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ENDOMETRIAL STROMAL SARCOMA WITH INVERSION OF UTERUS - A RARE CASE REPORT

ANGELINE GNANA MALAR S
Department of Obstetrics and Gynaecology, CHRISTIAN MEDICAL COLLEGE

Abstract : Endometrial stromal sarcomas (ESS) are rare malignant tumors of the uterus accounts for 0.2 of all genital tract malignancies. They occur more often in premenopausal women. Most of the information available in literature is based on small series or case reports hence there are limited information regarding the clinical management and final outcome of the disease. Non-puerperal inversion of the uterus is an uncommon presentation of ESS. We report this case of ESS with inversion of uterus in a 23 year old unmarried girl due to its rarity of clinical presentation. She presented with abnormal uterine bleeding associated with abdominal pain and mass descending PV. On examination she was obese, anemic and no masses palpable per abdomen. Local examination showed a large mass lying outside the introitus with foul smelling blood stained vaginal discharge. Biopsy showed a high grade endometrial stromal sarcoma. With the diagnosis of high grade ESS - stage IIB with inversion of uterus, she underwent surgical management - abdomino vaginal approach. Histopathology confirmed the diagnosis of high grade ESS. She received postoperative adjuvant radiotherapy in view of extra-uterine disease. Endometrial stromal sarcomas are aggressive tumors with poor prognosis. The 5 year disease free survival is 25. Total hysterectomy, bilateral salpingooophorectomy with pelvic lymphadenectomy is the optimal treatment in cases of ESS. Ovarian conservation may be possible in young women and with early stage disease and the role of lymphadenectomy is controversial. Adjuvant therapy is useful to prevent recurrence of disease but their role in survival is questionable.

Keyword : Endometrial stromal sarcoma, non-puerperal uterine inversion, uterine sarcomas.

INTRODUCTION:

Endometrial stromal tumours are rare tumours accounting for less than 10% of uterine sarcomas and approximately one percent of uterine malignancies.(1). The World Health Organization classifies endometrial stromal tumors into three categories: endometrial stromal nodule (ESN), endometrial stromal sarcoma (ESS) and undifferentiated endometrial sarcoma (UES). Endometrial stromal nodule is a benign condition with absence of lymphovascular invasion. Endometrial stromal sarcoma formerly termed as low grade ESS and it is distinguished from high grade ESS by a mitotic rate of less than 10MF/10 HPF. High grade ESS or undifferentiated endometrial sarcoma exhibits greater than 10MF/10 HPF and no evidence of stromal differentiation. Endometrial stromal tumors occur primarily in perimenopausal women between 45-50 years of age and one third occurs in postmenopausal women. Non puerperal inversion of uterus is a rare clinical condition with less than 30 case reports were available in the literature. Most common cause of inversion is fibroid uterus but sometimes endometrial stromal sarcomas also have similar effect due to thinning of uterine wall thereby predisposing to uterine inversion.

CASE REPORT

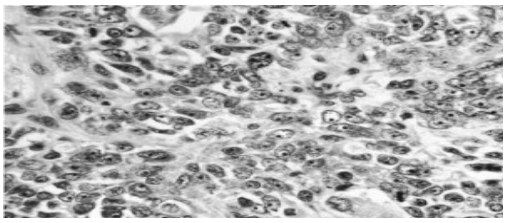
23 year old, unmarried girl presented with continuous bleeding per vaginum with lower abdominal pain for 2 months in November 2011. On examination she was morbidly obese (BMI 40 kg/m2) with no mass palpable per abdomen. She was evaluated elsewhere for the same complaints and MRI was done which was reported as 8x7cm mass noted in the cervix which was protruding through the vagina. She had undergone biopsy from the mass which was reported as decidual tissue with hemorrhagic necrosis with islands of cartilage with few degenerated hyalinized degenerated trophoblastic villi suggestive of products of conception. Same MRI was reported in our Institution as a large well defined mass noted in cervix with extension into the vaginal vault and into the lower uterine segment with myometrial invasion and extension into the parametrium. She was posted for examination under anesthesia (EUA) and repeat biopsy of the mass was done. Biopsy was reported as high grade endometrial stromal sarcoma with cartilaginous elements.

In view of high grade endometrial stromal sarcoma and MRI showing parametrial invasion, it was decided for neoadjuvant chemotherapy. She received 4 cycles of chemotherapy with Doxetaxel and Ifosfamide. Her last cycle of chemotherapy was on 19th June 2012.

ESS - mass lying outside with inversion of uterus

She presented again with excessive bleeding per vaginum and foul smelling vaginal discharge with mass descending per vaginum in the same month June 2012. On examination she was pale, vitals were stable and there was no mass palpable per abdomen. On local examination, a necrotic mass lying outside the introitus measuring 8x7cm with long pedicle with foul smelling blood stained discharge. On per rectal examination, the rectal mucosa was free and there was no parametrial invasion. With the clinical diagnosis of high grade endometrial stromal sarcoma – stage IIB with chronic inversion of uterus, she was posted for surgery. Preoperatively she received 3 units of packed cell transfusion because of low Hb (5.6gms). She underwent Kustner's incision and excision of the mass with body of the uterus vaginally and then proceeded to laparotomy with removal of cervix and peritubal nodule under GA on 24th July 2012. Ovaries are conserved because of younger age of the patient. Intra-operatively, there was 15x10cm polypoidal mass protruding outside the introitus with foul smelling bloodstained vaginal discharge with complete inversion of uterus. At laparotomy, ovaries were found to be elongated but normal and there was a left peritubal nodule of 2x1.5cm. She received antibiotics in the per-operative period. She developed superficial wound gaping which was managed with dressing and then resuturing was done and she was discharged with good general condition. Suture removal was done 10 days after resuturing and wound was healthy.

