

CASE REPORT

Gynecologic Oncology

High-grade endometrial stromal sarcoma: A case report

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Abstract

Background: High-grade endometrial stromal sarcoma (HG-ESS) is a homologous uterine sarcoma of a mesenchymal origin. Its natural course is poorly understood, making preoperative diagnosis difficult.

Case presentation: A 55-year-old nulliparous presented to the gynecological oncology ward as a referral with progressive abdominal distension and pain, constipation, and postprandial vomiting. Computed tomography (CT) scan of the abdomen revealed a bulky uterus with multiple calcified masses with cystic degeneration. A chest CT scan revealed bilateral upper and lower lobe nodules with

mediastinum and hilar adenopathy. She was scheduled for total abdominal hysterectomy and bilateral salpingo-oophorectomy. A histopathological examination confirmed HG-ESS. The patient survived for three months following surgery.

Conclusion: HG-ESS is rare. This case highlights the nonspecific presentation, aggressive nature of the tumor, rapid progression of the disease, and the challenges clinicians face in managing patients who do not honor their appointments with resultant poor overall survival in the late stage.

Keywords: HG-ESS, uterine sarcoma

Introduction

Uterine sarcomas are rare tumors that account for 3 - 9% of all malignant uterine tumors (1). They arise from the connective tissue and smooth muscles of the uterus. Five broad types include leiomyosarcomas, carcinosarcomas, endometrial stromal sarcomas, adenocarcinomas, and undifferentiated stromal sarcoma, constituting 55-70%, 30%, 20%, 5%, and 10% of the uterine sarcomas, respectively (2). The World Health Organization (WHO) classifies endometrial stromal sarcomas (ESS) into low-grade endometrial stromal sarcoma (LG-ESS), high-grade endometrial stromal sarcoma (HG-ESS), and undifferentiated uterine sarcoma (UUS) (2,3). This is a case of a high-grade uterine sarcoma in a 55-

year-old nulliparous who also had an incident diagnosis of coronavirus disease 2019 (COVID-19).

Case presentation

A 55-year-old nulliparous presented to the gynecological oncology ward at Kenyatta National Hospital as a referral with progressive abdominal pain and swelling, early satiety, postprandial vomiting, and constipation for three months. She did not have abnormal vaginal bleeding, difficulty in breathing, or cough. She attained menarche at 15 years and had a prior regular 28-day cycle. She had no history of contraceptives, tamoxifen, alcohol and cigarette use, previous surgical procedures, radiation, blood transfusion, or Papanicolaou (Pap) test. She was HIV seronegative with no history of hypertension, diabetes, or cancer. On examination,

she was mildly pale and dehydrated. Her blood pressure was 120/85mmHg, pulse rate 115 beats per minute (BPM), respiratory rate 15 BPM, temperature 36.3°C, and oxygen saturation 96%. A speculum examination was routine. Her hemoglobin level was 10.8g/dl, and she had normal liver and renal function tests. No tumor markers were evaluated. She had a grossly distended abdomen with a firm mass corresponding to a term-size uterus, slightly mobile, smooth, and nontender (**Figure 1**).



Figure 1: Massive abdominal distention by a giant mass.

Contrast-enhanced computed tomography (CT) of the pelvis and abdomen revealed a bulky uterus measuring 30.1 x 16.7 x 25cm, multiple uterine masses (largest 18 x 19cm), with calcification and cystic degeneration, distorted uterine outline, marked mass effect with compression and displacement of the bowel loops and urinary bladder. No peritoneal or pelvic nodes were seen. Chest CT revealed bilateral upper and lower lobe nodules (largest 1.87 x 2.16cm) with mediastinal and hilar adenopathy. Electrocardiogram and echocardiogram were normal. She tested positive for SARS-CoV-2 on a preoperative workup. She was scheduled for total abdominal hysterectomy (TAH) and bilateral salpingo-oophorectomy (BSO). Intraoperatively, the uterine mass extended to the xiphisternum, irregular with omental adhesions anteriorly and laterally to the pelvic sidewalls (**Figure 2**). Additionally, omentectomy and adhesiolysis were performed, and specimens were taken for histopathology analysis.

