CASE REPORT OF A RARE ENTITY: ENDOMETRIAL STROMAL SARCOMA

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ABSTRACT

Endometrial stromal sarcoma (ESS) is a rare malignancy arising from the stromal cells of the endometrium. We present a case of a 38-year-old female who was admitted to the Emergency Room (ER) with heavy and foul-smelling vaginal bleeding. Further workup (examination) revealed a bulky uterus with a fibroid extending into the cervix. Ultrasound and MRI pelvis showed a large submucosal uterine fibroid with concerning features of possible rupture. The patient underwent total abdominal hysterectomy with bilateral salpingectomy. Intraoperatively, the fibroid was found to be necrotic and macerated, coming out of the cervical os. Histopathology examination revealed a low-grade ESS. The patient was discharged in stable condition. Follow-up imaging and clinical evaluations have shown no evidence of recurrence at three-months post-surgery.

Introduction

Endometrial stromal sarcoma (ESS) is a rare malignant tumor arising from the stromal cells of the endometrium. Less than 10% of all uterine mesenchymal neoplasms and 1% of all uterine malignancies are thought to be accounted for by endometrial stromal sarcoma.[1] ESS can present with variable symptoms, including abnormal vaginal bleeding, pelvic pain, and a palpable mass. Menorrhagia is the most typical presenting symptom of high-grade endometrial sarcoma and occurs at an average age of 61 years.[2] The diagnosis is confirmed through histopathological examination and the management is primarily surgical. Adjuvant therapies such as radiation and chemotherapy may be used in certain cases.[3]

Case Report

A 38-year-old married woman with five living children presented to the ER with heavy and foul-smelling vaginal bleeding. She had no previous medical history and was not on any medications. On physical examination, the patient was afebrile, and her vital signs were stable except for a high pulse rate. Laboratory tests revealed low hemoglobin levels. Ultrasound examination showed a bulky uterus with a fibroid extending into the cervix. MRI pelvis was performed, which revealed a large submucosal uterine fibroid in the right lateral wall displacing the endo-metrium to the left side with concerning features of possible rupture.

The patient underwent a total abdominal hysterectomy with bilateral salpingectomy. Intraoperatively, the fibroid was found to be necrotic and macerated, coming out of the cervical os.

Histopathology examination revealed a low-grade ESS with no lymphovascular invasion.

Immunohistochemical stains were positive for CD10, estrogen receptor (ER), and progesterone receptor (PR), while smooth muscle actin (SMA) and desmin were negative.

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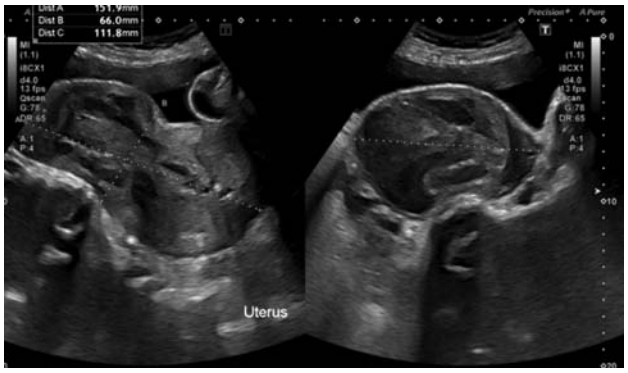
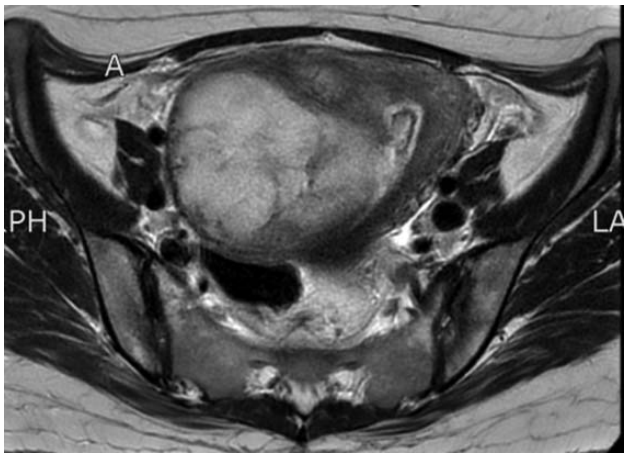


Figure 1: Ultrasound pelvis showed bulky uterus with a fibroid displacing the endometrium.



