



Case report

A giant metastatic low-grade endometrial sarcoma requiring surgical management

Freddy Houéhanou Rodrigue Gnanon^{a,*}, Ismaïl Lawani^b, Habib Ndome Natta N'tcha^a, Edwin Kelly Haag^a, Francis Moïse Dossou^b, Delphin Kuassi Mehinto^a

^a Department of Visceral Surgery, National University Hospital Hubert Koutoukou Maga (CNHU-HKM), Cotonou, Benin

^b Department of General Surgery, Departmental University Hospital Oueme-Plateau, Porto-Novo, Benin

ARTICLE INFO

Keywords:

Low-grade endometrial stromal sarcoma
Cytoreduction surgery
Pulmonary embolism
Case report

ABSTRACT

Introduction and importance: Low-grade endometrial stromal sarcomas are relatively rare tumors. We here report a case of a woman presenting with a giant metastatic low-grade endometrial stromal sarcoma with thromboembolic complications requiring urgent surgical management.

Case presentation: A 58-year-old obese female was admitted, with a voluminous abdominopelvic mass, due to complications related to its size and extent. The tumor derived from the uterus and invaded the ureters, bladder and rectum. It compressed the right iliac vessels causing both deep vein thrombosis and pulmonary embolism. She developed a painful irreducible umbilical. We proceeded with a debulking surgery (hysterectomy with bilateral salpingo-oophorectomy). Histological findings were consistent with a low-grade endometrial stromal sarcoma.

Clinical discussion: Low-grade endometrial stromal sarcomas are generally low-grade malignant neoplasms with an indolent clinical course. Surgery is the cornerstone of treatment. In low-income countries, malignancies are more often diagnosed at a late stage, which limits therapeutic options. Cytoreduction is recommended in advanced tumors with extrauterine manifestation, depending on symptoms and with palliative intent.

Conclusion: Low-grade endometrial stromal sarcomas are indolent uterine malignancies with metastatic potential. Even in advanced cases, cytoreduction must be considered.

1. Introduction

Endometrial stromal sarcomas (ESS) are relatively rare tumors. They account for 7–25% of all uterine sarcomas and less than 1% of all uterine malignancies [1–3]. The World Health Organization (WHO) classifies ESS into three categories: low-grade endometrial stromal sarcoma (LG-ESS), high-grade endometrial stromal sarcoma (HG-ESS), and undifferentiated uterine sarcoma (UUS) [4]. LG-ESS are indolent tumors with metastatic potential and a favorable prognosis [5]. Their treatment is largely surgical.

Late presentation of cancers is commonplace in most low-income countries in sub-Saharan Africa [6]. We here report the surgical management of a 58-year-old woman presenting at the National University Hospital of Cotonou (Benin) with a giant metastatic LG-ESS with an embolic complication. The particularity of this case lies not only in the size of the primary tumor, but also in the indication for surgical

treatment for a metastatic uterine cancer. This work has been reported in line with the SCARE criteria [7].

2. Case presentation

A 58-year-old obese black woman (BMI = 37 kg/m²) was admitted to the intensive care unit of the cardiology department with respiratory distress. She had been carrying an abdominal mass growing for the past four years associated with recent metrorrhagia and hematuria. She had been menopausal for 18 years with no estrogen replacement therapy and had a history of hypertension.

Her physical examination found a low oxygen saturation with polypnea and tachycardia, a large abdominopelvic mass and a swollen right lower limb.

Computed tomography revealed a massive bilateral proximal pulmonary embolism, a giant abdominopelvic mass and lung metastasis

Abbreviations: ESS, endometrial stromal sarcoma; LG-ESS, low-grade endometrial stromal sarcoma.

* Corresponding author at: 01 BP 188 Cotonou, Benin.

E-mail address: fredgnang@yahoo.fr (F.H.R. Gnanon).

<https://doi.org/10.1016/j.ijscr.2022.107163>

Received 25 March 2022; Received in revised form 1 May 2022; Accepted 1 May 2022

Available online 4 May 2022

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(FIGO stage IVB). The tumor predominantly liquid, was multi-partitioned (Fig. 1). The mass compressed the right iliac vessels. It infiltrated the posterior wall of bladder and the ureters with bilateral ureterohydronephrosis. Laboratory blood tests revealed renal failure and elevated D-dimers.

The patient received anticoagulant treatment at a curative dose in the cardiology intensive care unit. She developed a painful irreducible umbilical hernia and was referred to the visceral surgery department. We decided on a resection of the mass. During the operation, we discovered a tumor estimated at 50 cm (Fig. 2) deriving from the uterus. A cystic mass of 5 cm was also found on the right ovary. We decided to perform a debulking surgery. We proceeded with a hysterectomy with bilateral salpingo-oophorectomy. The surgery being palliative, and given local and distant extension, the portion of the tumor invading the bladder, ureters and rectum was not resected. A double-J ureteral stent was put on the right side but could not be completed on the left ureter. The total duration of surgery was 4 h. The intraoperative blood loss was significant and required the transfusion of 3 units of red blood cells. The postoperative course was marked by left pyelonephritis successfully managed by antibiotics. The patient was discharged from the hospital with oral anticoagulant treatment (Rivaroxaban 20 mg per day) and instructions for follow-up by the cardiologist.

