Case Report

A Case of High-grade Endometrial Stromal Sarcoma: A Poignant Allegory

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Abstract

Endometrial stromal sarcomas (ESS) are rare tumors of endometrial stromal origin that account for approximately 1% of all uterine malignant neoplasms and are responsible for a significant proportion of mortality due to uterine malignancies. There are immense case reports on low-grade ESS, but there is a paucity of data on high-grade ones, probably, because these cases generally present in advanced stages and have a high case fatality rate. Moreover, there have been several refinements in the classification of these tumors. We, herein, provide an update on this topic and discuss the poignant outcome of a case of high-grade ESS.

Keywords: *Allegory, endometrial stromal sarcoma, high grade*

Introduction

Endometrial stromal sarcomas (ESS) are rare tumors of endometrial stromal origin that account for approximately 1% of all uterine malignant neoplasms and are responsible for a significant proportion of mortality due to uterine malignancies.[1,2] There have been many changes in the classification of ESS and the most recent one categorizes them into three types: low-grade endometrial stromal sarcoma (LGESS), high-grade endometrial stromal sarcoma (HGESS), and undifferentiated uterine sarcoma (UUS).[3] The basic characteristic of these tumors is that they remain in disguise unless proven by histopathology after hysterectomy and hence often leads to preoperative misdiagnosis. There is no single investigation till date which can make a definite preoperative diagnosis of ESS. Even dilatation and curettage or endometrial biopsy fails to diagnose it as the curetting is taken from the endometrium and ESS tends to grow toward the myometrium.[4] Herein, we provide an update on this topic and discuss the poignant outcome of a case of high-grade ESS. Many case reports are there on low-grade tumors diagnosed after hysterectomy, but there is a paucity of literature on cases of high-grade and undifferentiated stromal sarcomas.

Case Report

A 55-year-old female, P3003, presented with postmenopausal spotting on and off for the past 6 months, cough with expectoration, and significant weight loss for past 2 months. For these complaints, she had been put on empirical antitubercular drugs at some outside hospital. She took these medications for 2 months but had no improvement in her chest condition and rather the bleeding episodes and pain in lower abdomen increased significantly. She was then referred to our center. The patient was nondiabetic and nonhypertensive, and there was no history of long-term exposure to estrogen. She was postmenopausal for 3 years. On examination, her general condition was poor. She was cachexic with weight 27 kg and body mass index of 15 kg/m^2. On auscultation, bilateral air entry was decreased and rhonchi were present. Abdominal examination revealed a scaphoid abdomen and a lump corresponding to 14 weeks size gravid uterus, hard in consistency, and restricted mobility in the suprapubic region. On per speculum examination, the cervix could not be visualized as it was pointing acutely forward. Hence, cervical screening and endometrial biopsy could not be done. On bimanual examination, a hard mass of around 14 weeks was felt, which was nonmobile and nontender.

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Her hemoglobin was 7.1 g%. Rest all investigations were within normal limits. Ultrasonography showed evidence of a well-defined heterogenous hypoechoic mass lesion 20 mm × 17 mm in part of the myometrium of the fundal region likely intramural fibroid and bulky uterus. However, the magnetic resonance imaging (MRI) report was suggestive of endometrial carcinoma involving more than half of the myometrium. Computed tomography (CT) of the chest was advised to rule out metastasis. However the patient was lost to follow-up for about 2 weeks when she returned back with CT report suggestive of traction bronchiectasis and discrete random nodules with feeding vessel signs in the right lower lobe of lung, likely metastases. Surprisingly, there was enlargement of uterine mass to approximately 20 weeks size in 2 weeks duration with increase in pain abdomen and fall in hemoglobin to 5.2 g %. A repeat ultrasonography was done which showed a large lobulated mass lesion with heterogeneous echotexture, size 13.7 cm × 7.3 cm with increased vascularity on Doppler. The uterus with bilateral adnexa could not be separately seen.

At the same time, the patient complained of severe pain in the left leg with unilateral edema. Homan's sign was negative. Doppler of the leg was normal. Hence, deep vein thrombosis was excluded. Seeing the rapid growth of mass and falling hemoglobin, a working diagnosis of leiomyosarcoma was made. After receiving two units of packed red blood cells, the patient was posted for exploratory laparotomy.

Intraoperatively, the mass was up to the umbilicus, adherent to the omentum and sigmoid colon. It had actually ruptured inside the abdomen with tumor tissue and necrosed material adherent to the bowel, abdominal wall, and lateral pelvic wall [Figure 1a]. Adhesiolysis was done. Total abdominal hysterectomy with bilateral salpingo-oophorectomy and infracolic omentectomy was done. The specimen was sent for histopathology.

