

# Low-Grade Endometrial Stromal Sarcoma in a Young Woman: A Case Report

Maryam Talayeh<sup>1</sup> , Maliheh Arab<sup>1\*</sup>, Somayyeh Noei Teymoordash<sup>2</sup> , Noushin Afsharmoghadam<sup>3</sup> ,  
Masoomeh Raoufi<sup>4</sup>, Behnaz Ghavami<sup>5</sup> , Mahsa Asghari<sup>1</sup>

1. Department of Gynaeco-oncology, Imam Hossein Medical Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran
2. Department of Gynaeco-oncology, Firuzgar Medical Center, Iran University of Medical Sciences, Tehran, Iran
3. Department of Pathology, Imam Hossein Medical Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran
4. Department of Radiology, Imam Hossein Hospital, Shahid Beheshti University of Medical Sciences, Tehran, Iran
5. Fellowship of Laparoscopy, Tehran, Iran



## Article Info

 [10.30699/jogcr.7.6.569](https://doi.org/10.30699/jogcr.7.6.569)

**Received:** 2021/06/03;

**Accepted:** 2021/10/16;

**Published Online:** 09 Sep 2022;

Use your device to scan and read the article online



## Corresponding Information:

**Maliheh Arab,**  
Department of Gynaeco-oncology, Imam Hossein Medical Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran

**Email:** [drmarab@sbm.ac.ir](mailto:drmarab@sbm.ac.ir)

## ABSTRACT

Endometrial stromal sarcoma is a rare condition, constituting nearly 0.2% of all reproductive tract malignancies. The condition mainly affects the peri-menopausal population; however, it can occasionally be found in younger women and adolescents. Here, this is a case of a 20-year-old primiparous female who was referred to us with the main complaint of vaginal bleeding (menometrorrhagia), ongoing for 6 months after delivery. Her sonography report indicated a 5 cm intra-cavitary mass suspicious for myoma or placental polyp. Given the pathology report of low-grade endometrial stromal sarcoma (LGESS) on samples obtained through hysteroscopy and D&C, she underwent total abdominal hysterectomy and bilateral salpingo-oophorectomy (TAH+BSO). Endometrial stromal sarcoma is a rare uterine malignancy of mesenchymal origin that should be considered, even in very young patients.

**Keywords:** Endometrial stromal sarcoma, Hysteroscopy, Low-grade, Uterine sarcoma



Copyright © 2022, This is an original open-access article distributed under the terms of the Creative Commons Attribution-noncommercial 4.0 International License which permits copy and redistribution of the material just in noncommercial usages with proper citation.

## Introduction

Constituting only 0.2% of uterine malignancies and nearly 20% of uterine sarcomas, low-grade endometrial stromal sarcoma (LGESS) is an uncommon neoplasm of the reproductive tract in women (1).

While other uterine cancers show age predilection towards the postmenopausal population, ESS mainly presents in peri-menopausal women. Clinically, ESS is often misdiagnosed as benign conditions, e.g., leiomyoma; the diagnosis is usually confirmed post-operatively based on histopathological evaluation of tumor specimens (2, 3).

Given its nonspecific clinical manifestations, diagnosis of ESS is often delayed, negatively affecting its prognosis (4).

ESS has recently been reclassified by WHO into three categories, namely: low-grade ESS (LG-ESS),

high-grade ESS (HG-ESS), and undifferentiated uterine sarcoma. Lesion's morphological similarity to endometrial stromal cells in their proliferative phase, the mitotic rate, and clinical behavior were the three factors on which this classification was based (5).

Given its post-hysterectomy discovery in most cases and the rarity of the condition, hysterectomy has consistently been the treatment approach considered for ESS in literature (6).

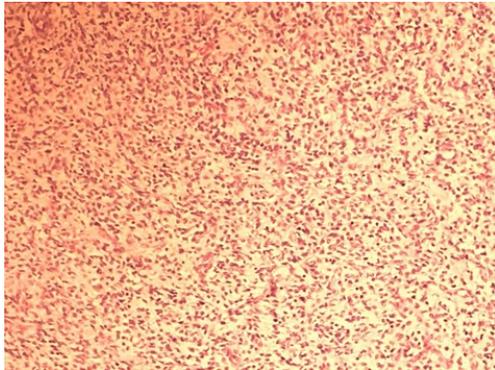
The present case is reported due to the patient's young age at diagnosis and the disease's rarity.