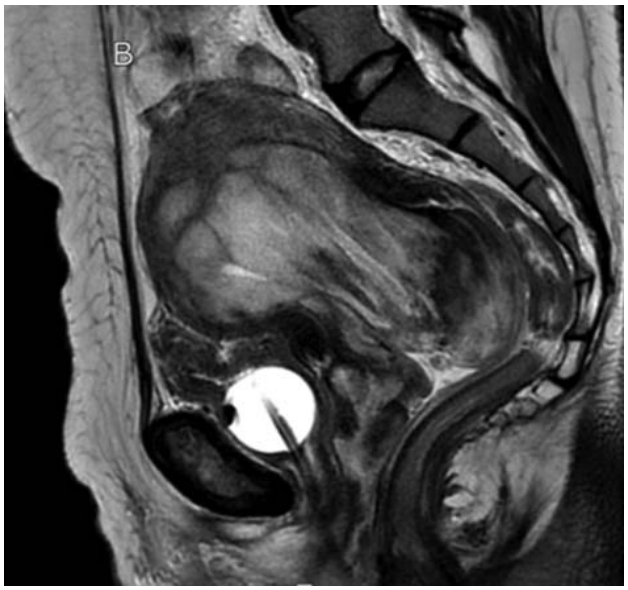
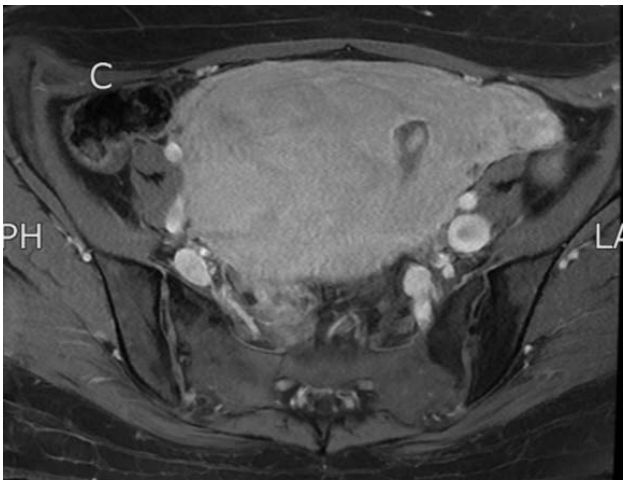


Figure 2: MRI pelvis: Image **A** and **B** (T2 axial and sagittal) demonstrate a well-defined heterogeneously hyperintense lobulated lesion with indistinct inferior margin originating from right lateral wall of uterus, extending medially and submucosally, protruding into the endometrial cavity, extending inferiorly, causing widening of the cervical canal and hanging into the vagina. It was pushing the endometrium towards the left side.



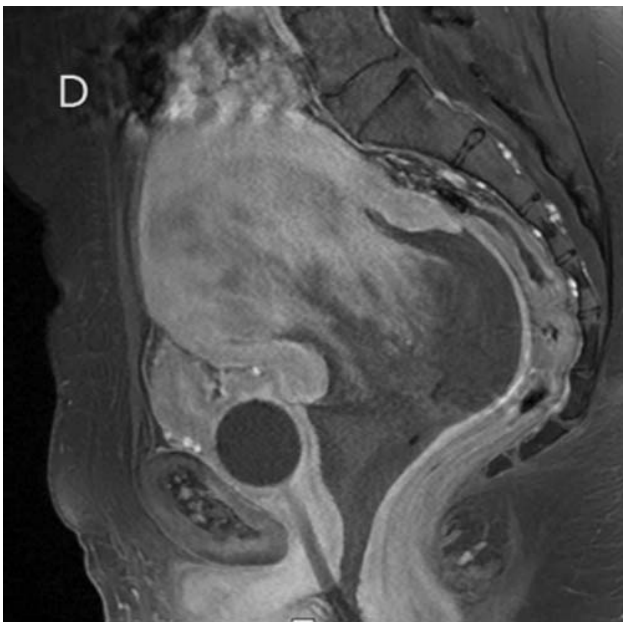
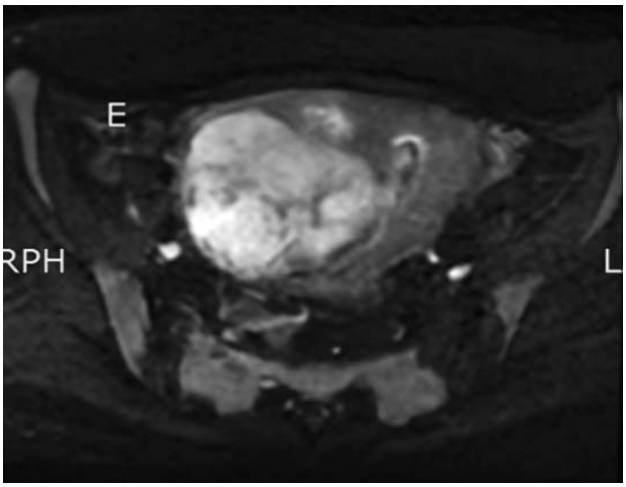


