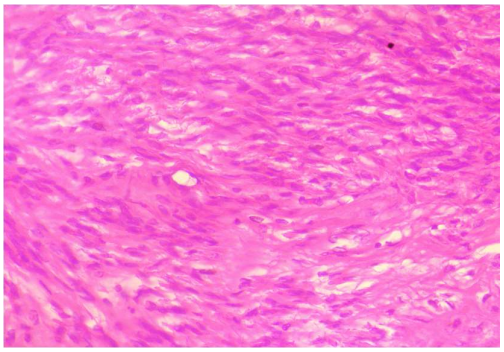


Figure 4. Low-power photomicrograph (Hematoxylin and Eosin, ×10) demonstrating uniform proliferation of endometrial stromal cells arranged in sheets and short fascicles within abundant myxoid stroma.

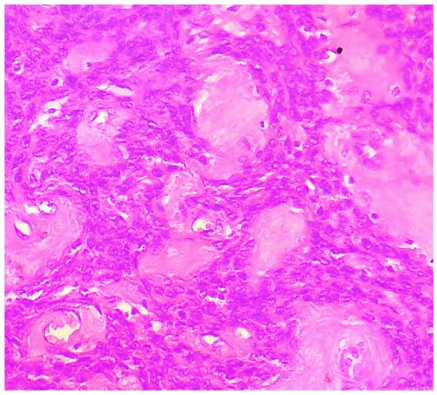
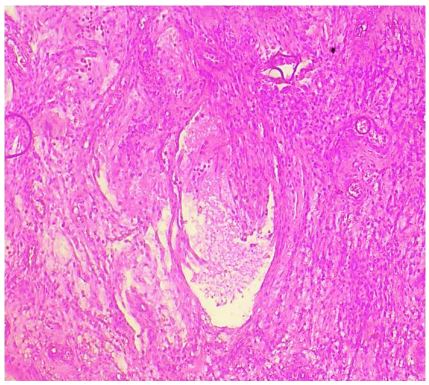


Figure 5. High-power photomicrograph (Hematoxylin and Eosin, ×40) showing monotonous round-to-oval tumour cells with scant cytoplasm, finely dispersed chromatin, inconspicuous nucleoli, and minimal cytological atypia.



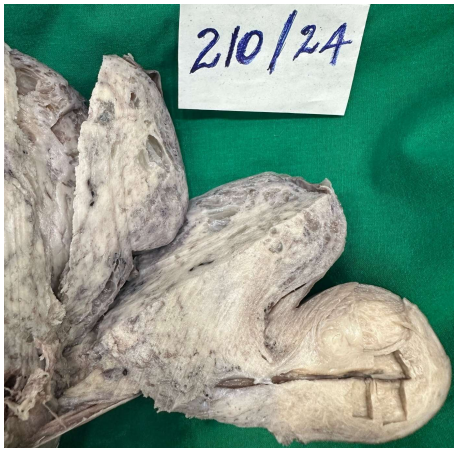


Figure 6. High-power photomicrograph (Hematoxylin and Eosin, ×40) demonstrating characteristic lymphovascular invasion by tumour cells, a hallmark feature of low-grade endometrial stromal sarcoma.

References

1. Schoolmeester JK, et al. Low-grade endometrial stromal sarcoma: expanding the morphologic spectrum and clinicopathologic correlations. **Am J Surg Pathol.** 2025. PMID: 40434060.
2. Smith ES, Jansen C, Miller KM, Chiang S, Alektiar KM, Hensley ML, et al. Primary characteristics and outcomes of newly diagnosed low-grade endometrial stromal sarcoma. **Int J Gynecol Cancer.** 2022;32(7):882-890. doi:10.1136/ijgc-2022-003383. PMID:35641004.
3. Kikuchi A, Sato S, et al. Clinical characteristics and prognostic factors of endometrial stromal sarcoma and undifferentiated uterine sarcoma confirmed by central pathological review: a multi-institutional retrospective study from the Japanese Clinical Oncology Group. **Gynecol Oncol.** 2023;171:64-72. PMID:37478616.
4. Kolin DL, et al. Molecular alterations in low-grade endometrial stromal sarcoma and their clinicopathologic significance. **Mod Pathol.** 2023.
5. Huang X, Peng P. Hormone therapy reduces recurrence in stage II–IV uterine low-grade endometrial stromal sarcomas: a retrospective cohort study. **Front Oncol.** 2022;12:922757. doi:10.3389/fonc.2022.922757.
6. Borella F, Bertero L, Cassoni P, Piovano E, Gallio N, Preti M, et al. Low-grade uterine endometrial stromal sarcoma: prognostic analysis of clinicopathological characteristics, surgical management, and adjuvant treatments. **Front Oncol.** 2022;12:883344. doi:10.3389/fonc.2022.883344.
7. Wu Y, Li N, Zhang R, Bai P. Primary low-grade extrauterine endometrial stromal sarcoma: analysis of 10 cases with a review of the literature. **World J Surg Oncol.** 2022;20:17. doi:10.1186/s12957-021-02474-1. PMID:35027071.
8. Li X, et al. Prognosis of uterine and extrauterine low-grade endometrial stromal sarcoma: an observational cohort study. **Int J Surg.** 2024.