## Case Presentation

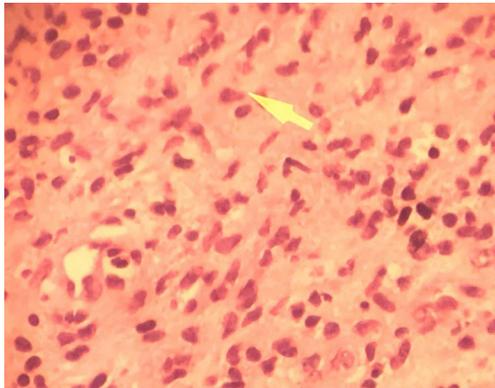
We report a 20-year-old primiparous female with a history of vaginal delivery 6 months before her referral with a complaint of vaginal bleeding

(menometrorrhagia). On transvaginal sonography, she had an intracavitary lesion measuring about 5 cm, suspicious of myoma or placental polyp. She underwent hysteroscopy, D&C, and colposcopy. On hysteroscopy, a 5 cm mass was found on the anterior wall of the uterus, which was resected and sent for pathologic examination and the colposcopy biopsy result was normal. The pathology report showed low-grade endometrial stromal sarcoma (Low-grade ESS)

(Figure 1 A, B). A pathology review confirmed low-grade ESS. IHC study indicated positive Vimentin and CD10 (Figure 1) but was negative for CK and WT1. Her case was discussed in a multidisciplinary session, and she decided to get a second pathology review. A pathology review could not discriminate between endometrial stromal nodule and low-grade endometrial stromal sarcoma.



A: Photomicrograph of low-grade stromal sarcoma showing proliferation of spindle shaped cells (H&E staining ×100)



B: High power field view of low-grade stromal sarcoma composed of mild atypical spindle shaped cells (H&E staining ×400).