Image **C** and **D** (post contrast axial and sagittal) lesion shows heterogeneous post contrast enhancement and no enhancement was seen in the inferior aspect of this lesion on image **D**.



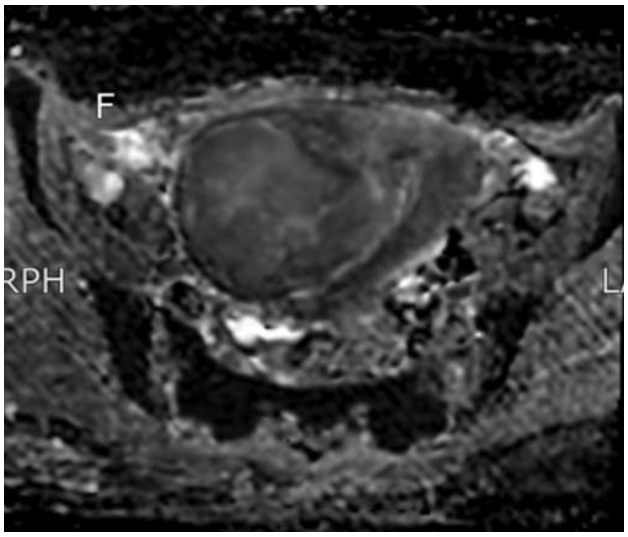


Image **E** and **F** (diffusion weighted images) show patchy areas of diffusion restriction in the lesion.

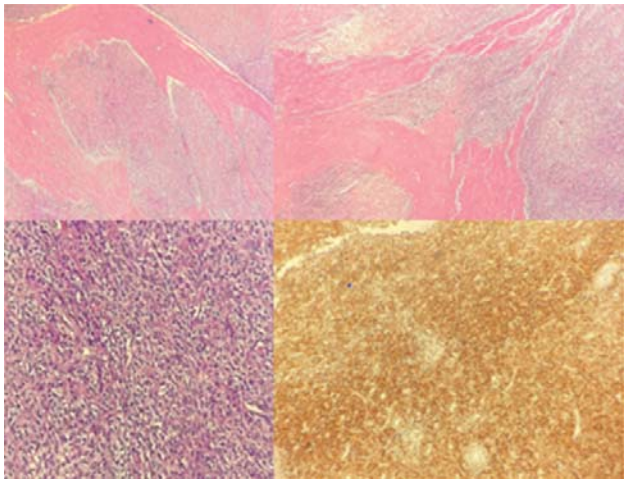


Figure 3: 4X Magnification: Low grade endometrial stromal sarcoma exhibiting widespread tongue like permeation into the myometrium. 10X Magnification: Monotonous oval to spindled cells with minimal cytologic atypia, vesicular chromatin and scant cytoplasm. Positivie expression of Immunohistochemical stain CD10 at 4X Magnification.

Patient was discharged on postoperative day six with instructions for wound care, medications, diet, and physical activity. However, a few days later she presented with wound dehiscence and pus discharge from the vagina and was readmitted for further management. She was vitally stable and afebrile. She was treated with resuturing of the abdominal wound, broad-spectrum antibiotics, antifungals for yeast in blood culture, and antiemetics and eventually discharged in stable condition.No adjuvant therapies were administered.

The patient was followed up at three months post-surgery and showed no recurrence or metastatic disease on CT scan chest abdomen and pelvis or clinical evaluation.

Patient underwent bilateral oophorectomy and bilateral pelvic lymph node dissection for complete staging after 1 month and histopathology revealed normal both ovaries and omentum with benign reactive pelvic lymph nodes. The patient is scheduled for regular follow-up visits with imaging studies.

