

Endometrial Stromal sarcoma masquerading as uterine fibroid**Dr. Richa Jindal, Dr. Sripriya Krishnan, Dr. Priya Jain, Dr. Sheetal Malpani, Dr. Molly Joseph**
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Abstract: Endometrial stromal sarcoma is a rare malignant tumour in the uterine cavity occurring in the perimenopausal period mostly. Generally it is commonly misdiagnosed as uterine fibroid, and hysterectomy is needed for final diagnosis. We report a case of 46 year old female with low grade endometrial stromal sarcoma presenting as uterine fibroid with abnormal bleeding. Histopathology confirmed its diagnosis. Patient is under close follow up after surgery and received radiotherapy. As the tumour is rarely encountered, the management protocols are still questionable. Although rare, endometrial stromal sarcoma should be considered in differential diagnosis of a rapidly growing fibroid.**Keywords:** Endometrial stromal sarcoma, endometrial stromal nodule, leiomyoma, immuno histochemistry.

INTRODUCTION:

Endometrial stromal sarcoma is a rare malignant tumour in the uterine cavity with an incidence of 0.2% of all uterine malignancies. It occurs in the age group of 40 to 50 years [1]. They account for less than 10% of the uterine mesenchymal neoplasm and 10 to 25% of affected women are premenopausal [1]. The preoperative diagnosis is difficult, as it mimics fibroid in the clinical presentation. Hysterectomy is needed for definitive diagnosis in most cases. Prognosis also depends on the stage of the disease.

CASE REPORT:

A 46 years old female, P3L3 presented in gynaecology O.P.D with complaints of heavy bleeding for 5 days associated with passage of clots and pain abdomen on and off. She had a previous history of bleeding per vaginum for 1 month which was controlled with antifibrinolytics. There was no history of hormonal mediations. Her haemoglobin levels were 8 gm/dl.

On per vaginum examination she had a bulky uterus. Ultrasound revealed her uterus to be anteverted and a bulky fibroid in the posterior wall. Average uterine thickness was 8.1mm. Pap smear was normal. Endometrial biopsy was done which showed secretory endometrium with features of disintegration.

Total abdominal hysterectomy with bilateral salphiniophorectomy was planned and the specimen was sent to our department. Grossly, uterus was bulky measuring 10x9.5x9 cms. External surface of uterus and cervix were unremarkable. Cut section showed an irregular thickened soft grey white mass measuring 3x2.5cms within the myometrium, pushing the endometrial cavity. Bilateral adnexa were normal.

Microscopic examination showed a tumour composed of nodules and sheets of cells resembling endometrial stromal cells infiltrating the myometrium. The cells were round to ovoid with bland nuclear chromatin and moderate amount of eosinophilic cytoplasm. At places, they were arranged concentrically around spiral arterioles. Invasion into lymphovascular spaces was identified. Mitotic activity is inconspicuous (<10/10 HPF). No necrosis was present.

DISCUSSION:

Endometrial stromal sarcoma is a rare tumour. About 20 % are low grade and 6 % are high grade tumour. The age of presentation varies from 42 to 53 years [1]. They are generally divided into 3 types depending on the mitotic activity. They are divided into endometrial stromal nodule (ESN), low grade endometrial stromal sarcoma (LGESS) and high grade or undifferentiated endometrial stromal sarcoma.

Puliyah *et al.*; reported a case of ESS in a 30 years old female where ultrasound and Doppler were suggestive of fibroid uterus [2]. Women with LGESS are younger in age varying from 45 to 57 years and do not have risk factor for endometrial carcinoma [5]. The surgical staging is the most significant prognostic marker for recurrence and survival in LGESS. They tend to grow slowly and over many years.

Differential diagnosis of ESS includes several soft tissue neoplasms demonstrating arborizing vasculature, cellular leiomyoma, ESN and cellular intravenous leiomyomatosis. The microscopic appearance of LGESS and ESN appear similar. ESS differs from ESN, since in former the margins are infiltrative and distinct growth as worm like cords are seen whereas, well differentiated margins are seen in

ESN. Hence extensive sampling of the tumour margins and detecting vascular invasion is very important. LGESS is differentiated from high grade ESS on the basis of mitotic activity and necrosis. Mitotic activity is low (<10/10HPf) in ESS and in high grade ESS, the mitotic activity is high (>10/10HPf).