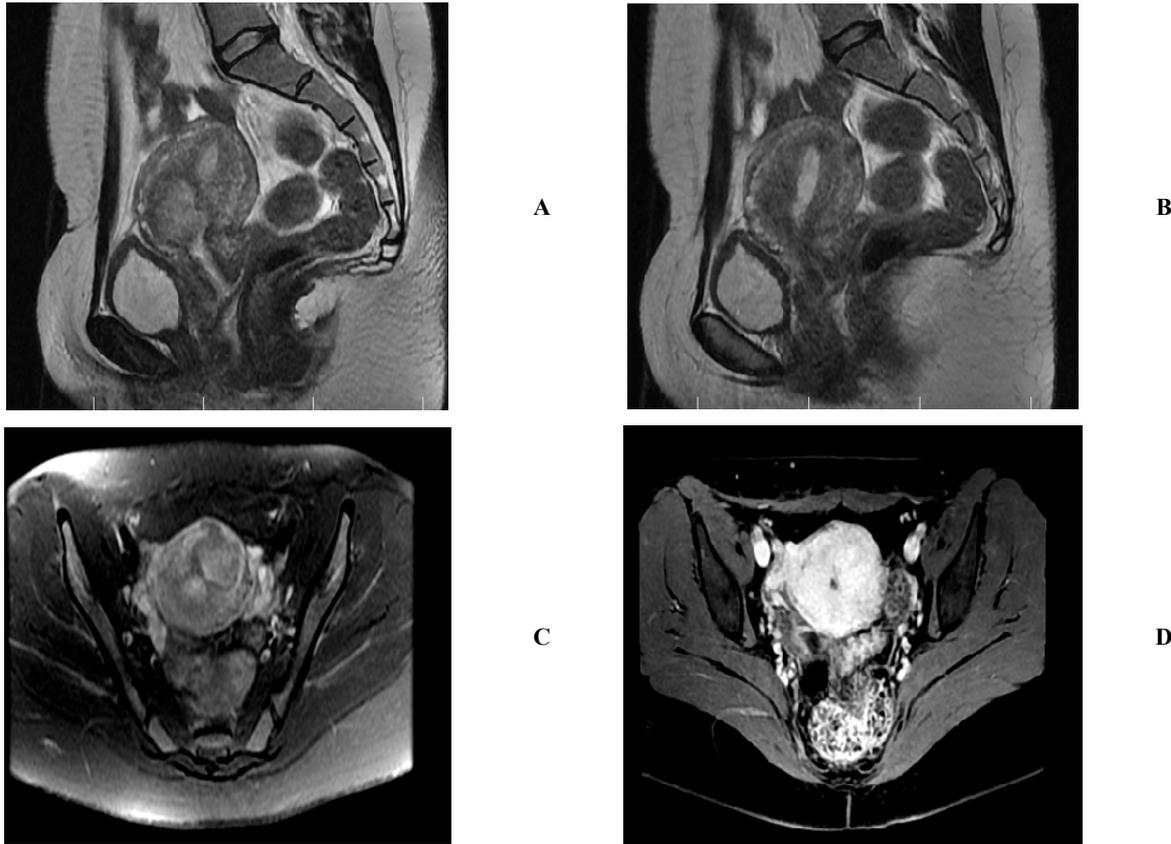


C: Positive immunostaining for CD10 in low grade stromal sarcoma

**Figure 1. Photomicrograph of low-grade stromal sarcoma**

Chest and abdominal computed tomography (CT) were normal. Transvaginal sonography, 3 months after the first hysteroscopy, showed 35×24 mm hyperheterochronic mass in the left anterolateral region of the uterus, extending from the endometrial layer to serosa. Pelvic MRI revealed a 43×40 mm intramural myoma with homogenous enhancement and pressure effect on the uterus's endometrium from the left anterolateral mass (Figure 2).

The patient was a candidate for complete resection of the mass by laparotomy and hysteroscopy. Hysteroscopy revealed endometrial hyperplasia, and two separate endometrial nodules were resected. In laparotomy, a 3 cm intramural myoma in the left anterolateral wall of the uterine body with pressure effect into the uterine cavity in hysteroscopy was excised abdominally. Endometrial curettage just beneath the mass was done.



**Figure 2.** T2 sagittal pelvic images: A iso-signal to high signal mass lesion in lower segment of uterus anterior wall (A) is seen without clear connection to endometrial line and no serosal invasion (B). Axial T1 pre and post contrast fat suppression images show no significant tumoral enhancement, near to myometrial enhancement (C, D).

Low-grade ESS was confirmed on pathologic examination of mass, nodules, and attached endometrium. Taking this report into account, along with a multidisciplinary team decision and consultation with the patient, she underwent a total abdominal hysterectomy and bilateral salpingo-oophorectomy (TAH+BSO) after fertility preservation. Four oocytes were retrieved, and 3 embryos were reserved.

There was no residual tumor in the hysterectomy specimen, and IHC was positive for ER and PR.

After 27 months of follow-up, she has not developed any problems in physical examination or chest and abdominopelvic CT scans.

## Discussion

Endometrial stromal sarcoma is a rare condition, constituting nearly 0.2% of all reproductive tract malignancies, its incidence is about 2 per million women; in comparison, endometrial cancer affects 700 per million women (6).

In 2014, endometrial stromal tumors were classified by the WHO into four categories, namely: endometrial stromal nodule (ESN), low-grade endometrial stromal sarcoma (LGESS), high-grade endometrial stromal

sarcoma (HGESS) and undifferentiated uterine sarcomas (UUS) (2).

ESS is an uncommon tumor, mainly affecting perimenopausal women, with a median age at presentation ranging between 45 and 55 years. However, younger women and adolescents are not spared, as was the case in our 20 years old patient. Reviewing the medical literature, five cases of young patients with low-grade ESS were reported (Table 1).

Usual presenting features for the condition include abnormal uterine bleeding, postmenopausal bleeding, abdominal pain, and pelvic mass (7). In our reported case, the patient had abnormal uterine bleeding about 6 months after vaginal delivery. Her ultrasound indicated myoma or placental polyps, which, at that stage, were the most likely diagnoses in this patient.

The most common site of occurrence of ESS is the uterine corpus though extra-uterine sites are not spared. Compared to the other two types of uterine sarcoma, women with low-grade endometrial stromal sarcoma (LGESS) often present at a younger age, as exemplified by our 20 years old patient. Local spread of ESS to the vagina, fallopian tubes, ovaries, bladder, and ureters is possible, and the disease can metastasize distally to extra-pelvic sites such as lungs and heart. At

presentation, the extra-uterine disease is observed in up to 30% of cases with low-grade ESS (8).

