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Giant leiomyosarcoma: A case report

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ABSTRACT

INTRODUCTION: Uterine leiomyosarcoma is a rare uterine malignancy. Most of the patients lack symptoms or present with a rapidly enlarging pelvic mass.
PRESENTATION OF CASE: We report on a very large leiomyosarcoma in a woman presenting with a 3 months history of rapidly growing adominal mass and fatigue. Laparotomy was performed and diagnosis was confirmed by pathologic and histologic analysis. Patient refused chemotherapy after surgery and died from recurrence at 4th postoperative month.
DISCUSSION: Uterine leiomyosarcomas may follow a rapid clinical course with a doubling time of four weeks. There is no reliable method to distinguish uterine sarcoma from benign leiomyomas preoperatively.
CONCLUSION: This case represents the largest leiomyosarcoma reported in the literature.

1. Introduction

Leiomyosarcomas (LMS) are rare malignancies of uterus that account for only 1–2% of all uterine malignancies [1,2]. Most women with LMS lack symptoms or present with a rapidly enlarging pelvic mass [3]. In most cases, the diagnosis of leiomyosarcoma is made by pathological examination of hysterectomy or myomectomy specimen [4,5]. The incidence of LMS in a series of hysterectomies performed for presumed uterine leiomyomas is approximately 0.1–0.3% [6].

The cornerstone of the treatment for LMS is surgery. The resection of the localised disease by hysterectomy is regarded as a gold standard. Pelvic and para-aortic lymphadenectomy is not routinely indicated. The incidence of lymphatic spread is only about 3% in early-stage uterine LMS [4,7]. However, lymph-node involvement is often present in the advanced disease. Chemotherapy or pelvic radiation may be considered following surgery. However, whether any form of adjuvant therapy improves survival compared with observation is unknown. Although the majority of women have a uterus-limited disease at the time of diagnosis, patients are at a substantial risk for both local and distant recurrence of the disease.

Herein, we report a case of a leiomyosarcoma of the uterus weighed 57 kg with a diameter of 40 cm. To the best of our knowledge, this case presents the largest leiomyosarcoma reported in the literature.

2. Presentation of case

A 62-year-old, grand multiparous woman presented at our clinic with complaints of enlarged mass and fatigue for last 3 months. She had no significant medical or family history and she had used no medications. She has a body mass index (BMI) of 41.2 kg/m^2 (Fig. 1).

Physical examination revealed a huge, palpable, mass with restricted mobility in whole abdomen from symphysis pubis to up to the level of the processus xiphoideus, with associated tenderness. Laboratory studies showed an elevated cancer antigen 125 (CA-125) level of 557 U/ml. All other laboratory tests, including CA 19-9, CEA, AFP were within normal limits.

Transabdominal ultrasonography revealed a complex mass with solid areas, measuring more than 25 × 25 cm. Because of anxiety, MR or computered tomography (CT) imaging was not available.

The patient underwent surgery with the presumed diagnosis of an ovarian or uterin malignancy. Exploratory laparatomy revealed a huge uterine mass similar to that of a leiomyoma. A total hysterectomy and bilateral salpingooopherectomy were performed (Fig. 2).

Gross characteristics of the mass such as loss of the whorl pattern, homogeneous texture, yellow color and soft consistency felt different than a leiomyoma. Intraoperative frozen section analyses suggested of a high grade leiomyosarcoma. The surgery included peritoneal washing, omentectomy, systematic pelvic and paraaortic lymphadenectomy and peritoneal biopsies.

The removed uterus measured 68 × 55 × 33 cm and weighed 59 kg, while the mass measured 27 × 42 × 30 cm and weighed 57 kg (Fig. 3).

