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Endometrial Stromal Sarcoma Mimicking as Uterine Leiomyoma

Short Running title: Endometrial stromal sarcoma

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Abstract: ***Endometrial stromal sarcoma (ESS) is a rare malignant tumour of endometrium, occurring in age group of 40-50years. We report a case of ESS in a 29-year-old woman with a clinical diagnosis of multiple leiomyoma. Total Laparoscopic hysterectomy was done under general anaesthesia. Histopathology and Immunohistochemistry helped to arrive at a final diagnosis of a low-grade Endometrial stromal sarcoma. As this tumour is rarely encountered, management protocols are questionable. Although rare, ESS should be considered in the differential diagnosis of a rapid enlargement of a uterine Leiomyoma. This case report is aimed at its rarity and difficulties faced in establishing the diagnosis.***

Keywords: Endometrial stromal sarcoma, ESS

1. Introduction

Endometrial stromal sarcoma, also known as Endolymphatic stromal myosis, is a rare neoplasm comprising approximately 0.2% of all uterine malignancies and 10% of all uterine sarcomas.[1] The tumours are composed of cells resembling normal endometrial stromal cells in proliferative phase. It affects younger women and mean age is 42-58years.[2]The pathogenesis of ESS is unknown, but exposure to Tamoxifen, unopposed estrogens and conditions such as PCOD are implicated.[3]

In the latest WHO classification, Endometrial stromal tumours are divided into three types based on mitotic activity, vascular invasion and prognosis. They are Endometrial stromal nodule (ESN), Low-grade endometrial stromal sarcoma (LGESS) and Undifferentiated Endometrial stromal sarcoma (UES). ESN and LGESS fall in the lower end of the spectrum of this group of tumours. At the other end of spectrum is UES, which is a very high grade endometrial stromal sarcoma (HGESS) not resembling the proliferative endometrium.

ESN is confined to uterus with pushing margins, <3 mitotic fig/10hpf and absence of lymphovascular invasion. It has a good prognosis with no reported recurrences or deaths following surgical removal of tumour. LGESS is an infiltrative stromal tumour with <10 mitotic fig/10hpf frequently extending into and growing within the vascular spaces. It has a 5-year survival rate of 100%. High-grade ESSis characterised by >10mitotic fig/10hpf. It is lethal with a clinical course and 5-year survival rate of 55%.

2. Case Report

A 29-year-old woman P_1L_1 presented with menorrhagia for 8 months and dysmenorrhoea for 5months. A Transvaginal USG was done which revealed 3 myomas in the posterior aspect of uterus measuring 6x5cm, 4x3cm and 2x2cm. Cervix and ovaries were normal. So, an impression of Multiple Leiomyoma of uterus was given.

Routine blood and radiological investigations were normal. Vitals were stable and systemic examination was unremarkable. Laparoscopic myomectomy was attempted but the masses had no clear demarcation capsule (fig-1). So, Total laparoscopic hysterectomy was performed followed by bilateral salpingectomy. The operated uterus and bilateral fallopian tubes were sent for histopathological examination. Grossly, uterus, cervix measured 13x10x4.5cm with multiple friable and nodular growth obliterating endometrial cavity, largest measuring 4.5x4.5cm and another measuring 3x3cm (fig-2). Two tubes measured 4x3x1cm each appeared normal.

Microscopically, the tumour had an infiltrative margin and permeated the myometrium in irregular tongues (fig-3). More than 50% myometrial invasion was observed (fig-4). The tumour cells were small, uniform with round to oval nuclei and scant cytoplasm arranged in sheets and whorls around arterioles (fig-5).Mitotic activity was <3/10hpf and there was lack of haemorrhage or necrosis. One of the parametrial margins was invaded by tumour cells. There were multiple foci of lymphovascular invasion. Cervix, fallopian tube and vaginal cuff margin were free from

tumour. An impression of Low grade Endometrial stromal sarcoma was made from the histopathological findings.

Immunohistochemistry

The tumour cells showed strong diffuse positivity for CD10 which is a cell-surface neutral endopeptidase (fig-6). This was helpful in distinguishing it from Leiomyoma. At the same time ESS cells showed positivity for both ER and PR receptors.

3. Discussion

The usual clinical presentation of Endometrial stromal sarcoma is abnormal vaginal bleeding. An asymptomatic ESS occurs in 25% individuals.[4] Abdominal pain and uterine enlargement may occur. Our patient came with the chief complaint of menorrhagia and dysmenorrhoea. The main tumour mass is almost always intramyometrial, rarely ESS can be present at an extrauterine site most commonly ovary.