The resected specimen measured 13 cm × 13 cm × 4 cm. The outer surface was congested with multiple surface deposits. Cut section showed dilated endometrial and endocervical cavity. Endometrial cavity was filled with friable gray–brown growth [Figure 1b]. Multiple small growths were also seen in the myometrial cavity.

Microscopic examination

Tumor was arranged in sheets and lobules and separated by fibrous septae of varying thickness. Individual tumor cells were small to medium sized, round having round to irregularly contoured, vesicular nuclei, small nucleoli, and scant to moderate amounts of eosinophilic to granular cytoplasm. Brisk mitotic activity is seen. Several bizarre cells and a few multinucleated tumor giant cells and large areas of necrosis were seen. Hemorrhage, fibrinous deposits, and several lymphovascular emboli were seen. Morphologically features were suggestive of HGESS [Figure 2]. The left ovary and the left fallopian tube also showed metastatic tumor deposits with lymphovascular emboli. The tumor involved the muscularis propria of the appendix.

Immunohistochemistry was positive for CD 10 [Figure 3] and negative for estrogen and progesterone receptors.

The patient received three units of packed cells, eight units of fresh frozen plasma, and four units of platelet intraoperatively because of massive blood loss and subsequent hypotension. There was generalized oozing from all the sites. Hence, the pelvis was packed to give a tamponade effect and abdomen closed. She was kept on ventilator support in the intensive care unit. After 48 h, we reopened the abdomen to remove the pack.

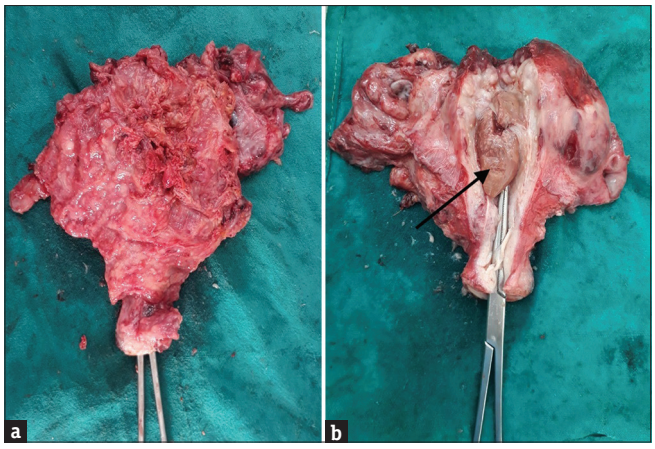

Figure 1: (a) Gross appearance (b) cut section with arrow showing a friable growth within the endometrial cavity

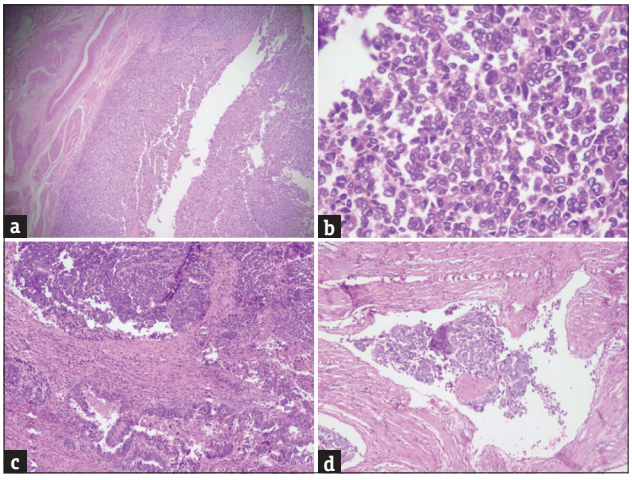

Figure 2: High-grade endometrial stromal sarcoma. (a) Tumor (H and E stain, ×10) (b) tumor (H and E, ×40), (c) tumor invading the muscularis layer of the uterus H and E × 10, (d) lymphovascular invasion

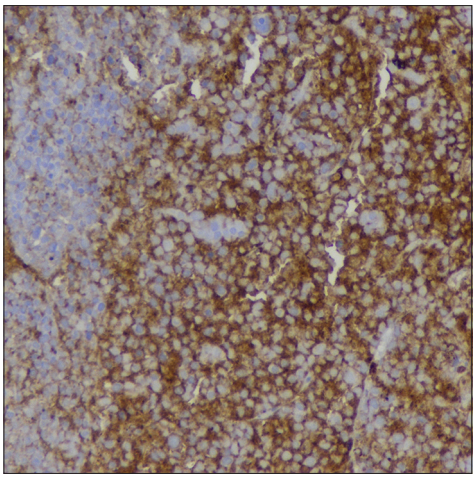

Figure 3: Immunohistochemistry showing positivity for CD10

There was no bleeding, but she had developed ascites. A drain was kept *in situ*. In the postoperative period, the patient was conscious and responsive but could not be extubated due to her poor chest condition. She developed herpes zoster on the 4th postoperative day and that justified the pain that she had preoperatively to be preherpetic neuralgia. Intravenous acyclovir was given for 7 days.

