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CASE REPORT

# Endometrial Stromal Sarcoma: Case Series with Emphasis on Gross Features

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## Introduction

Endometrial stromal sarcomas (ESS) constitute around 10% of uterine sarcomas, but only 0.2% of all uterine malignancies. ESS usually present in the peri-menopausal age group, as compared to other uterine malignancies which are more common in postmenopausal women [1]. ESS are often clinically misdiagnosed as benign entities like uterine leiomyomas and are usually diagnosed postoperatively, after a histopathological examination of a hysterectomy or polypectomy specimen [2]. This calls for a repeat surgery for definitive management.

ESS are indolent tumours, but can often present with late recurrence and metastasis [3]. Differentiating endometrial stromal nodule (ESN) from ESS is of utmost importance from the management point of view and is done on the basis of myometrial and/or vascular invasion. The recognition of gross features that could lead to a suspicion of an endometrial stromal tumour versus a leiomyoma is of vital importance, so that extensive sampling can be performed on initial evaluation to rule out a malignancy [4].

We are presenting a case series of three cases of ESS, reported within a month period, all clinically misdiagnosed as uterine leiomyomas.

## Results

### Clinical Features

All three cases were in the age group of 30–35 years and presented to the Gynaecology OPD with chief complaint of abnormal uterine bleeding. Clinico-radiologically, these cases were diagnosed as fibroid and underwent hysterectomy, sparing bilateral adnexa. Table 1 shows salient clinical features of these cases.

### Gross

On initial evaluation, pale grey-white tumour was seen distorting the endometrial cavity in the first two cases. The third case showed a grey-white intramural tumour.

On re-evaluation, first case showed a yellowish, fleshy, polypoid tumour growing into the endometrial cavity (Fig. 1a). The uterine wall was thickened due to myometrial permeation by nodules and cords of tumour (Fig. 1b). Multiple discrete yellow nodules were also seen in the cervix (Fig. 1c). Second case showed a relatively well circumscribed yellow, polypoid tumour growing into the endometrial cavity (Fig. 1d). The cut surface was bulging and fleshy unlike the whorled, firm appearance seen in a leiomyoma (Fig. 1d, e). Cervix was grossly unremarkable. In the third case, uterus and cervix were in multiple pieces, with yellowish tumour permeating into the myometrium (Fig. 1f). No areas of necrosis/ haemorrhage were grossly identified in any of the cases.

### Microscopy

On microscopic examination, case 1 showed tongues and nodules of tumour cells invading into the myometrium (Fig. 2a). Tumour showed a plexiform capillary network (Fig. 2b), an occasional foci of hyalinization (Fig. 2c) and xanthomatous change (Fig. 2d). Tumour cells resembled the normal endometrial stroma, were round—oval, bland, with scant cytoplasm, and a small round nucleus (Fig. 2e). Nodules of tumour cells were also seen in the cervix (Fig. 2f). No areas of necrosis were identified in the tumour, mitosis being < 1/10HPF. Case 2 showed similar microscopic findings (Fig. 2g). A focus of lympho-vascular

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**Table 1** Salient clinical features

| Serial number | Age | Signs and symptoms | Clinical and radiological diagnosis | Size on gross | Surgery performed |
|---|---|---|---|---|---|
| 1. | 34 years | Abnormal uterine bleeding | Submucosal fibroid | 2 lesions, measuring 9 × 8 × 5 cm, and 2 × 1 × 1 cm | Total abdominal hysterectomy |
| 2. | 35 years | Abnormal uterine bleeding | Submucosal fibroid | 11.5 × 9.5 × 9 cm | Total abdominal hysterectomy |
| 3. | 35 years | Abnormal uterine bleeding | Intramural fibroid | 6 × 5.5 × 5 cm | Non descent vaginal hysterectomy |

invasion was also identified in the second case (Fig. 2h). Case 3 showed similar findings (Fig. 2i). The cervix was free of tumour.

The tumour cells showed cytoplasmic positivity for CD10 (Fig. 2j), and nuclear positivity for oestrogen receptor (ER) (Fig. 2k) and progesterone receptor (PR) (Fig. 2l). The tumour cells were negative for smooth muscle actin.

