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Case Report

Endometrial stromal sarcoma: a rare form of undifferentiated endometrial cancer

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ABSTRACT

Uterine sarcomas are a rare for uterine cancers. They account for 0.2% of uterine cancers. The median age group is 40 to 60 years. They run an indolent course. About 60% women recur after a long period. Metastasis may occur even after 20 years. They can be classified into low grade, high grade and undifferentiated types. Low grade ESS has good prognosis. Surgery with adjuvant hormonal therapy is the mainstay of treatment. Adjuvant radiotherapy and chemotherapy have no role in management. The role of lymphadenectomy is not clear. The first line treatment for recurrence is a repeat surgery. Patients require a long term follow up to detect recurrence. Here we present a case of perimenopausal women presenting as a case of AUB. MRI initially diagnosed it as a case of fibroid. Patient underwent TAH with BSO. Subsequent hispathology and immunohistochemistry revealed it to be Low grade ESS. Although rare, endometrial stromal sarcoma should be considered as a differential diagnosis in perimenopausal and postmenopausal women presenting as AUB.

Keywords: Endometrial sarcoma, Endometrial stromal sarcoma

INTRODUCTION

Uterine sarcomas are a rare type of neoplasm arising from the uterus. Endometrial stromal sarcoma (ESS) is a rare type of sarcoma with an incidence of 0.2%.[1] Median age at diagnosis is 40-60 years. It has an indolent course and about 60% of women recur after a long duration and metastasis can occur even after 20 years. Purpose of this case report is to add to the existing literature which is in form of small studies and case reports.

CASE REPORT

A 50-year-old female para 2 with previous caesarean section presented with case of heavy and frequent menses since 1 year. She is a known case of hypertension on medication. She has no history of diabetes asthma thyroid disorder. On examination vitals stable, mild pallor present, systemic examination was NAD. Per abdomen revealed an uterus of 12 weeks size. Per vaginal examination revealed free fornices. Patient was investigated. MRI pelvis revealed an intramyometrial mass without extra uterine involvement. Ovaries were unremarkable likely features suggested? Fibroid biopsy was done which was inconclusive. After preoperative workup patient was taken up for laparotomy. Patient underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy postoperative period was uneventful. Specimen was sent for HPE. On histopathology it was stage IC endometrial stromal sarcoma low grade (FIGO). Subsequent immunohistochemistry revealed CD10 ER/PR positivity. Patient was reviewed by oncologist. Due to high rate of metastasis and late recurrence patient was started on high

dose medroxyprogesterone acetate. She was counselled regarding regular follow up and risk of recurrence.

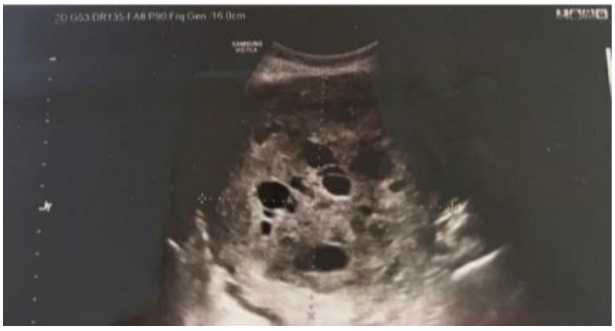

Figure 1: Endometrial stromal sarcoma.

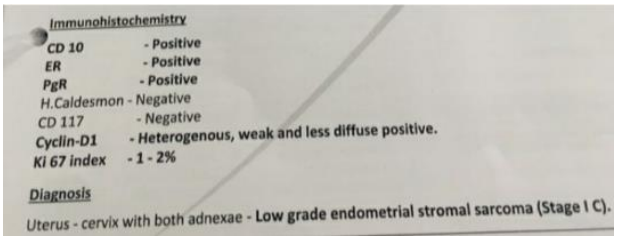

Figure 2: Report.

DISCUSSION

Uterine sarcomas are a heterogeneous group of rare tumors of the uterine musculature and uterine connective tissue. Endometrial stromal cell sarcomas are classified into low grade and high grade tumors. In the current WHO classification published in 2014, LG-ESS are classified as endometrial stromal tumors, along with benign endometrial stromal nodules (ESN), HG-ESS, and undifferentiated uterine saroma (UUS). ESS are staged along with uterine leiomyosarcomas in accordance with the FIGO and TNM classifications. Most frequent presenting symptom is vaginal bleeding. The diagnosis is by endometrial sampling which can be challenging due to resemblance with proliferative endometrial stroma and diagnosis is certain only in 2/3rd of cases. Stage of the disease is most important factor which determines prognosis. [2] Because sarcomas have high risk of metastasis imaging modalities like CT and chest Xray are important. In addition to arising in the uterus, LG-ESS can also develop in extrauterine locations such as the ovaries, the pelvis, or the abdominal cavity, and also the vagina or vulva. Endometriosis is found in 50% of these cases, giving rise to the suspicion that stromal sarcomas can develop out of endometriosis. Rare cases have been reported of the tumor developing in connection with tamoxifen or estrogen administration, as well as after radiotherapy. Obesity, diabetes mellitus, and early menarche are reported to be associated with an increased risk of LG-ESS.[1] Total abdominal hysterectomy with bilateral salpingo-oophorectomy with omental biopsy with peritoneal fluid sampling is the standard treatment. BCOR is a diagnostic immuno-histochemical marker for high grade variety.[3] Immunohistochemically the tumor cells are typically positive for CD 10 vimentin Actins ER and PR.[1] The most common genetic abnormality in low grade ESS is t(7;17) (p15;q21) .patient with low grade early stage ESS have a good prognosis. 5 years survival being 90%. As tumors are positive for ER/PR hormone treatment with progesterone s is effective for prevention of recurrence.[4] In case of recurrences hormone therapy with or without resection is indicated.[4] If tumor is palpable in parametrium radical hysterectomy is required. Nodal involvement is rare. Because of rarity of tumor there is lack of consensus regarding optimal management. Hence treatment should be individualized. Adjuvant chemotherapy has no role.

CONCLUSION

Endometrial stromal sarcoma are a rare group of tumors and a diagnostic dilemma. Many cases are diagnosed only after hysterectomy. These are associated with late recurrences. Hence adequate staging and long term follow up is necessary. Follow up is with clinical assessment and imaging.

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