

## Case Report

# Uterine low grade endometrial stromal sarcoma: a case report with review of literature

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### ABSTRACT

Endometrial stromal sarcomas are rare uterine tumours usually seen in perimenopausal females. We report here a case of low grade endometrial stromal sarcoma in a 45 year female with a clinical diagnosis of fibroid uterus and final histological diagnosis of low grade endometrial stromal sarcoma.

**Keywords:** Uterine leiomyoma, Low grade endometrial stromal sarcoma, Uterine sarcoma

### INTRODUCTION

Endometrial stromal tumors of uterus are the second most common mesenchymal tumors of the uterus. Endometrial Stromal Sarcoma (ESS), is a rare neoplasm, constituting to about 0.2% of all uterine malignancies and account for less than 10% of all uterine sarcomas, composed of cells closely resembling normal proliferative endometrial stromal cells.<sup>1</sup>

It may be mistaken for leiomyoma, as identifying it clinically is difficult and is more often diagnosed postoperatively after a histopathological examination. We present a case of low grade endometrial stromal sarcoma, diagnosed clinically as leiomyoma.

### CASE REPORT

A 45 year old post-menopausal female presented with pain abdomen and mass per abdomen since 5 months. Per abdominal findings are a 26 weeks size mass which is firm, tender with smooth well defined borders. She underwent total abdominal hysterectomy with bilateral

salpingo-oophorectomy for bulky uterus with suspicion of fibroid clinically.

On gross examination, the hysterectomy specimen (Figure 1) measured 20x18x12 cm with bilateral adnexae with each ovary measuring 3x2x1 cm with attached 4cm tube. Cut section of uterus revealed an irregular nodular mass of 12x8 cm with gray white, gray brown, yellowish areas and firm to soft consistency (Figure 2). Cut section of both ovaries and tubes were normal.

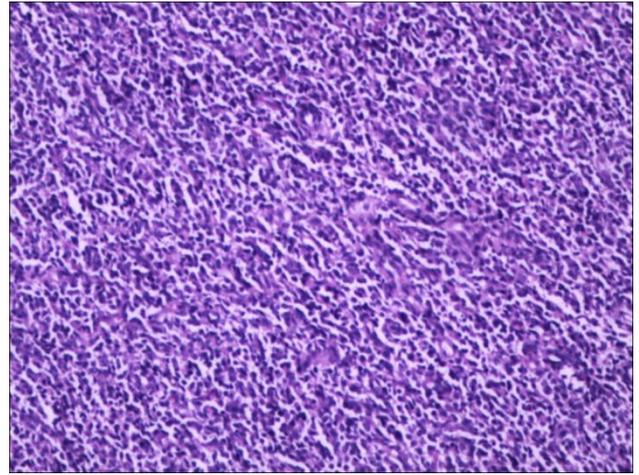
Histologically, multiple sections from the nodular mass show sheets of round to oval cells (Figure 3). Individual tumor cells have scanty cytoplasm, round to ovoid and hyperchromatic nuclei. There are foci of highly cellular areas and foci of necrosis. There are 1-3 mitotic figures per 10 high power fields, occasional multinucleate giant cells seen. Some of the tumour areas show high vascularity with proliferation of small blood vessels and arterioles (Figure 4). There are foci of tumor infiltration into adjacent myometrium (Figure 5).



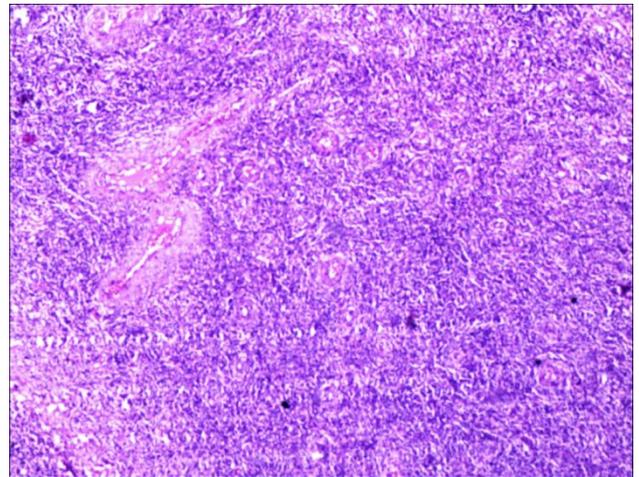
**Figure 1: Gross hysterectomy specimen showing a nodular mass in endometrial cavity.**



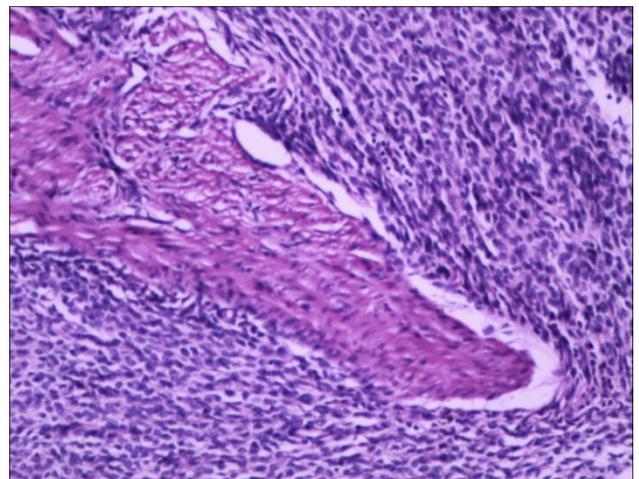
**Figure 2: Cut section of nodular mass showing gray white, gray brown and yellowish areas.**