**Table 1. Five cases of low-grade ESS in young patients reported in the literature**

Author	Age	Parity	Size	Diagnosis	Treatment	Follow-up	Recurrence
Arab <i>et al.</i> (the present case)	20 y/o	1	5 cm	Pathologic examination after myomectomy (hysteroscopic + laparotomy)	TAH* and BSO* + letrozole	27 months	No recurrence at 27-months
Chang <i>et al.</i> , (11)	27 y/o	0	4.6 cm	Pathologic examination after hysterectomy	TAH and salpingectomy+ medroxyprogesterone acetate + tamoxifen	11 months	No recurrence at 11-months
Dong <i>et al.</i> , (12)	25 y/o	0	5 cm	Final pathologic and IHC examination	Fertility-preserving local mass resection and endocrine therapy	18 months	No recurrence after 3 years
GÜZİN <i>et al.</i> , (13)	21 y/o	1	9 cm	Final pathologic examination after myomectomy	TAH and BSO, pelvic lymph-node dissection, an appendectomy, omentum biopsy	3 years.	No recurrence after 3 years
Koskas <i>et al.</i> , (10)	34 y/o	0	15 mm	Pathologic examination after hysteroscopic resection	Hysteroscopic resection. Resection of recurrent masses + Letrozole (refused hysterectomy by patient)	6 months	Severe peritoneal recurrence after pregnancy

\* TAH, total abdominal hysterectomy; BSO, bilateral salpingo-oophorectomy; ICSI, Intracytoplasmic sperm injection;

Its resemblance to conditions such as leiomyoma makes the clinical and radiological diagnosis of ESS challenging. As was the case in our patient, ultrasound lacks specificity and can suggest an incorrect diagnosis of more prevalent conditions such as adenomyosis or leiomyoma. Even histologically, especially in the presence of myxoid, epithelioid, and fibrous changes, ESS may be incorrectly identified as other conditions such as leiomyoma, uterine leiomyosarcoma (LMS), or other sarcomas, making the pathological identification a challenge too. Three experienced pathologists reviewed the specimens in the present case to achieve an accurate diagnosis. IHC study can prove useful in diagnosing ESS. While muscle-specific Actin, SMA, and Desmin are expressed in ESS, leiomyoma, and LMS, diffuse CD10 immunoreactivity is a valuable marker of ESS. CD10 is a sensitive marker of normal endometrial stroma and ESS; our patient had positive CD10 on immunohistochemistry. CD10 is negative in most cases of cellular leiomyoma (8).

Total abdominal hysterectomy with bilateral salpingo-oophorectomy is the surgery recommended as the final resort for LGESS treatment. The presence of steroid receptors in ESS raises concerns about the stimulation of residual disease by estrogen secreted by the ovaries; thus, the recommendation for oophorectomy (9). Koskas *et al.*, (10) in their case study, concluded that definitive surgery should be considered as soon as ESS is diagnosed and should not be postponed. This was based on their finding of a dramatic progression of LGESS in the form of severe peritoneal recurrence in their 34-year-old patient who became pregnant after her ESS was conservatively managed. Thus, in our patient, given the positive

estrogen and progesterone receptors, complete abdominal hysterectomy with bilateral salpingo-oophorectomy was suggested and performed, despite her young age.

Hormonal therapy may have a role in treating cases with advanced disease (6).

A third to a half of LGESS patients develop a recurrence (1). Recurrences are reported to have occurred between 3 months and 23 years after treatment, with a median of about 3 years (10).

Although controversial, many factors are suggested to be prognostically significant for the condition, including age, menopausal status, tumor size and stage, mitotic count, histologic grade, and involvement of surgical margins by tumor (7).

For LGESS with recurrent disease, medroxy-progesterone, gonadotropin-releasing hormone (GnRH) analogue, and aromatase inhibitors are the suggested hormonal therapies. Compared to other types of uterine sarcoma, LGESS is reported to have a higher survival rate (1).

With a 5-year survival rate of 60%, low-grade ESS has a prognosis similar to that of endometrial carcinomas; the rate drops to 25% for patients with high-grade lesions. Behtash *et al.*, (7). Reported a five-year survival rate of 96% for LGESS patients in their study.

Given its rarity, only a few case reports and retrospective analyses with small sample sizes have been reported for ESS (7).

