University Journal of Surgery and Surgical Specialities

ISSN 2455-2860 2021, Vol. 7(4)

# Endometrial Stromal Sarcoma - A Rare Case Report

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**ABSTRACT**

Endometrial Stromal Sarcoma (ESS) is a rare malignant tumour of the Endometrium occurring in the age group of 40 – 50 years. A proper preoperative diagnosis is difficult and in most cases the diagnosis is confirmed after hysterectomy. This is a case of low grade endometrial stromal sarcoma presenting as post-menopausal bleeding for which fractional curettage was done. HPE report showed high grade serous endometrial carcinoma. CT report showed heterogeneous enhancing lesion in the endometrial cavity. As surgery was main treatment of endometrial stromal sarcoma, the staging laparotomy was done. HPE revealed Low grade endometrial stromal sarcoma with right pelvic node involvement.

**KEY WORDS**

Endometrial Stromal Sarcoma, Uterine Sarcoma, Staging Laparotomy

**INTRODUCTION**

Cancers arising from mesodermal structures like muscles and connective tissue are called sarcomas. Sarcomas of the uterus are uncommon, and may arise from connective tissue, smooth muscle or the endometrial stroma. Uterine sarcoma is a rare form of malignancy, occurring in 2–5% of all patients with uterine malignancy, with an incidence of approximately one to two cases per 100,000 women in the general population. Endometrial stromal sarcomas (ESSs) are very rare malignant tumors that make up approximately 10% of all uterine sarcomas but only around 0.2% of all uterine malignancies[(1)].

**CASE REPORT**

A 55-year-oldpost-menopausal woman para 4, live 4, all full term normal delivery, sterilized, last child birth 27 years back. She was attained menopause 15 years back. She was a known case of DM for 5 years on treatment and known case of HT recently diagnosed on treatment. She was presented with bleeding per vagina for 3 months and complaints of abdominal pain and white discharge per vagina.

O/E patient was conscious, oriented, afebrile, no pallor, not dyspnoeic, not tachypnoeic, cardio vascular and respiratory system was normal. Per abdomen was soft. In bimanual examination cervix pointing downwards, uterus anteverted, bulky, fornices free. Per rectal examination parametrium and rectal mucosa free. Transvaginal ultrasound showed uterus appears bulky with thickened Endometrium. Her other investigations were normal. Fractional curettage was done. HPE report showed high grade serous endometrial carcinoma. CT report revealed heterogeneous enhancing lesion in the endometrial cavity (Figure 1).

For which staging laparotomy was done. A Total abdominal hysterectomy with bilateral salphingo oophorectomy was performed through a vertical midline incision. The findings were as follows; 1. bulky Uterus 14 weeks in size (Figure 2) 2. Both adnexa normal 3. Right pelvic node size of about 3×2 cm 4. No ascites and peritoneal nodule 5. Liver and Spleen were normal.

Saline wash for cytology was done. Proceeded to Extra Fascial Type I Pan Hysterectomy bilateral pelvic node dissection with InfracolicOmentectomy was done. Peritoneal biopsy was taken from bilateral paracolic gutter and bilateral sub diaphragm.

Microscopically no malignant cells identified in the peritoneal washing. Section from uterus showed characteristically uniform round to oval cells with eosinophilic cytoplasm and vesicular nuclei suggestive of low grade endometrial stromal sarcoma infiltrating the myometrium was noted (Figure 3). Myometrial thickness was 2.3 cm. tumour invasion was 2 cm. a section from both fallopian tubes and both ovaries were normal. The cervix showed chronic cervicitis. One right pelvic node revealed metastatic tumour deposits. Left pelvic nodes were normal. Right paracolic gutter sub diaphragmatic peritoneal biopsy and omental biopsy showed only fibrofatty tissue. This case was confirmed to be a low grade endometrial stromal sarcoma stage 3. We could be planned the treatment accordingly. Post-operative period was uneventful.