The surgical pathology specimen was reported as high grade ESS, body of uterus with maximum tumor size of 12cm with 20% tumor necrosis with increased mitotic activity (8-10/HPF). Myometrium with infiltration of tumor. LVSI was not evident. Cervix was free of tumor and peritubal nodule with metastatic tumor deposits. She was referred to radiotherapy for adjuvant radiotherapy in view of extra uterine spread of the disease. She completed radiotherapy in December 2012 after which she developed oliguria and swelling of both lower limbs. She was under evaluation for the same however she expired in 20th December, 2012.



High grade ESS with anaplastic neuclei and mitotic figures

DISCUSSION

Uterine sarcomas are rare tumors of mesodermal origin. They constitute 2-6% of uterine malignancies. There are three histological variants of uterine sarcomas – carcinosarcoma, leiomyosarcoma and endometrial stromal sarcoma (ESS) out of which ESS constitutes 15% and carcinosarcoma and leiomyosarcoma each constitute approximately 40%. ESS occurs primarily in perimenopausal women between ages 45-50 years and one third occurs in postmenopausal women. But in our case, ESS occurs in reproductive age group which is a rare presentation. ESS is hormone sensitive tumors and hyper-estrogenic state could act as a growth stimulus.(2). Because of rarity of these tumors there are limited reports are available in the literature in terms of clinical management and final outcome of these disease. Endometrial stromal tumors were historically characterized as either low-grade or high-grade neoplasm, and this classification is used in older literature. However, high-grade endometrial stromal tumors are more commonly referred to as undifferentiated endometrial sarcomas (UES) reflecting their composition of anaplastic cells with little or no evidence of endometrial stromal differentiation. Most of the uterine inversions are puerperal and non-puerperal inversions are uncommon but this condition is associated with a polypoidal mass. Leiomyomas (71.6%), sarcomas (13.6%), and carcinomas (6.8%) have all been found present with this condition. No etiology is identified in 8% of affected patients.(3). Approximately 30 cases of non-puerperal uterine inversion due to sarcomas have been reported in the literature. One of the mechanisms is that contraction of the uterine musculature due to tumor prolapsing into the vagina combined with tumor-related weakening of the uterine wall results in inversion. Nulliparity may also cause a reduction in thickness of the uterine wall which matches with our case.

The usual clinical presentation of ESS is abnormal uterine bleeding and pelvic pain or abdominal pain. Our patient had similar complaints. The amount of bleeding ranges from spotting to menorrhagia and sometimes associated with foul smelling vaginal discharge. Rarely the tumor may prolapse through the cervical os into the vagina and even outside the introitus presenting as inversion of uterus as in our case report. Although the main tumor mass is almost always intramyometrial, most ESS involve the endometrium and uterine curettage may be helpful in preoperative diagnosis. However, when the lesion is completely within the myometrium, the curetting may not be helpful.(4). Due to the great similarity of ESS with normal endometrium, it may be impossible to diagnose it with certainty on curettage fragments, and the definitive diagnosis can be made only on a hysterectomy specimen. The pathologic diagnosis of endometrial stromal tumors requires the evaluation of vasculature and borders for infiltration and invasion. Endometrial stromal tumors are classified by tumor invasiveness with respect to margins and the degree of stromal differentiation and immunohistochemistry. High grade ESS is characterized by marked cytologic atypia, nuclear pleomorphism, high mitotic activity (>10MF/HPF) and extensive invasion.(5). It lacks features of normal endometrial differentiation and exhibit hemorrhage and necrosis and destructive myometrial invasion. UES shows increased staining for proliferation markers (Ki67, p16, and p53) and do not generally exhibit immunoreactivity against CD 10, ER, PR, desmin or SMA. UES also expresses the receptor tyrosine kinase CD117 and HER2 which are not typically found in ESS. Unlike ESS, there are no known chromosomal abnormalities associated with UES. On Ultrasound, Endometrial stromal sarcoma characterized as a heterogeneous hypoechoic endometrial mass, which can show extensive myometrial involvement. On MRI, these tumors appear as large masses with or without evidence of myometrial invasion. Inversion of uterus can be seen in MRI as absence of uterine fundus and U shaped uterine cavity.(6). Treatment of UES is total abdominal hysterectomy and bilateral salpingo-oophorectomy with pelvic lymphadenectomy.(7). In advanced stage of the disease adjuvant radiotherapy or chemotherapy can be given. If sarcoma is associated with inversion of the uterus, combined abdomino-vaginal approach is the

treatment of choice similar to our case. Adjuvant radiation is beneficial in decreasing pelvic recurrences but it has no effect on survival which was proved in randomized studies. Similarly, there is no effect on survival with adjuvant chemotherapy. The commonly used drugs are Doxorubicin, Ifosfamide, Doxetaxel and Gemcitabine which are under clinical trials.(8). These tumors lack ER and PR receptors hence they will not respond to hormonal therapy. The UES has more aggressive clinical course and poor prognosis than ESS. The 5 year disease free survival is 25%. Even in our case, our patient had very short period of survival after treatment.

CONCLUSION

Endometrial stromal sarcoma 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. Multianalysis from a large group of patients is necessary for predicting prognosis and to define proper treatment of endometrial stromal sarcoma. Although the most common cause of non-puerperal uterine inversion is a leiomyoma, a high index of suspicion for a coexisting malignancy must be maintained.

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