Postoperatively, the patient was isolated due to coronavirus disease. She was managed with antibiotics and analgesics and was discharged stable on day 10. The histopathology specimen revealed a sizeable nodular mass measuring 30 x 30 x 22cm, attached fatty tissue measuring 28 x 10mm, and weighing 9.5kg (21lb). The cut surface

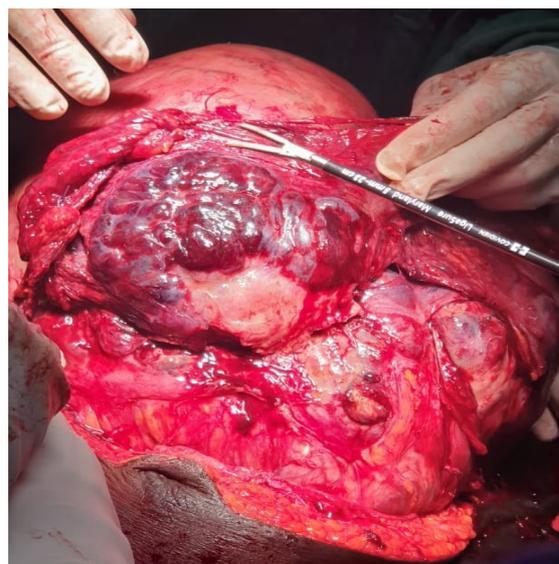


Figure 2: Intraoperative findings of a highly vascularized tumor.

of the tumor was white with a whorled appearance and areas of extensive hemorrhage and cystic change (**Figure 3**). Microscopically, the sections showed a highly vascular tumor composed of plump-spindled cells arranged in a haphazard, storiform, and herringbone patterns, polygonal cells with an epithelial appearance. They were moderately pleomorphic with brisk mitoses (720 per 10 high power field (HPF)) and areas of coagulative necrosis (**Figure 4**). A diagnosis of HG-ESS was made. A review a month later reported a hemoglobin level of 11.5g/dl. A plan of adjuvant chemotherapy with doxorubicin/ ifosfamide was made. However, she was temporarily lost to follow-up, and on her second review two months later, she was weak with a hemoglobin level of 9.1g/dl. A plan to transfuse and start chemotherapy was made, but she succumbed two weeks later before commencing treatment.

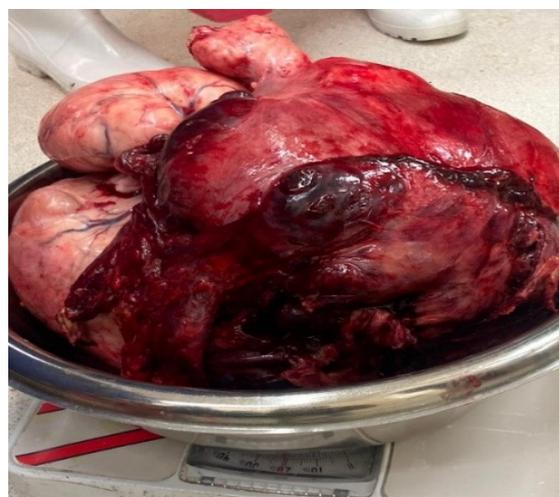


Figure 3: Gross appearance of the specimen.

