

Case Report

High grade endometrial stromal sarcoma: a clinician dilemma

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ABSTRACT

Uterine sarcomas are relatively rare tumors of mesodermal origin. ESS occurs primarily in perimenopausal women in 4th and 5th decade of their life; about one third occurs in postmenopausal women. Here in we describe a case of 44 years old patient presented with one month history of foul smelling discharge per vagina and a pelvic mass. Ultrasound and MRI gave possibility of a large anterior wall and fundal fibroid with degeneration versus neoplastic endometrial thickening. The patient underwent exploratory laparotomy with total abdominal hysterectomy with bilateral salpingoophorectomy with pelvic lymphadenectomy. Histopathology showed tumor cells with round to oval nuclei with high mitotic activity, blood vessel proliferation between the tumor cells and extensive lymphovascular invasion. The pathological diagnosis was HG-ESS stage IB. The patient was referred to radiotherapy department for adjuvant therapy. HG-ESS is a rare clinical entity and considered as important differential diagnosis.

Keywords: HG-ESS, Mesenchymal, Uterine sarcoma

INTRODUCTION

Endometrial stromal sarcomas (ESS) account for 7–25% of all uterine mesenchymal tumours and less than 1% of all malignancies arising in the uterus.^{1,2} Uterine sarcomas are relatively rare tumors of mesodermal origin. The incidence of uterine sarcomas is 1.5–1.7/100,000 females, with a slight increase over time.³ ESS is the second most common type of uterine mesenchymal neoplasm after leiomyosarcoma.⁴ There is dispute whether it is an invasion from endometrium or due to metaplasia of myometrial cells. Most ESS originate in the uterus, whereas rare extrauterine cases, presumably arising from endometriosis, are also encountered.⁵ ESS occurs primarily in perimenopausal women in 4th and 5th decade of their life; about one third occurs in postmenopausal women.^{6,7}

In 2014, World Health Organization (WHO) reclassified the endometrial stromal tumors (ESTs) on the basis of

immunohistochemistry and molecular findings into 4 subtypes- endometrial stromal nodule (ESN), low grade ESS (LG-ESS), high grade ESS (HG-ESS) and undifferentiated uterine sarcoma (UUS).⁶ This was based on the finding that at molecular level both LG-ESS and HG-ESS exhibit relatively simple karyotype whereas defined chromosomal rearrangement is lacking in UUS. High grade ESS or undifferentiated endometrial sarcoma is distinguished from low grade ESS by a mitotic rate greater than 10 mitotic figures per 10 high power microscopic fields.⁷ Detection by cytology is extremely difficult because the cells lack sufficient atypia and mimic normal endometrial stromal cells. This possibility should be kept in mind by the cytopathologist to avoid missing the diagnosis.⁵ High grade ESS has a much more aggressive clinical course and poorer prognosis than low grade ESS. Their 5-year disease free survival is about 25%.⁷ Here in, we report a case of High grade-ESS in a woman who presented with foul smelling discharge and a pelvic mass.

CASE REPORT

44 years old patient P2+0 with one month history of foul smelling discharge per vagina. On examination the patient was obese and her vitals were normal. On per abdominal examination there was a mass arising from the pelvis about 12 weeks gestational size with well defined margins and restricted mobility from above downwards. No other organomegaly or ascites was present. On Per speculum, cervix was suspicious looking and bleeding through the os was present. On per vaginal examination, the cervix was slightly irregular to feel and indurated.

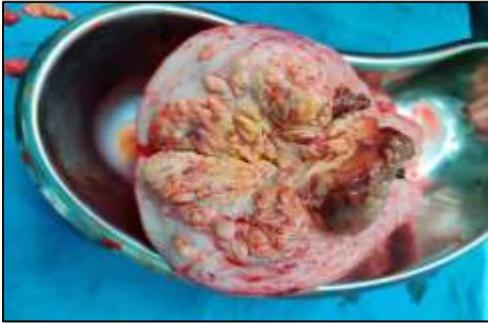


Figure 1:

The uterus was grossly enlarged to 12-14 weeks gestational size and the mass felt per abdomen was felt in continuation with the uterus. Bilateral adnexa were not palpable. On per rectal examination the rectal mucosa was free. Hemoglobin was 10.0 gm/dl and blood group was A positive. Her renal function and liver function tests were normal along with the normal coagulation profile and viral markers. Ultrasound showed a large fibroid with left ovarian cyst of 6×6 cm with a thickened endometrium. MRI showed large well defined heterogeneous mass lesion with few interspersed hyperintensities arising from the anterior wall and the fundus indenting the endometrium with the loss of endometrial -myometrial outline protruding into the endometrial cavity. MRI gave possibility of a large anterior wall and fundal fibroid with degeneration versus neoplastic endometrial thickening.