About 30% of the women with low grade ESS are diagnosed as uterine leiomyoma [3]. Cellular leiomyomas are composed of cells with spindle shaped nuclei and fascicular growth pattern. At times it is

difficult to differentiate from leiomyomas and ESS. Hence to differentiate, immunohistochemistry is used which shows immunoreactivity with CD10, inhibit, smooth muscle actin and desmin [4].The immunohistochemical markers such as h-caldesmon, beta catenin, and CD10 solves the problem as CD 10 and beta catenin staining is positive in ESS but not in leiomyoma. Surgery is the final treatment for ESS. It is often misdiagnosed as fibroid. Hormone therapy with progesterone, tamoxifen, GnRH is used for LGESS and recurrent disease.

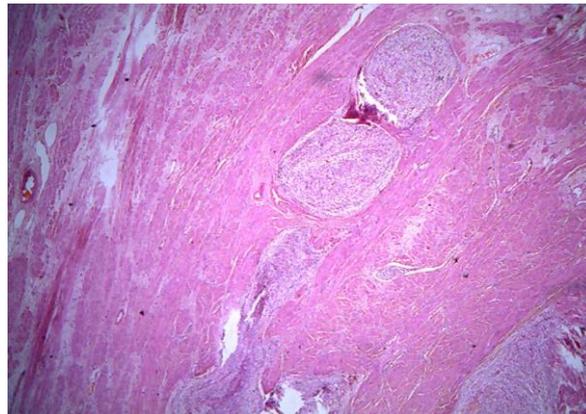


Fig-1: Sheets and nests of endometrial stromal cells invading into the myometrium

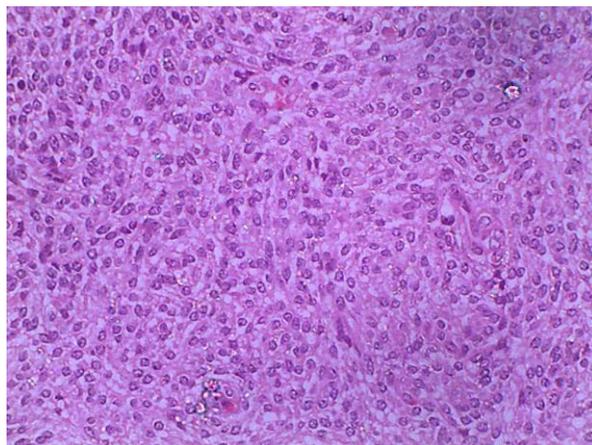


Fig-2: round to ovoid cells with bland nuclear chromatin and moderate amount of eosinophilic cytoplasm

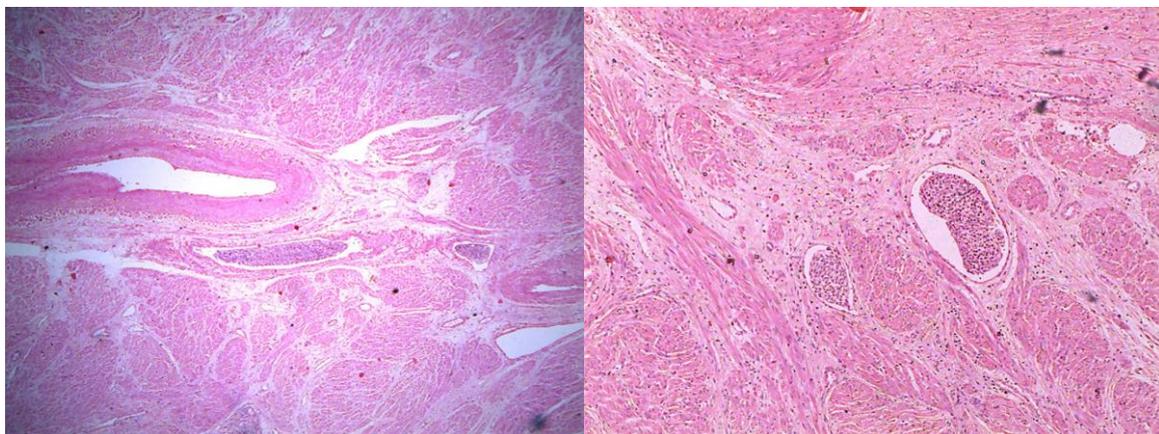


Fig-3 and 4: lymphovascular invasion

CONCLUSION:

Endometrial stromal sarcoma is a rare malignant tumor, usually presenting like a uterine fibroid. Final definitive diagnosis is received only after histopathology of uterus. Hence by reporting our case. We would like to stress on the necessity for a high degree suspicion even in women of younger age. This really would help in patient's treatment and survival.

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