

## Endometrial stromal sarcoma- A case report

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### Abstract

Endometrial stromal sarcomas are rare malignant tumours of the uterus. Most details about this tumour is obtained from previous case reports. In most of the cases, diagnosis is made only postoperatively. A 29 year old female, came with complaints of heavy menstrual bleeding, lower abdominal pain, mass in lower abdomen for 2 months. Clinical examination and investigations showed bulky irregularly enlarged uterus probably Fibroid Uterus. Patient was planned for Myomectomy. Intraoperatively, uterus was irregularly enlarged with cauliflower like growth of size 7 x 5 cm over fundus suggestive of Carcinoma. Total abdominal hysterectomy with bilateral salphingo oophorectomy with bilateral pelvic lymphnode dissection with infracolic omentectomy was done. Histopathology examination revealed as Low Grade Endometrial Stromal Sarcoma with Negative Lymph Node Spread. Postoperatively, Patient was evaluated for ER sensitivity and started on Tab. Anastrozole.

**Keywords:** Uterine sarcoma, Endometrial stromal sarcoma, Malignant uterine tumour

### Introduction

Endometrial stromal sarcomas constitute around 0.2% of all uterine malignancies. They constitute second most common mesenchymal uterine tumours. It usually involves age group of 40-58 years and most women are premenopausal. World health organisation classifies ESS into 1) Endometrial Stromal Nodule 2) Low Grade ESS 3) High Grade ESS 4) Undifferentiated ESS. It usually presents late with local metastasis. Distant metastasis can occur even 20 years after initial diagnosis. Surgery and hormonal therapy are mainstay of treatment. Prognosis is good in case of early diagnosis.

### Case Report

A 29 year old female was admitted in Department of Obstetrics and Gynaecology, Vinayaka Missions Medical College and Hospital, Karaikal, Pondicherry with complaints of heavy menstrual bleeding, lower abdominal pain, mass in lower abdomen for 2 months. Clinical examination revealed uterus enlarged upto 14-16 weeks size, firm, mobile from side to side, irregular margins with irregular surface. Ultrasonogram of abdomen showed uterus of size 10.3\*8.0\*7.8 cm, intramural fibroid of size 6.7\*6 cm in anterior wall. Patient was planned for Myomectomy. Intraop findings-uterus irregularly enlarged, cauliflower like growth of size 7 x 5 cm seen over fundus, edematous right fallopian tube. Bilateral ovaries were cystic and unhealthy. On suspicion of malignancy, Total Abdominal Hysterectomy with Bilateral Salphingo Oophorectomy with Bilateral Pelvic Lymphnode Dissection with Infracolic Omentectomy was done. Histopathology examination report came as Low Grade Endometrial Stromal Sarcoma with Negative Tumour Deposit. Patient underwent Immunohistochemistry studies and found to be ER positive. Patient was started on Tab. Anastrozole 1 mg OD. Patient is in regular follow up.



Fig. 1: Endometrial Stromal Sarcoma

### Discussion

Endometrial stromal sarcoma (ESS) is second most common Malignant Pure Mesenchymal Uterine Tumor (MPMUT) constituting about 0.2% of all uterine tumours.<sup>1</sup> It arises from submesothelial pluripotent müllerian cells.<sup>2</sup> ESS is classified into i) Endometrial Stromal Nodule (ESN) ii) Low Grade ESS (LGESS) iii) High Grade ESS iv) Undifferentiated ESS (UES).<sup>3</sup> Cytogenetic Abnormality of ESS is recurrent translocation t (7;17)(p15;q21).<sup>4</sup> Low grade tumours are indolent tumours with favorable prognosis.<sup>5</sup> LGESS occurs in age group of 28-67 years and presents as uterine enlargement, pelvic mass, abnormal uterine bleeding.<sup>6</sup> High grade tumours have features intermediate between LGESS and UES.<sup>7</sup> Microscopically, HGESS have round cells with spindle cell fibromyxoid component.<sup>6</sup> Undifferentiated ESS usually present as postmenopausal bleeding. UES are highly aggressive and carry poor prognosis.<sup>8</sup> ESS is common in extrauterine sites also. Among them, ovary is first involved usually 76%, extraovarian sites constitute 24%.<sup>9</sup> Histology of extrauterine extraovarian ESS show multinodular growth

patterns.<sup>10</sup> Imaging studies of ESS are usually nonspecific.<sup>11</sup> Positive labelling for CD10, PR, ER with negativity for CD117,CD34 is confirmative diagnosis.<sup>12</sup> Mainstay of treatment is total abdominal hysterectomy with bilateral salphingo oophorectomy. In case of pregnancy associated with sarcomas, laparoscopy can be done safely.<sup>13</sup> Abdominal surgery can also be done safely in case of pregnancy associated ESS.<sup>14</sup> In case of inoperable, recurrent tumours, hormonal therapy is helpful. It includes medroxyprogesterone acetate, GnRH agonists, aromatase inhibitors.<sup>15</sup> In earlier studies, oral medroxyprogesterone acetate was used.<sup>16</sup> In recent times, combination containing Doxorubicin is used.<sup>17</sup> Ifosfamide is helpful in chemoresistant sarcomas.<sup>18</sup> Tamoxifen, estrogens are contraindicated as they cause stimulative effect on disseminated endometrial stromal cells.<sup>19</sup> Dydrogestrone is hormonally active, non androgenic, synthetic steroid.<sup>20</sup> It lacks estrogenic, androgenic, glucocorticoid, mineralocorticoid properties. Dydrogestrone is preferred to other progestins due to lack of side effects.<sup>21</sup> Long term use of GnRH agonists can cause osteoporosis.<sup>22</sup> Anastrozole is the preferred aromatase inhibitor. They cause muscle pain, nausea, vomiting.<sup>23</sup> There is noncompliance of Letrozole while usage in breast cancer patients.<sup>24</sup> 5 year survival rate of patients with stage I and II is 90%, stage III and IV is 50%. Follow up is done once in 3 months for first year, half yearly for next 4 years, thereafter yearly follow up.<sup>25</sup>

**Conflict of Interest:** None.

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