MRI is an useful pre-operative diagnostic tool showing bands of low-signal intensity within the areas of myometrial invasion.[5] Ultrasound is not reliable as it can lead to wrong diagnosis of Adenomyosis or Uterine leiomyoma. Another feature is extension of tumour into vessels, fallopian tube, ligaments and ovaries.

ESS has been divided into low and high-grade according to cell morphology and mitotic count. Boardman CH et al[6] defined low-grade ESS by cell uniformity, <3/10hpf mitoses and lack of haemorrhage or necrosis.However, there are controversies regarding the various criteria for distinction. Several authors have concluded that two separate entities exist- A low-grade sarcoma and an Undifferentiated or high grade sarcoma.

The differential diagnosis of ESS includes several soft-tissue neoplasms demonstrating arborizing vasculature like highly cellular leiomyoma, cellular endometrial polyp, low-grade mullerianadenosarcoma, leiomyosarcoma and adenomyosis.[7] Extragenital ESS maybe confused with gastrointestinal stromal tumours, hemangiopericytoma, lymphangiomatosis, or mesenchymal cystic hamartoma of lung.

ESS like other sarcomas is most effectively treated by surgery. The effect of adjuvant therapy is not yet proven. Hormone therapy is effective due to rich expression of estrogen and progesterone receptors in the tumour. Recurrences develops in about one-third of patients with ESS and are usually limited to pelvis and lower genital tract. Distant metastasis to lungs may occur after several years.[8]

Prognosis mainly depends upon the stage of disease at the time of diagnosis. Prognostic factors are age, race, tumour size and stage, nuclear atypia, mitoses, tumour necrosis, lymphovascular invasion, expression of hormone receptors etc. Older patients (more than 50years), black race, advanced stage, lack of primary surgery, nodal metastasis, >5 mitoses per 10 hpf, CD10 negative or low expression and lack of estrogen and progesterone receptors were independent risk factors for poor survival in a multivariate analysis. ESS has a better life expectancy than other sarcomas.[9]

CD10 was initially thought to be a specific marker of Endometrial stromal tumours. It is expressed in other smooth-muscle tumours of uterus like highly cellular leiomyoma and leiomyosarcoma. Other useful antibodies are h-caldesmon, histone deacetylase-8 and smooth-muscle myosin.[10] The neoplastic endometrial stromal cells are typically immunoreative for vimentin, muscle-specific actin and smooth-muscle actin. In our case, tumourcells showed positivity for CD 10, ER and PR hormone receptors.

4. Conclusion

Endometrial stromal sarcoma is a rare uterine tumour. It has a large variation in pathological features and hence difficult to diagnose.Extensive sampling is very important in distinguishing endometrial stromal neoplasia from an endometrial stromal sarcoma. A panel of antibodies should be used for confirmation. Immunohistochemistry should always be correlated with the histomorphological appearance of the tumour cells and interpreted accordingly.

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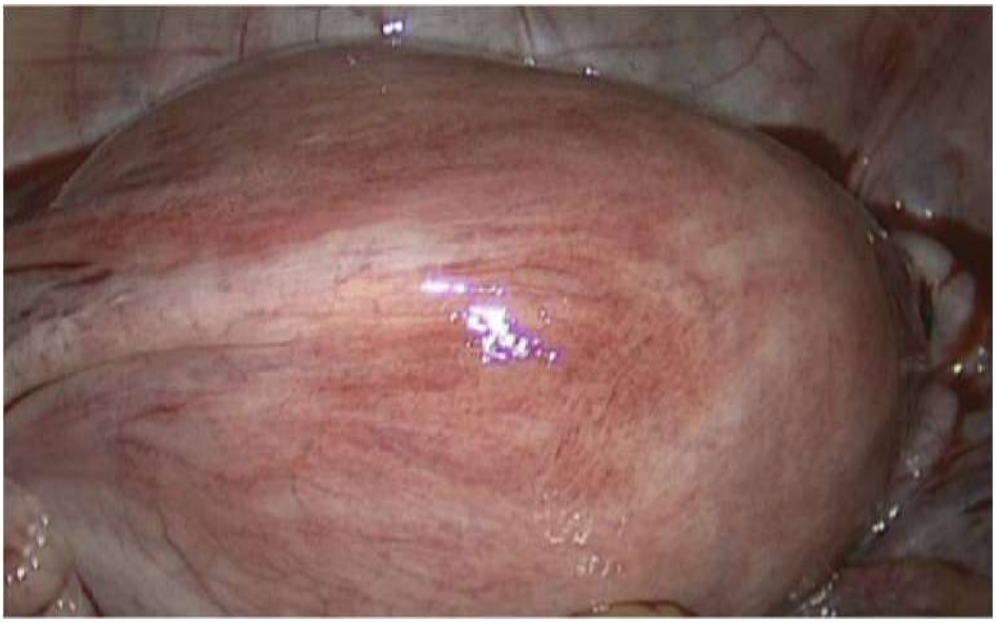