Extubation was tried many times, but she failed to maintain without ventilatory support. Bronchial aspirate revealed the presence of *Acinetobacter* species for which cefoperazone-sulbactam and colistin were given for 8 days. To wean her off the ventilator, tracheostomy was done. However, despite all efforts, patient expired on day 28 of operation.

Discussion

Uterine sarcomas are rare malignant uterine neoplasms that are responsible for a large majority of uterine cancer-associated deaths. They are basically of two types: mesenchymal tumors which include leiomyosarcoma, endometrial stromal sarcoma, undifferentiated endometrial sarcoma, and smooth muscle tumors of uncertain malignant potential; mixed epithelial and mesenchymal tumors which include carcinosarcoma, mullerian adenosarcomas, carcinofibromas, adenofibromas, and adenomyomas.[3] ESS are the second most common mesenchymal tumors after leiomyosarcoma. The common age of presentation is between 40 and 60 years.

In general, patients do not have identifiable risk factors; however, some studies suggest an increased risk for uterine sarcoma in women with a history of obesity, diabetes mellitus, younger age at menarche, or exposure to tamoxifen.[5] Our patient did not have any of the mentioned risk factors. Women with low-grade ESS usually present with leiomyoma-like symptoms such as abnormal uterine bleeding, abdominal or pelvic pain or pressure symptoms due to enlarging abdomen, abnormal or foul-smelling vaginal discharge, or postmenopausal bleeding.[5] Hence, they are generally operated in view of symptomatic leiomyoma, and it is only after hysterectomy that the actual diagnosis comes into picture. Sometimes, this may be an incidental finding in the uterus operated for uterovaginal prolapse.[6] Hence, the clinical course of LGESS is usually indolent, and the overall prognosis is also better. However, cases of high-grade uterine sarcoma generally present in advanced stage (III/IV) as it was in our case and have a very aggressive clinical course. Our patient was also diagnosed with intramural fibroid initially and within a matter of few weeks, turned into highly malignant tumor. The overall survival is <2 years.

The differential diagnosis between uterine sarcoma and benign leiomyoma is difficult when made only by ultrasonography or MRI; it usually requires an additional preoperative diagnostic procedure. Ultrasound-guided needle biopsy may be a reliable preoperative diagnostic procedure for such uterine tumors having heterogenous appearance and suspected malignancy.[7]

Since approximately more than 50% of patients of HGESS present with advanced stage disease (Stage III/IV), whenever possible, surgery, i.e., hysterectomy and bilateral salpingo-oophorectomy should be undertaken in order to remove maximum disease and performing accurate staging, followed by adjuvant radiation and/or chemotherapy.[8,9] The risk of lymph node metastases has been reported to be between 3% and 11%; thus, routine pelvic lymphadenectomy is not routinely indicated.[10]

If we compare the histologic picture, high-grade ESS comprises of a tumor composed of atypical cells resembling endometrial stromal cells but lacking the degree of atypia and pleomorphism of UUS, whereas UUS exhibits myometrial invasion, lacking any resemblance to proliferative phase endometrial stroma, with severe cytologic atypia, often with multinucleated and bizarre cells and brisk mitotic activity (>20 mitoses/10 HPFs) and with no specific type of differentiation. Our case had the histologic features of HGESS.

To differentiate endometrial stromal tumors from smooth muscle tumors, a panel of markers in immunohistochemistry may be needed including smooth muscle markers (smooth muscle actin (SMA), desmin, H-caldesmon) and stromal markers (CD10, Bcl-2) in an attempt to delineate the cell of origin.[11]

In our patient, we did optimal cytoreduction and had planned for adjuvant chemotherapy, but the poignant allegory was that despite all treatment, the patient did not survive. The survival of the patient depends on the stage of disease. Had she come earlier, may be, she could have had better survival. This is the allegory of these cancer patients. By the time they reach an apt place for treatment, they are already in advanced stage.

Conclusion

High-grade endometrial sarcomas are rare and aggressive tumors having a very poor prognosis. Early diagnosis in the initial stages is of paramount importance to increase the overall survival.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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