On pathological examination (Fig. 3), the proliferation resembled a cytogenic chorion, with monomorphic cells endowed with an oval nucleus. Some of the cells were spindle-shaped. Cellular atypia was discreet. Mitotic activity was low (less than 5 per 10 high-powered fields). The tumor infiltrates the entire thickness of the myometrium. Lymphovascular space invasion was identified. Tumor necrosis was estimated at less than 50%. A dermoid cyst was found on the right ovary. The fallopian tubes and left ovary were histologically normal. Immunohistochemistry showed a strong and diffuse expression of CD10. The histological findings were consistent with a LG-ESS.

According to guidelines [8], hormone therapy with an aromatase inhibitor was started at a daily dose of 2.5 mg. Six months after surgery, the patient had no clinical complication and showed no sign of disease progression. However, two months after that she passed away at home, from an acute abdominal pain as reported by the family, before she could reach hospital.

3. Discussion

The term “endometrial stromal tumor” applies to neoplasms typically composed of cells that resemble endometrial stromal cells of the proliferative endometrium. LG-ESS are generally low-grade malignant neoplasms with an indolent clinical course. It frequently occurs in women from 40 to 55, and more than 50% of patients are premenopausal [9]. This was the case for our patient. Obesity, diabetes mellitus, and early menarche are reported to be associated with an increased risk of LG-ESS [10]. Our patient had morbid obesity. These tumors often become apparent through pathologic vaginal bleeding, which is sometimes combined with uterine enlargement and associated symptoms such as lower abdominal pain [9].

As they increase in size, symptoms may appear due to compression of the surrounding anatomic structures, resulting in a significant increase in the incidence of thromboembolism and urinary retention [11,12].

Most cases of LG-ESS (60%) present with FIGO stage I disease, with only 20% presenting with stage IV, metastatic disease [2]. In low-income countries, malignancies are more often diagnosed at a late stage due to poor access to healthcare, lack of awareness, diagnostics, and healthcare capacity [6]. The patient in this case lived for several years with a voluminous abdominal mass and did not seek medical care until she had complications.

Staging is the most important prognostic factor, with stage I and II tumors having a 5-year survival rate exceeding 90%. In contrast advanced-stage tumors have a 5-year survival rate of 40 to 50% [2,9,13]. According to a large population-based study conducted in the United States [2], black people had the lowest disease-specific survival rates when compared to the other races.

Hysterectomy is the cornerstone of the treatment of LG-ESS at an early stage [14]. Cytorreduction is recommended in advanced tumors with extrauterine manifestations [15,16]. The extent of surgery has to be decided on an individual basis, depending on symptoms and with palliative intent [13]. In our case, surgery was indicated to manage the painful irreducible umbilical hernia and the thromboembolic complications of the uterine mass. Moreover, cytorreduction significantly improved the outcome and the quality of life of our patient.

Although surgical castration is a logical intervention for a hormone-sensitive disease, the clinical benefit remains unproven [2,17,18]. Bilateral salpingo-oophorectomy in premenopausal women always needs to be critically discussed. This issue has less importance in



Fig. 1. Computed tomography, transversal (a) and sagittal (b) sections showing a 32 × 28 × 18 cm abdominopelvic heterogenous mass (white arrow).