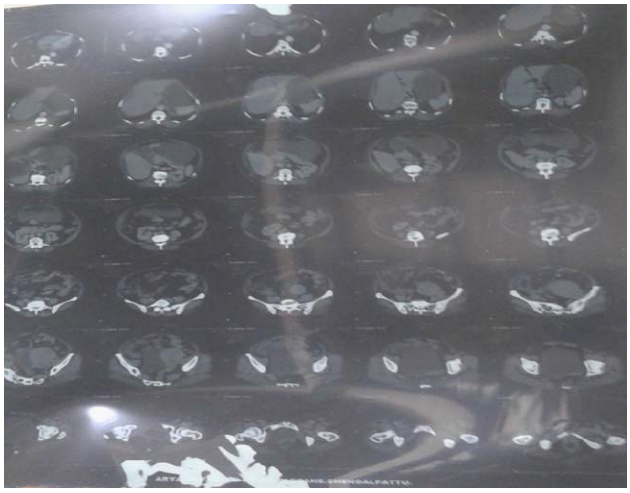

**Figure 1. CT Abdomen showsheterogeneous enhancing lesion in the endometrial cavity**

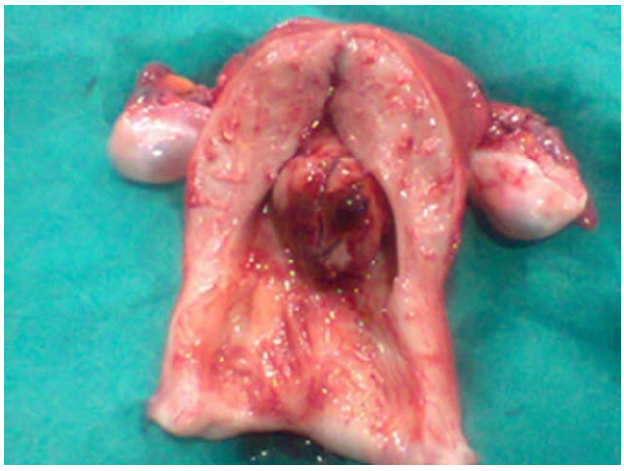

**Figure 2. Macroscopic appearance of uterus, endometrial growth obliterates the endometrial cavity**

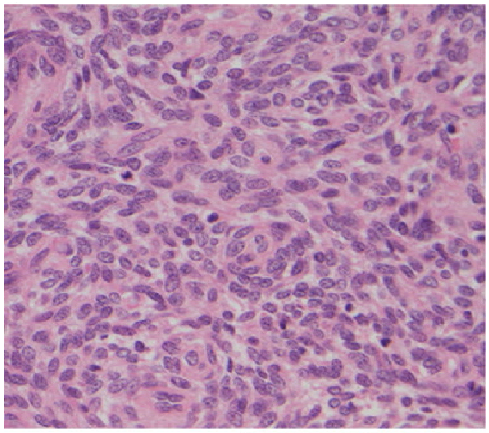

**Figure 3. Microscopic appearance of uterus shows characteristically uniform round to oval cells with eosinophilic cytoplasm and vesicular nuclei.**

## DISCUSSION

ESSs are very rare malignant tumors. Based on tumor margin status and cytological features, the WHO has classified endometrial stromal tumor into benign endometrial stromal nodule (ESN) and endometrial stromal sarcoma. ESN does not infiltrate the myometrium. They are well circumscribed, with a pushing margin. ESSs infiltrate the myometrium and are characterized by proliferation of uniform small cells closely resembling those of endometrial stroma in the proliferative stage [(2)]. ESS can be divided into low-grade and high-grade tumors according to cell morphology and mitotic count. Low-grade ESS usually occur in the young population (mean age 39 years), contrary to the high-grade ESS (mean age 61 years). Low-grade ESSs have less-frequent mitosis (<3 per 10 high-power fields) and they do not show hemorrhage and necrosis [(3)]. Some authors have regarded high grade endometrial stromal sarcoma (HGESS) as an undifferentiated sarcoma[(4)].

Most ESSs involve the endometrium, and uterine curettage usually leads to diagnosis [(2)]. The main differential diagnosis of low-grade ESS includes ESN, cellular leiomyoma and cellular intravenous leiomyomatosis[(5)]. The microscopic appearance of ESS and ESN are identical. Infiltrative margins and distinctive growth as worm-like cords are noted in low-grade ESS, whereas the margins are well demarcated in ESN. Hence, extensive sampling of tumor margins and detecting vascular invasion are extremely important in distinguishing between the two. Cellular leiomyomas are composed of cells with spindle-shaped nuclei with a fascicular growth pattern, thick muscular-walled vessels, cleft-like spaces and showing focal merging with the adjacent myometrium[(6)]. In low-grade ESS, cords of tumor cells infiltrate between smooth muscle and within lymphatic spaces. The neoplastic stromal cells resemble those of the

proliferative endometrium, are monotonous in appearance and have a uniform size and shape. The nuclei are round to ovoid, with fine chromatin, and small nucleoli is seen. A small amount of cytoplasm is present and cell borders are indistinct. Mitotic activity is low (<10/10 high-power fields)[5].About a third to a half of the low-grade ESS have extrauterine spread at the time of diagnosis. When there is a difficulty in diagnosing between ESS and cellular leiomyoma, immunoreactivity with antibodies to CD10 and smooth muscle actin and desmin are used[7].

Surgery is the final resort for diagnosis and primary treatment of ESS. Preoperative diagnosis is high grade serous endometrial carcinoma. Ultrasound and CT Scan are inconclusive. If the diagnosis is known, the extent of surgery is planned according to the stage of the tumour. The FIGO staging for carcinoma of the corpus uteri has been applied to ESS.

The treatment is total abdominal hysterectomy, bilateral salpingo-oophorectomy and pelvic and periaortic selective lymphadenectomy. Cytologic washings are obtained from the pelvis and abdomen. Hormone therapy with medroxy progesterone, tamoxifen, gonadotropin releasing hormone (GnRH) analogues and aromatase inhibitors are suggested for low-grade ESS stage 3–4 and for recurrent disease[8,9].

RT in the form of brachytherapy with or without pelvic radiation can be used as adjuvant therapy. This will be useful for control of local recurrences but with limited effect on surveillance [10,11]. It is not recommended routinely in FIGO stage-I and stage-II disease. However, radiotherapycanbe considered for advanced or recurrent cases[12,13].

## CONCLUSION

ESS is a rare uterine tumor. Because of the large variation in pathologic characteristics, combined with scarcity of patients, there is insufficient information about an optimal management. Study on prognostic factors is also not satisfactory. Hormone therapy is a new promising adjuvant treatment modality. In post-menopausal woman presenting with bleeding per vagina high suspicious of endometrial carcinoma. Therefore, fractional curettage was done. HPE revealed high grade serous endometrial carcinoma for which staging laparotomy was done. HPE report revealed endometrial stromal sarcoma. This case highlights necessity for proper pre-operative diagnosis for post-menopausal woman with complaints of bleeding per vagina and surgery is the definitive diagnosis and treatment for endometrial stromal sarcoma. Multianalysis from a large group of patients is necessary for predicting prognosis and to define proper treatment of endometrial stromal sarcoma.

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