Keeping in view of the above findings the patient underwent fractional curettage with cervical biopsy. Histopathology showed focal areas of dysplastic cells with endometrial glands showing squamous metaplasia. The patient underwent exploratory laparotomy with total abdominal hysterectomy with bilateral salpingoophorectomy with pelvic lymphadenectomy on 28/2/2020.

At exploratory laparotomy, the uterus was grossly enlarged to 16-18 weeks size. Right ovary was enlarged to 6×6 cm (approximately) smooth walled with no solid areas or papillary excrescences.

On cut section of uterus, macroscopically, there was a large 7×7 cm ?degenerated fibroid over the fundus with an

irregular polypoidal growth arising from the fundus and posterior uterine wall which was yellowish in color. The growth was invading into the outer half of myometrium but not reaching upto serosa. There were dense adhesions of the colon with the posterior uterine wall with endometriotic spots on the uterosacral ligaments and the posterior uterine wall.

Histopathology showed tumor cells arranged in large group of sheets separated by thick fibrous septa. Tumor cells had predominantly monomorphic round to oval nuclei with coarse open up chromatin with clear cytoplasm ill-defined cell borders. Blood vessel proliferation was seen in between the tumor cells. Extensive tumor necrosis, atypical mitotic figures were present along with lymphovascular invasion by tumor cells.

Bilateral adnexa were normal. Bilateral ovaries showed dermoid cysts. Cervix shows chronic nonspecific cervicitis. Lymph nodes showed sinus histiocytosis. The patient was referred to radiotherapy department for adjuvant therapy. The patient received postoperatively radiotherapy and six cycles of Gemcitabine and Docetaxal. She is alive and healthy and on regular follow up by radiotherapy and oncology department.

DISCUSSION

ESS is a rare pathological type of uterine sarcoma. This disease is usually misdiagnosed as uterine leiomyoma or endometrial carcinoma preoperatively due to a lack of characteristic imaging and clinical manifestations. High-grade endometrial stromal sarcoma is rare, constituting of less than 1% of uterine malignancies and less than 10% of uterine sarcomas.⁹ The common presenting symptoms of HG-ESS are abnormal vaginal bleeding, palpable masses, and pelvic pain.¹⁰ The clinical presentations in our patient were foul smelling discharge per vagina and pelvic mass. The average ages at HG-ESS diagnosis reported in the literature range from 40 years to 55 years which is consistent with 44 years age of our patient.

High-grade endometrial stromal sarcoma typically harbors t (10;17) (q22;p13) resulting in YWHAE-NUTM2A/B (previously known as YWHAE- FAM22) genetic fusion.^{10,11} YWHAE-rearranged HG-ESS represents a molecularly and prognostically distinct type of ESS, which is associated with an aggressive natural course.¹¹

The unique clinical behavior and treatment responses of this subset of ESS therefore merit pathologic evaluation to confirm the presence of the t(10;17)(q22;p13) translocation in cases of morphologically suspected HG-ESS.^{11,12} However, in our study the definite molecular genetic feature was unknown. Preoperative imaging is mandatory, because ESS tends to spread to the lungs and peritoneum. In our patient there was no metastasis to the peritoneum and pelvic lymph nodes. Treatment is primarily surgical with addition of adjuvant radiotherapy for local control or chemotherapy for systemic control.⁷

High grade ESS and UES are rarely positive for estrogen receptor and progesterone receptor.⁷ They are not responsive to hormonal treatment unlike low grade ESS. Our patient had total abdominal hysterectomy, bilateral salpingoophorectomy and pelvic lymphadenectomy followed by adjuvant radiotherapy and chemotherapy.

CONCLUSION

We describe a case of HG-ESS stage 1B diagnosed in a 44 years-old woman who presented with foul smelling discharge per vagina and a pelvic mass. Complete staging and surgery along with adjuvant radiotherapy and chemotherapy are associated with significantly improved disease free survival. Role of cytoreductive surgery is now well established in uterine sarcomas. Multicentre retrospective analysis has shown that cytoreductive surgery to less than 2 cm is associated with significantly improved survival in HGEES. Information regarding the natural course, prognostic factors and optimal treatment for HG-ESS is currently limited.

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