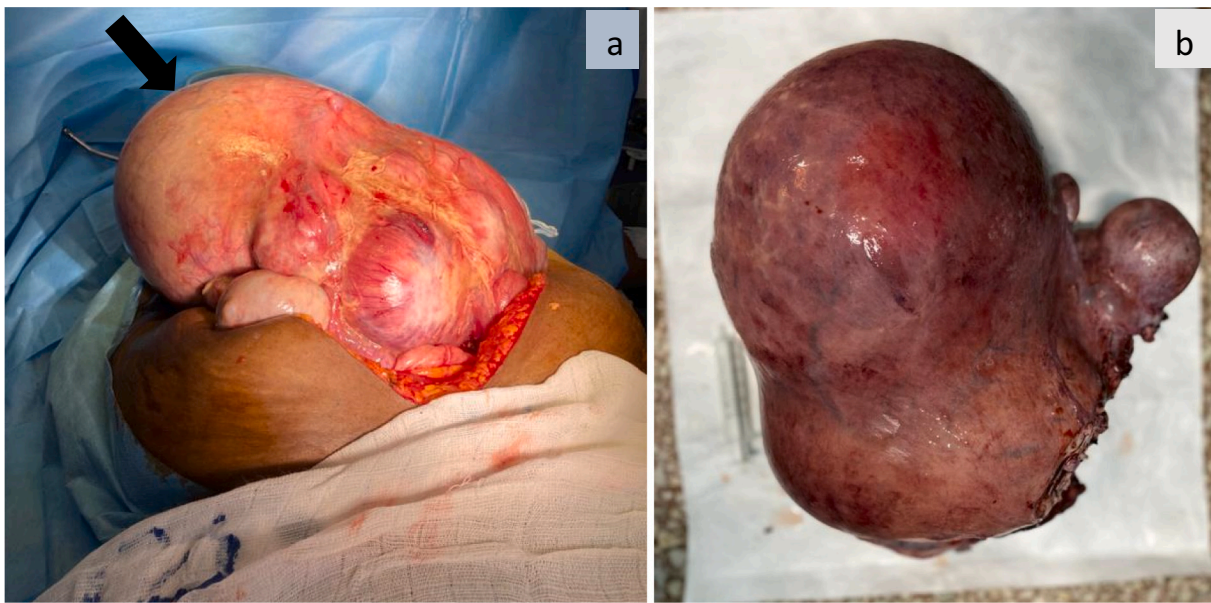


Fig. 2. a. Surgical extraction of a voluminous uterine tumor; b. the resected specimen 32 × 25 × 13 cm.

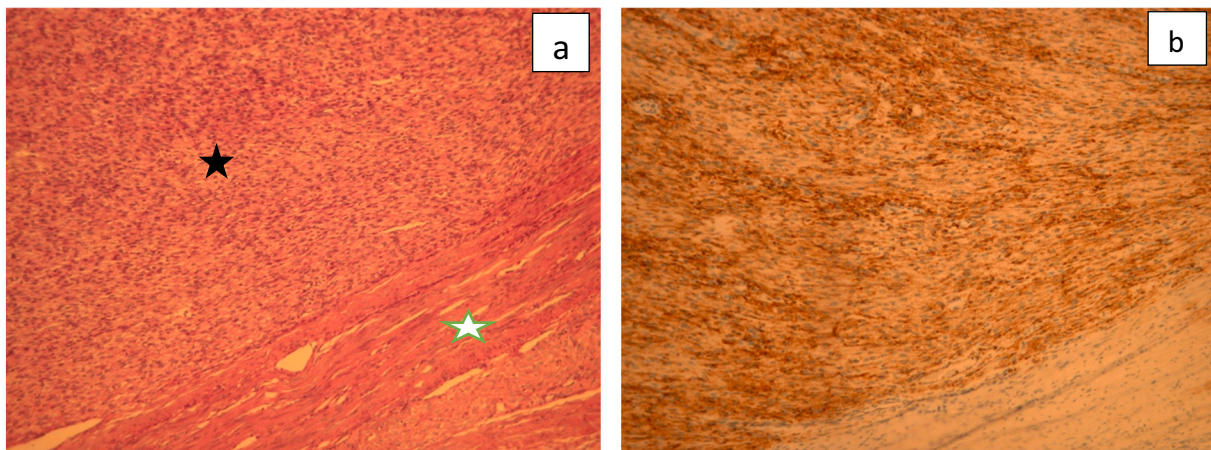


Fig. 3. Proliferation resembling a cytotrophoblastic chorion with discrete cellular atypia and low mitotic activity (black star), less than 5 per 10 high-powered fields; myometrium (white star) is infiltrated (a). Strong and diffuse staining of CD10 at immunohistochemistry (b).

perimenopausal and postmenopausal women [18].

Systematic lymphadenectomy for ESS is not indicated. Lymph-node involvement in tumors with other obvious extrauterine involvement, such as in our case, is unlikely to be predictive of prognosis [14,17].

The systemic treatment of metastatic LG-ESS exploits their sensitivity to hormonal therapies. Their efficacy has been well documented in recurrent and advanced disease settings. Typical hormone therapy includes aromatase inhibitors, and progestins (megestrol acetate or medroxyprogesterone acetate) [8,18,19]. We decided to use aromatase inhibitors due to their lower toxicity profiles and broader therapeutic index [19,20].

4. Conclusion

LG-ESS are relatively rare tumors, with an indolent course that can slowly grow into voluminous tumors and lead to complications. We reported the case of a locally advanced and metastatic LG-ESS presenting with pulmonary embolism and irreducible umbilical hernia. Cytoreduction can improve the outcome and is a reliable option even in metastatic disease.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Ethical approval

Not applicable.

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Guarantor

The guarantor who is responsible for the present case report is Gnangnon FHR.

Research registration number

Not applicable.

CRedit authorship contribution statement

Study conception and design: FHR Gnangnon, and I Lawani
 Acquisition of data: FHR Gnangnon, EK Haag and HN Natta N'tcha
 Analysis and interpretation of data: FHR Gnangnon and I Lawani
 Drafting of manuscript: FHR Gnangnon and EK Haag
 Critical revision: FM Dossou and DK Méhinto.

Declaration of competing interest

All authors declare no conflicts of interest associated with this manuscript.

Acknowledgement

We would like to thank Mr. Gnangnon José and Mrs. Avougnansou Sylvia.

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