## Conclusion

Endometrial stromal sarcoma is a rare uterine malignancy of mesenchymal origin. Although rare, diagnosis of endometrial stromal tumors should be considered whenever a patient comes with a large uterine polyp and fibroid-like symptoms.

## Acknowledgments

None.

## Conflict of Interest

The authors declare no conflict of interest.

## References

1. Verma M, Pandey L. Low grade endometrial stromal sarcoma: a case report. *Int J Curr Adv Res.* 2020;9(1):21040-2.
2. Mangla G, Diwaker P, Gogoi P. Endometrial Stromal Sarcoma: Case Series with Emphasis on Gross Features. *Indian J Surg Oncol.* 2019;10(1):91-4. [DOI:10.1007/s13193-018-0847-2] [PMID] [PMCID]
3. Ashrafganjoei T, Bahman A, Noei Teymoordash S, Aminimoghaddam S, Ebrahimi A, Talayeh M. The Incidence of Ovarian Involvement in Endometrioid Endometrial Adenocarcinoma: A Retrospective Analysis. *J Obstet Gynecol Cancer Res.* 2021;6(3):105-9. [DOI:10.30699/jogcr.6.3.105]
4. Angelos D, Anastasios L, Dimosthenis M, Roxani D, Alexis P, Konstantinos D. Endometrial stromal sarcoma presented as endometrial polyp: a rare case. *Gynecol Surg.* 2020;17(1):1-3. [DOI:10.1186/s10397-020-01073-4]
5. Abu Jamea GA, Al Rikabi AC, Akkour K. Primary low-grade endometrial stromal sarcoma arising in the vagina: report of an unusual case and literature review. *J Surg Case Rep.* 2017;2017(11):rjx238. [DOI:10.1093/jscr/rjx238] [PMID] [PMCID]
6. Chu MC, Mor G, Lim C, Zheng W, Parkash V, Schwartz PE. Low-grade endometrial stromal sarcoma: hormonal aspects. *Gynecol Oncol.* 2003;90(1):170-6. [DOI:10.1016/S0090-8258(03)00258-0]
7. Behtash N, Akhavan S, Gilani MM, Mousavi A, Ghaemmaghani F, Mazhari MM. Low grade endometrial stromal sarcoma of uterine: review of 17 cases. *Acta Medica Iranica.* 2011;49(9):619-24.
8. Devi P, Himaja S, Manasa R, Atla B. Uterine low grade endometrial stromal sarcoma: a case report with review of literature. *Int J Res Med Sci.* 2015; (3):2510-3. [DOI:10.18203/2320-6012.ijrms20150665]
9. Pandey P, Dixit A, Tanwar A, Mahajan NC. low grade endometrial stromal sarcoma: report of a rare uterine malignancy (case report). *Iran J Pathol.* 2013;8(1):64-70.
10. Koskas M, Morice P, Yazbeck C, Duvillard P, Walker F, Madelenat P. Conservative management of low-grade endometrial stromal sarcoma followed by pregnancy and severe recurrence. *Anticancer Res.* 2009;29(10):4147-50.
11. Chang Y, Li J, Fan Y. Ovarian function preservation of low grade endometrial stromal sarcoma at stage II: A case report and review of literature. *Res Rep Gynaecol Obstet.* 2018;2(1):5-8.
12. Dong R, Pang Y, Mao H, Yang N, Liu P. Successful pregnancy following conservative management of low-grade endometrial stromal sarcoma: A case report. *Oncol Lett.* 2014;7(4):1039-42. [DOI:10.3892/ol.2014.1858] [PMID] [PMCID]
13. GÜZİN K, YİĞİT A, AfŞAr S, ÖZerden E, DoĞAnyılmaz S, GÜÇLÜEr B. A Young Patient with Low Grade Endometrial Stromal Sarcoma: Case Report. *Clin Obstet Gynecol.* 2010;20(6):407-10.

## How to Cite This Article:

Talayeh M, Arab M, Noei Teymoordash S, Afsharmoghaddam N, Raoufi M, Ghavami B et al. Low-Grade Endometrial Stromal Sarcoma in a Young Woman: A Case Report. *J Obstet Gynecol Cancer Res.* 2022; 7(6):569-73.

## Download citation:

[BibTeX](#) | [RIS](#) | [EndNote](#) | [Medlars](#) | [ProCite](#) | [Reference Manager](#) | [RefWorks](#)