Figure 1: Laparoscopic view of uterine mass

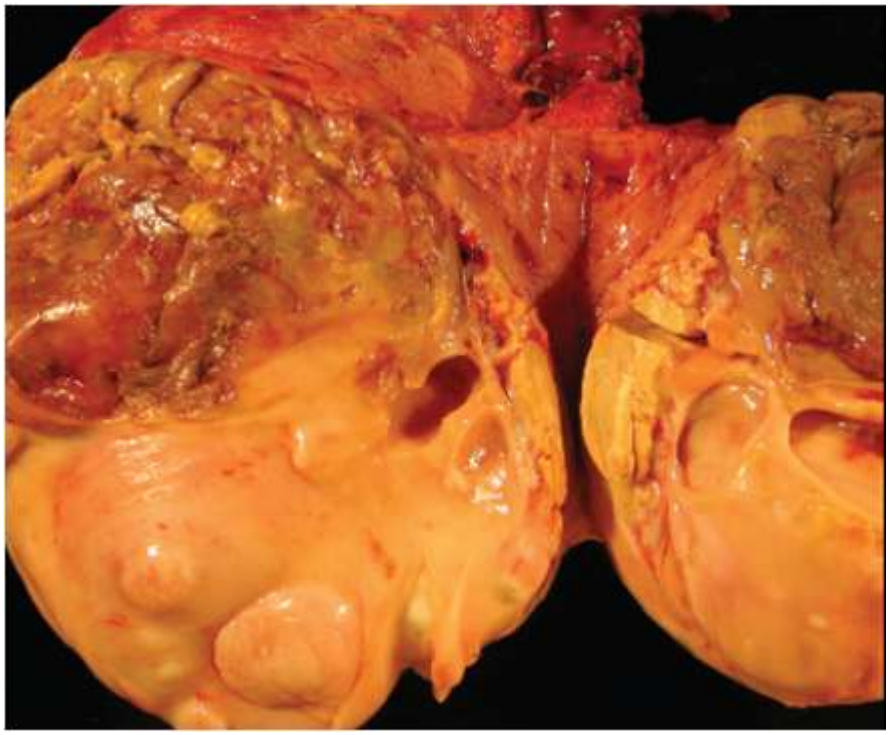

Figure 2: Uterine cavity showing multiple nodular growths

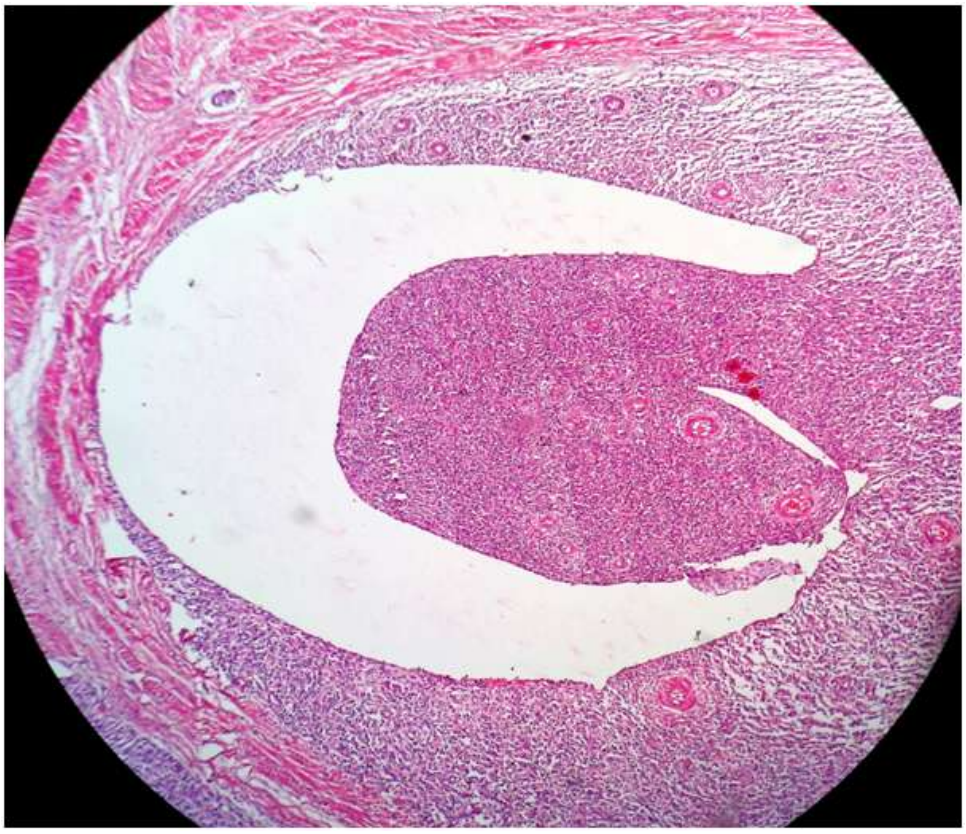