## Discussion

Endometrial sarcomas are a rare group of tumours, usually presenting in peri-menopausal women. Patients with low-grade endometrial stromal sarcoma (LGESS) commonly present at an earlier age than high-grade endometrial stromal sarcoma (HGESS) and undifferentiated uterine sarcoma (UUS), with a median age ranging from 45 to 55 years. They can occasionally arise in younger women and adolescents [2]. In the present series, all three women presented at a younger age, mean age being 34.6 years. Usual clinical presentation of ESS is abnormal uterine bleeding. Patients can also present with pelvic pain and dysmenorrhoea [1]. The uterine corpus is the most common site, although tumours can also present at extra-uterine sites like ovaries, pelvis and abdominal cavity [5, 6]. All three patients in the current series presented with abnormal uterine bleeding and had tumours of the uterine corpus. Clinical recognition of these tumours is difficult, and they are often misdiagnosed as leiomyoma [7]. Ultrasound is not a reliable investigation and can lead to an incorrect diagnosis of adenomyosis or uterine leiomyoma [1]. In this study, all the three cases were clinically and sonographically mislabelled as leiomyomas. Total abdominal hysterectomy and bilateral salpingo-oophorectomy with/without adjuvant therapy is the mainstay of treatment of endometrial stromal sarcomas [8]. In the present series, all three cases were diagnosed on histopathological examination of hysterectomy specimens, sparing bilateral adnexa. Hence, a

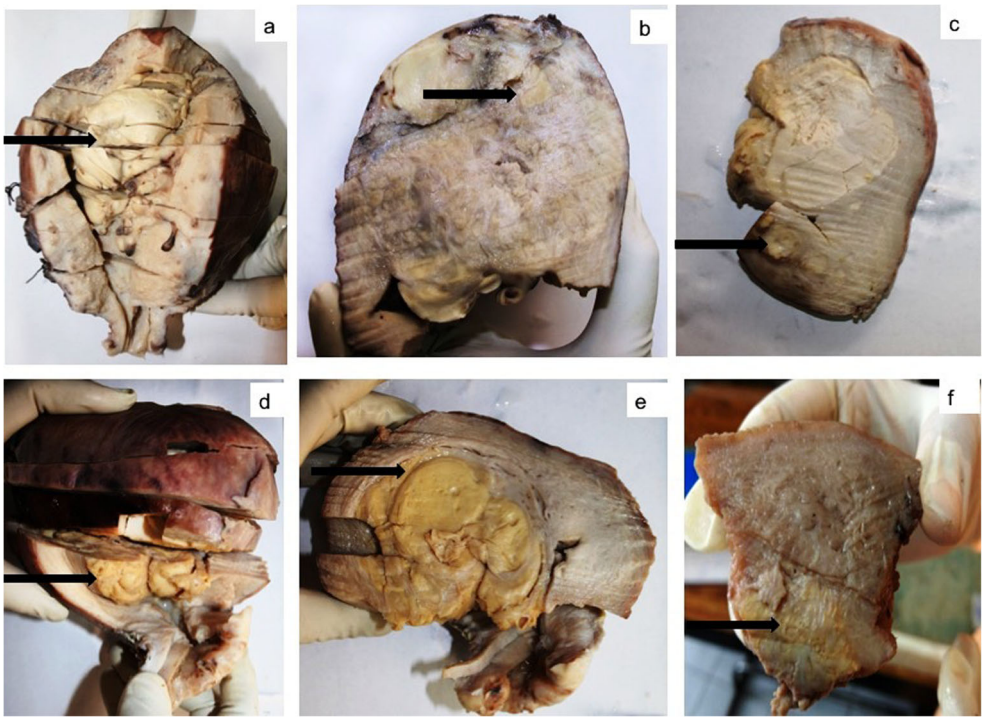


**Fig. 1** Gross examination. **a** Case 1: fleshy yellow polypoid tumour filling the endometrial cavity. **b** Yellowish areas also identified within the myometrium. **c** Discrete yellow tumour nodules in the cervix. **d** Case 2: yellow, polypoid tumour filling the endometrial cavity. **e** Relatively well circumscribed tumour margin. **f** Case 3: yellow tumour areas seen in the myometrium