**Figure 3: Microphotograph showing tumour cells relatively uniform, round to oval nuclei and scanty cytoplasm with less than three mitotic figures per high power field.**



**Figure 4: Microphotograph showing proliferation of small blood vessels and arterioles.**



**Figure 5: Microphotograph showing tumour invading the myometrium.**

## DISCUSSION

Endometrial Stromal Sarcoma (ESS) is a very rare malignant tumor, the annual incidence of ESS is 1-2 per million women and is characterized by sheets of cells with endometrial stromal cell differentiation.<sup>2</sup> Endometrial stromal tumours are divided into three types on the basis of mitotic activity, vascular invasion. The endometrial stromal nodule is a lesion confined to the uterus, with pushing margins, less than three mitosis per ten high power fields and absence of lymphatic or vascular spread. The disease usually has good prognosis with no reported recurrences or deaths following surgical removal of the tumor.<sup>3</sup> Low grade endometrial stromal sarcoma is defined as infiltrative stromal tumor, show less than ten mitoses per ten high power fields, frequently extending into and growing within large vascular spaces. It has a five year survival rate of 100%. High grade endometrial stromal sarcoma is characterized by more than ten mitoses per ten high power fields. It is a highly lethal neoplasm with aggressive clinical course and a five year survival rate of 55%.<sup>4</sup> Differentiation of endometrial stromal nodule and Endometrial Stromal Sarcoma (ESS) is done by establishing the well-circumscribed nature of the lesion in endometrial stromal nodule in contrast to irregular, infiltrating margins in ESS as observed in our case.

Low-Grade Endometrial Stromal Sarcoma (LGESS), also known as endolymphatic stromal myosis, has an infiltrating margin and commonly exhibits extensive worm-like lymphatic and venous vessel invasion. The first description was published by Norris and Taylor in 1966.<sup>5</sup>

ESS is an occasional diagnosis in a patient presenting as leiomyoma uterus. An early diagnosis is essential because patient survival is directly related to tumor stage. Uterine sarcoma most often affect postmenopausal women. Women with LGESS are younger with a median age of 45 and 55 years. The presenting features are similar to uterine leiomyoma; abnormal vaginal bleeding, pelvic mass or abdominal pain and pressure caused by an enlarging pelvic mass, and some patients may be asymptomatic.

ESS is malignant and can spread to the vagina, fallopian tubes, ovaries, bladder and ureters. Distant metastasis to lung, heart and to other sites has also been reported. Up to 30% of women with low grade ESS have an extra uterine disease at presentation.<sup>6</sup>

Preoperative diagnosis is often difficult and around 75% are diagnosed as benign leiomyoma as in our case.

Endometrial curettage and histopathological examination do not help due to similarity with normal endometrium. Besides, the tumor has a propensity to grow through intramural sections of the uterus instead of intracavitary part. This prevents accurate histopathological diagnosis

preoperatively. Ultrasound and magnetic resonance imaging are inconclusive and the diagnosis is usually uterine leiomyoma or pelvic mass.

ESS may be confused with leiomyoma, uterine leiomyosarcoma (LMS), or other sarcomas, especially histologically, when associated with myxoid, epithelioid, and fibrous changes. In addition, the immunohistochemical profile of ESS may have similarities with leiomyoma and LMS, with expression of muscle-specific actin, SMA and desmin. Diffuse CD10 immunoreactivity has proven to be a useful positive predictive marker for endometrial stromal sarcoma. CD10 is a sensitive and diagnostically useful immunohistochemical marker of normal endometrial stroma and of endometrial stromal neoplasms.<sup>7</sup> Most cellular leiomyomas are completely negative for CD10.

LGESS have a more protracted clinical course and have <10 MF/10 HPF. Recurrence occurs late and local recurrence is more (50% cases). Prolonged survival as well as cure is common despite the development of recurrent or metastatic disease. Total abdominal hysterectomy with bilateral salpingo-oophorectomy is optimum initial therapy and radiotherapy is recommended for inadequately excised or locally recurrent pelvic disease.

## CONCLUSION

Endometrial stromal sarcoma is a rare uterine malignancy of mesenchymal origin. Although rare, a clinical diagnosis of endometrial stromal tumours should be kept in mind, especially when a patient presents with a bulky uterus and symptoms similar to leiomyoma.

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## REFERENCES

1. M. Usha, Aarathi R. Rau, B. K. Sujani, T. Uravashi. Low grade endometrial stromal sarcoma presenting as a cervical polyp in a young female: a rare case report. Clin Cancer Invest J. 2014;3(3):257-9.
2. Puliath G, Nair MK. Endometrial stromal sarcoma: a review of the literature. Indian J Med Paediatr Oncol. 2012;33:1-6.
3. Amrith N. Pandya, Arpita Nisha, Hemali Tailor. A case of malignant low grade endometrial stromal sarcoma and review of the literature. Natl J Community Med. 2011;2(1):152-4.
4. Vaidya KA, Pawar JG, Byathanal AS. Low grade endometrial stromal sarcoma: a case report. Case Study Case Rep. 2012;2(2):18-22.
5. Vincent Landreat, Nicolas Paillocher, Laurent Catala. Low-grade endometrial stromal sarcoma of the uterus: review of 10 cases. Anticancer Res. 2008;28:2869-74.

6. Jain R, Batra S, Ahmad A, Elahi AA, Gupta M, Saith P. Low grade endometrial stromal sarcoma: a case report. *Iran J Med Sci.* 2015;40(1):81-4.
7. Jassal CD, Patnaik BL, Divya A, Prasad S. Low-grade endometrial stromal sarcoma in young age: a

clinicopathological report. *J Obstet Gynaecol India.* 2012;62(1):73-5.

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