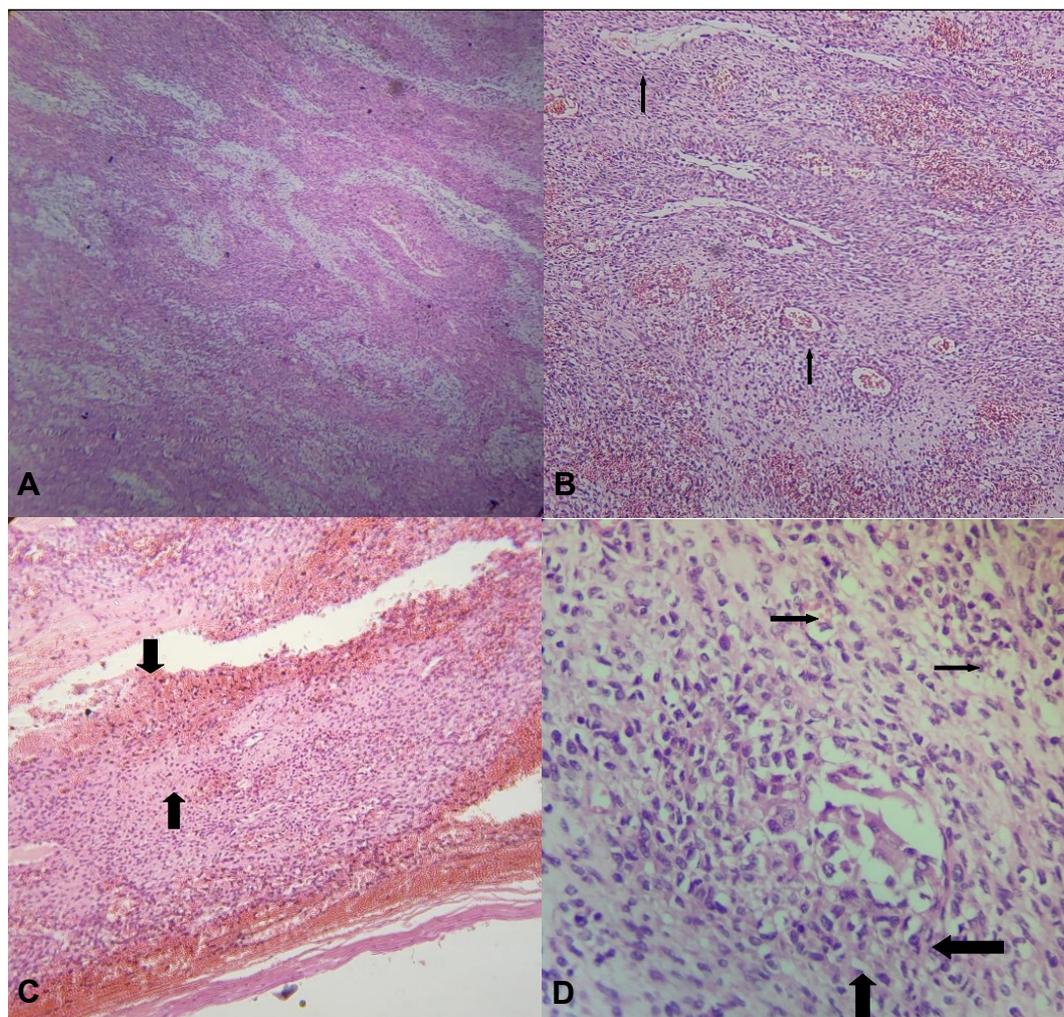


Figure 4: **A:** Highly cellular tumor (Hematoxylin and Eosin (H&E) stain x100 magnification); **B:** Highly vascular tumor (arrows), pleomorphic with plump-spindled cells (H&E x200 magnification); **C:** Coagulative necrotic and hemorrhagic areas (arrows), well-encapsulated (H&E x200 magnification); **D:** Mitotic figures (thin arrows), giant bizarre and vesicular cells (thick arrows) (H&E x400 magnification).

Discussion

Malignant neoplasms of the corpus uteri include undifferentiated sarcomas, smooth muscle tumors, mesenchymal tumors, endometrial stromal tumors, mixed epithelial tumors, carcinosarcomas, adenomyomas, and leiomyosarcomas; the latter being the most prevalent at 55-70% (1). HG-ESS lacks definitive risk factors, and diagnosis is often made upon routine histopathology after myomectomy or hysterectomy is performed because of suspected leiomyoma (4). Preoperatively, it is difficult to differentiate a sarcoma from a leiomyoma based on symptoms and examination. Diffusion-weighted magnetic resonance imaging (MRI) may differentiate degenerated leiomyomas from sarcomas (5). Intralesional hemorrhage may suggest sarcomas; however, it is not conclusive. Additionally, there are no specific tumor markers for sarcomas (4,5). Endometrial sampling has a remarkably lower

predictive value and yields a preoperative diagnosis in only 33-68% of the cases (3,6).

HG-ESS symptomatology may include abnormal postmenopausal bleeding, uterine mass, pelvic pain, and abdominal pain and distention (7). There is an increased risk of uterine sarcomas in women with diabetes mellitus, pelvic radiation, obesity, early menarche, and tamoxifen use (1,3). The patient in the presented case did not have any risk factors. Late presentation is common, as in this case, which requires an aggressive clinical course to resolve. In HG-ESS, TAH and BSO with optimal cytoreduction to <2cm of residual disease is done. Lymphadenectomy with preoperative imaging evidence of enlarged lymph nodes or intraoperative findings of lymphadenopathy is indicated in patients without the extrauterine disease (8). Adjuvant chemotherapy is effective with gemcitabine, docetaxel, ifosfamide, and doxorubicin (3). The role of radiotherapy is unclear but is an option in selected high-risk patients with

bulky tumors, advanced stage, deep myometrial invasion, and positive resection margins (2). Hormonal therapy with aromatase inhibitors, gonadotropin-releasing hormone analogs, and progestins is effective in recurrent, metastatic, or unresectable LG-ESS but has little effect on HG-ESS since it is aggressive and rarely expresses estrogen and progesterone receptors (9). The overall survival and progression-free survival of high-grade uterine sarcoma range between 11 - 23 and 7 - 11 months, respectively (10). This is primarily attributed to the aggressive course of the disease and poor response to adjuvant therapies of radiation and systemic chemotherapy regimens (4).

Conclusion

HG-ESS is rare. This case highlights the nonspecific presentation, aggressive nature of the tumor, rapid progression of the disease, and the challenges clinicians face in managing patients who do not honor their appointments with resultant poor overall survival in the late stage.

Consent for publication

Informed consent for publication was obtained from the patient.

Acknowledgement

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Declarations

Conflict of interests

The authors declare no conflicts of interest.

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