Figure 3: Tongue like protrusion of tumour cells into myometrium (HP, H & E, 100X)

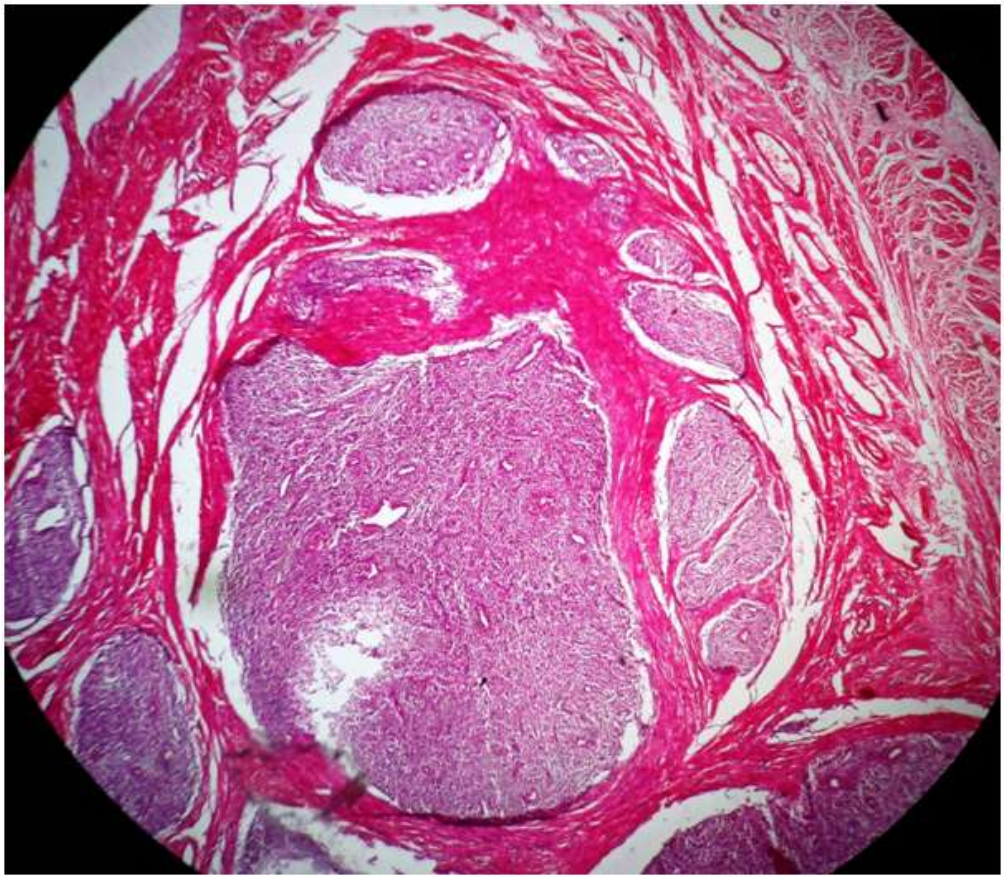

Figure 4: More than 50 % myometrial invasion (HP, H & E, 100 X)