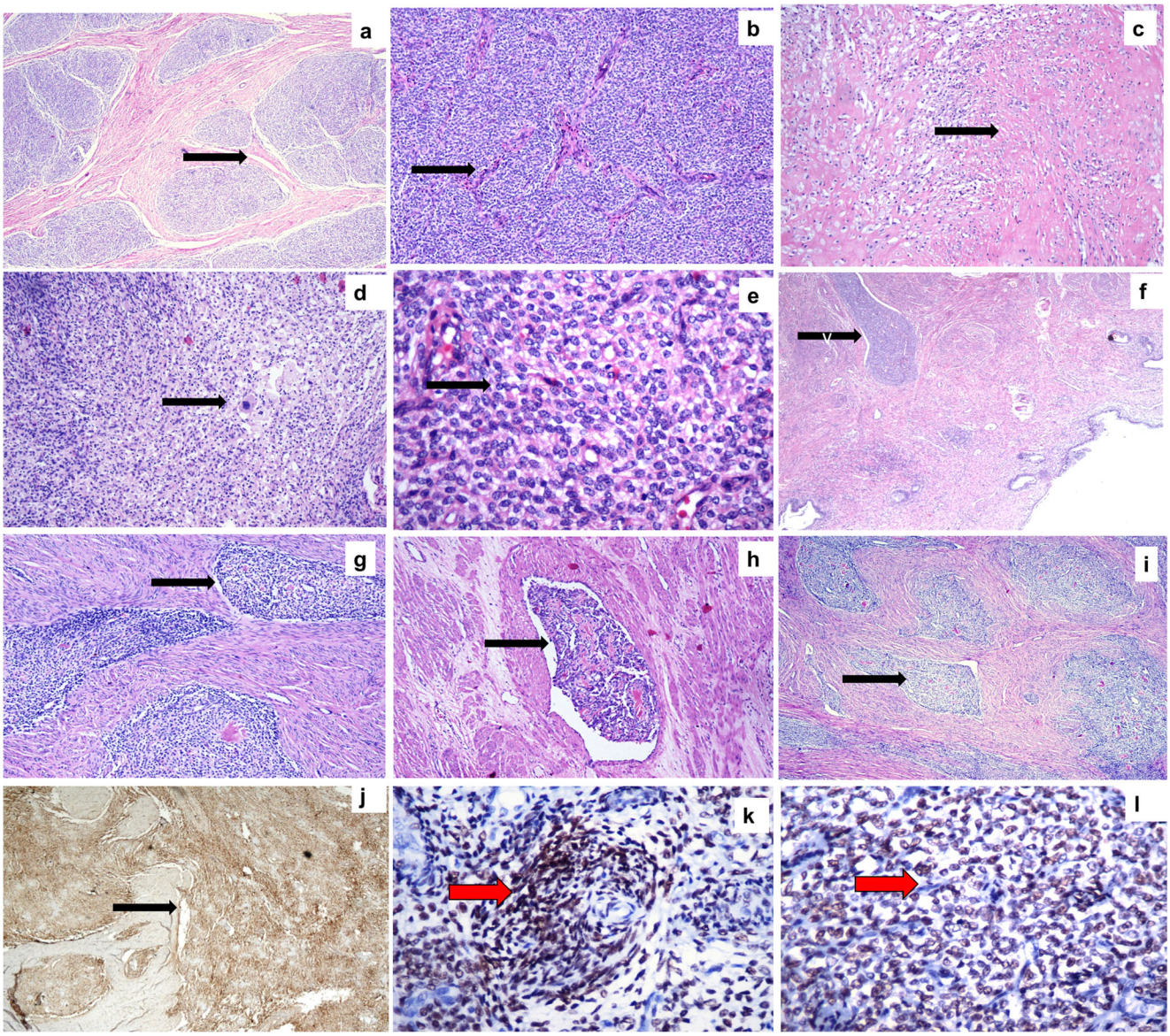


**Fig. 2** Microscopic examination, H&E stain. **a** Case 1: nodules and tongues of tumour infiltrating into the myometrium (× 40). **b** Plexiform capillary network seen in the tumour (× 100). **c** Foci of hyalinization (× 100). **d** Foci of xanthomatous change (× 100). **e** Tumour cells resembling the normal endometrial stroma (× 400). **f** Tumour nodules in the cervix (× 40). **g** Case 2: nodules and tongues of tumour infiltrating into the myometrium (× 100). **h** Foci of lympho-vascular invasion (× 100). **i** Case 3: nodules of tumour cells infiltrating into the myometrium (× 40). **j** Immunohistochemistry—tumour cells showing diffuse cytoplasmic CD10 positivity (× 40X). **k** Tumour cells showing nuclear positivity for ER (× 400). **l** Tumour cells showing mild to moderate nuclear positivity for PR (× 400)

repeat surgery was advised to rule out involvement of the extra-uterine structures.

Endometrial stromal tumours were traditionally divided into endometrial stromal nodule (ESN), low-grade endometrial stromal sarcomas (LGESS) and high-grade endometrial stromal sarcomas (HGESS) [9]. The 2003 WHO Classification eliminated the term "high-grade endometrial stromal sarcoma". The term endometrial stromal sarcomas (ESS) was used for tumours showing histological resemblance to proliferative endometrial stroma. Pleomorphic tumours, earlier classified as HGESS, showing no histological resemblance to normal endometrial stromal cells, were designated as undifferentiated endometrial sarcomas. However, 2014 WHO classification has reintroduced the term HGESS in view of the recent molecular findings. It classified endometrial stromal tumours 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). Presently, the term HGESS is reserved for stromal sarcomas having a round cell morphology, characteristic immunophenotype, and harbouring a distinct translocation t(10;17) (q22; p13) [4].

Grossly, ESN is a well circumscribed tumour, which may be intramural or submucosal filling the endometrial cavity, similar to a leiomyoma. The cut surface of ESN is soft, fleshy, bulging and yellow-tan in colour, in contrast to the firm-white whorled cut surface of a leiomyoma. LGESS generally have an ill-defined border showing myometrial permeation; however, some tumours may appear relatively well circumscribed. HGESS and USS usually present as fleshy polypoid masses, with extensive areas of haemorrhage and