Discussion

The endometrial stromal tumor usually affects postmenopausal women at an average age of 61 years.[2] And in literature we found that women with LGESS are usually younger with a median age of 45 and 55 years.[4] In our case the patient presented at 38 years, which is slightly towards the lower extreme. ESS does not show specific clinical manifestations or imaging characteristic features. The presenting feature in our case was menorrhagia which is usually the presenting complaint in cases of fibroids.

The initial diagnosis in our case was also submucosal fibroid with possible rupture as its inferior margin was indistinct and intraoperatively it was found to be necrotic and macerated coming out of the cervical os. However, the histopathology report revealed low grade endometrial stromal sarcoma. On initial imaging, there was no extrauterine spread of disease. Follow-up imaging did not reveal any features of disease recurrence and distant metastasis.

One of the case reports published on ESS reports an enlarged uterus with soft yellow necrotic and hemorrhagic tumor and in our case, we also found a necrotic and macerated fibroid coming out of the cervical os intraoperatively.[4]

A case of ESS was reported in a 41-year-old woman who underwent fertility testing but had no primary complaints. Imaging revealed a well-defined mass within the myometrium with multiple nodules giving a "nodule in nodule" appearance, and the patient was suspected of having LG-ESS and underwent total abdominal hysterectomy and bilateral adnexectomy. Histopathology identified LG-ESS as the cause.[5]

Another instance included a young woman who experienced menometrorrhagia for six months following delivery. Pelvic imaging revealed an intramural myoma

that was pressing on the endometrium. A pathology report and hysteroscopic removal of a 5 cm tumour from the uterus' anterior wall revealed low-grade endometrial stromal sarcoma. There was no sign of a residual tumour in the samples after TAH+BSO. After 27 months, a follow-up test and imaging revealed a disease-free period.[6] Toprak, U. et al reported a caseof a 24-year-old woman who complained of a menorrhagia and a slowly expanding sore mass in the lower abdomen. A cystic lesion in the left cornu was discovered by imaging workup, coupled with small intramural nodules, widespread thickening, and augmentation of the fallopian tube walls that extended through the broad and round ligaments to the peritoneum. An endometrial curettage sample had no signs of illness. In order to diagnose the low-grade endometrial stromal sarcoma without endometrial component, a thorough abdominal hysterectomy was done. However, there was a lymphangitic invasion of the cervical myometrium as well as other myometrial tissue throughout the uterus. The malignant process entered the peritoneum through the serosa. This patient's diagnostic imaging revealed nothing, and the lesions were labelled as being entirely myometrial. The stromal cells of the endometrium give birth to ESS malignancy, and histological analysis is used to confirm the diagnosis. Premenopausal women account for more than half of instances, especially in those with low-grade endometrial stromal sarcoma, which tends to affect younger patients (mean age, 39 years).[7]

Preoperative diagnosis of ESS is difficult and literature reports around 75% cases are diagnosed as benign leiomyoma. As the tumor grows in intramural course rather than intracavitary, therefore endometrial curettage and histopathological examination do not help due to similarity with normal endometrium.[8]

Because endometrial stromal tumours can have a variety of morphologies and frequently involve extrauterine locations, diagnosing one can be difficult. Studies using immunohistochemistry may aid in making the right diagnosis. Although surgery is the main form of treatment, adjuvant therapies including radiation and chemotherapy may be utilised in some circumstances. The prognosis for ESS is generally favourable, with a five-year survival rate for low-grade tumours of about 90%.

Previously, research works examined the many kinds of the disease and for a long time, the classification of endometrial stromal tumours was challenging, therefore it was not always possible to distinguish between LG-ESS, high-grade endometrial stromal sarcoma, and undifferentiated uterine sarcoma in previous investigations. Surgery with hysterectomy and adnexectomy is the first-line treatment for LG-ESS. The advantages of lymphadenectomy and tumour debulking remain unknown. Because radiation only provides locoregional control, and given the generally fair prognosis of LG-ESS patients, its benefits must be evaluated against its negative effects. Repeat surgery is the first option in the event of a recurrence.[9]

The patient in our case presented with menorrhagia for 6 months which is a chief complaint in most of the cases reported in literature while rarely it may present without any major complaint diagnosed on workup for unable to conceive.

Conclusion

ESS is a rare malignancy that presents with various symptoms, including abnormal vaginal bleeding and pelvic pain. The radiological findings are nonspecific for fibroid versus degenerating fibroid and diagnosis is confirmed through histopathological examination. The management is primarily surgical, However, adjuvant therapies may be consider in certain cases.

Conflict of Interest: Declared

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