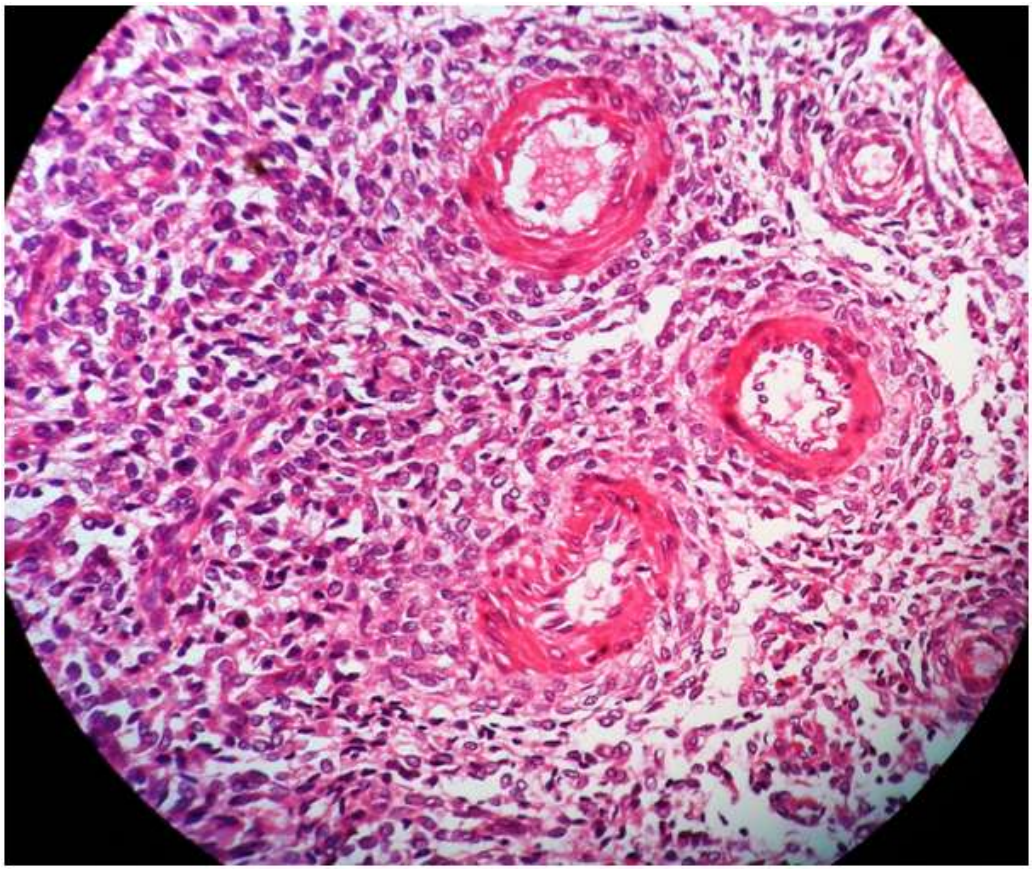

Figure 5: Tumour cells in sheets & whorls around arterioles (HP, H & E, 400 X)

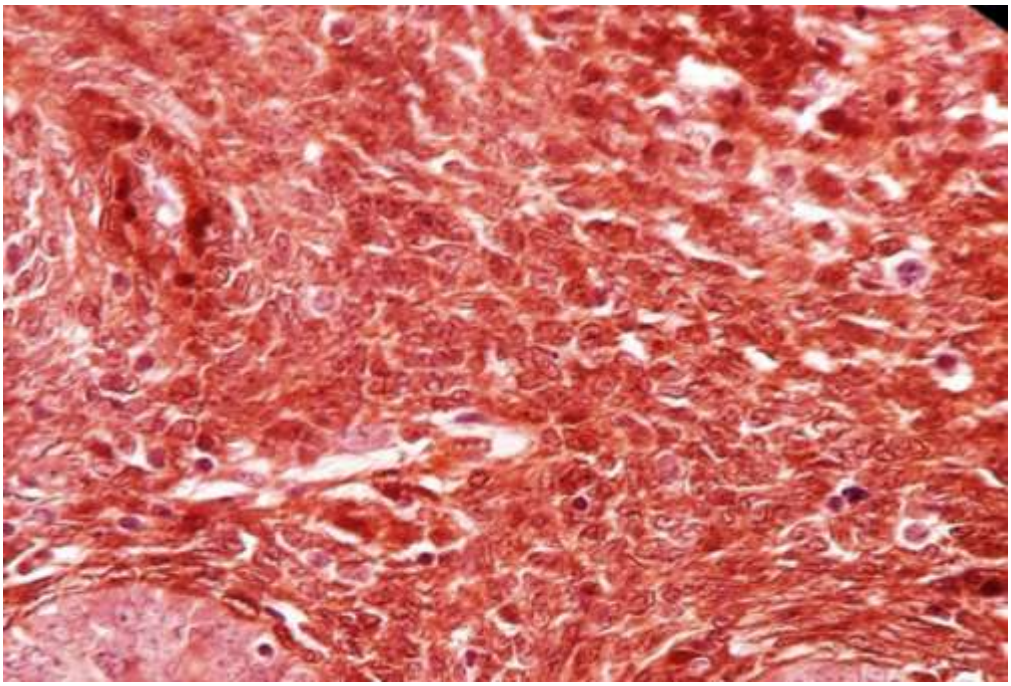

Figure 6: CD 10 strong & diffuse positivity (HP, IHC, 400 X)