necrosis [4]. Careful gross examination is essential to distinguish leiomyomas, from endometrial stromal tumours. Suspicion of endometrial stromal tumours at the time of grossing, alarms the pathologist to take appropriate sections to rule out myometrial/vascular invasion. However, in the present series, all the three specimens were initially grossed as leiomyoma. They were later re-evaluated and re-grossed based on a suspicion of endometrial stromal tumour on microscopy. This was due to clinical bias, and overlooking of the yellow–tan areas of the tumour, possibly due to under-fixation at the time of initial grossing. This leads to a delay in diagnosis of the tumour, due to the time taken for repeat evaluation and processing of the specimens. This indicates that the yellow-tan cut surface of endometrial stromal tumours is better appreciated in well-fixed specimens, and it is important to ensure that the hysterectomy specimens are adequately fixed at the time of grossing.

Microscopically, ESN and LGESS consist of sheets of cells resembling normal proliferative endometrial stroma, showing minimal cytological atypia. Myometrial and/or lympho-vascular invasion are the sole criteria for differentiating LGESS from ESN. This requires extensive gross sampling of all endometrial stromal tumours. LGESS shows numerous nodules and tongues of tumour cells infiltrating the myometrium. HGESS and UUS consist of pleomorphic cells, showing no histological resemblance to the normal endometrial stroma. The tumour cells show extensive permeation of the myometrium, with large areas of necrosis and haemorrhage [4]. In the present series, all the three cases showed bland cells resembling the normal endometrial stroma infiltrating the myometrium and were classified as LGESS. LGESS are positive for CD10, ER and PR [4]. In our study, all the three cases were positive for CD10, ER and PR.

## Conclusion

Through this case series, we wish to highlight the gross features of ESS. We conclude that hysterectomy specimens must be adequately fixed at the time of initial grossing, as the characteristic yellow areas of endometrial stromal tumours may be overlooked in an underfixed specimen. Tumours with a clinical diagnosis of leiomyomas should be carefully evaluated on gross examination, so as to not miss a possibility of endometrial stromal tumours at the time of initial grossing, allowing for a prompt diagnosis.



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