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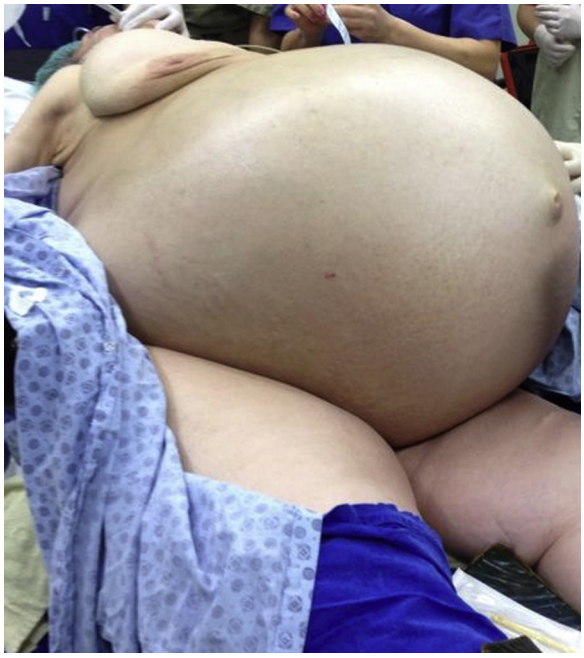

Fig. 1. Preoperative view of the patient in supine position.

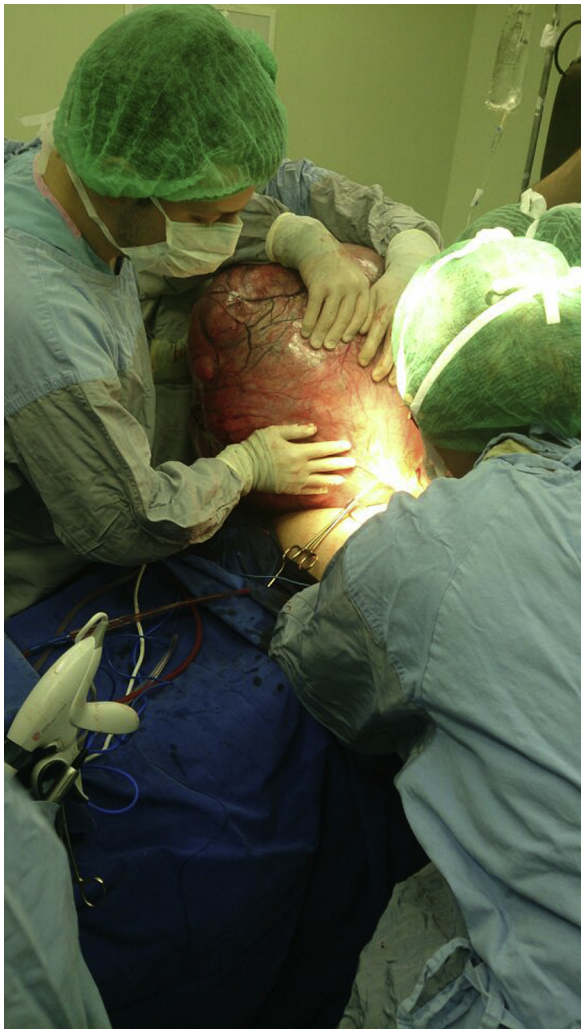

Fig. 2. Intraoperative view of the uterus.

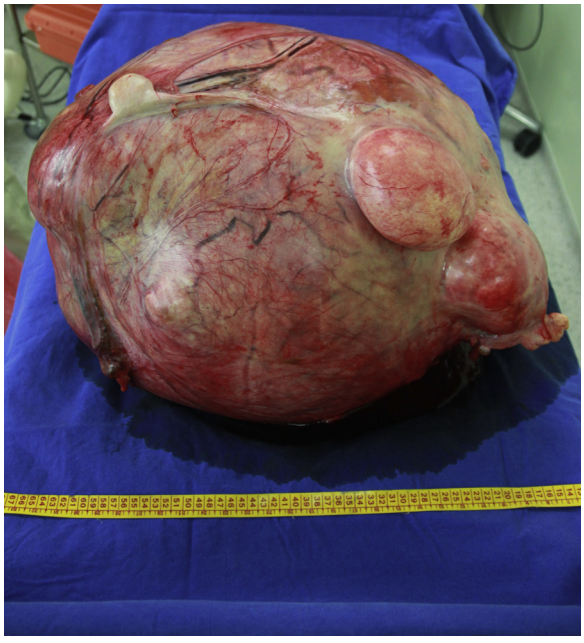

Fig. 3. Gross appearance of the tumor.

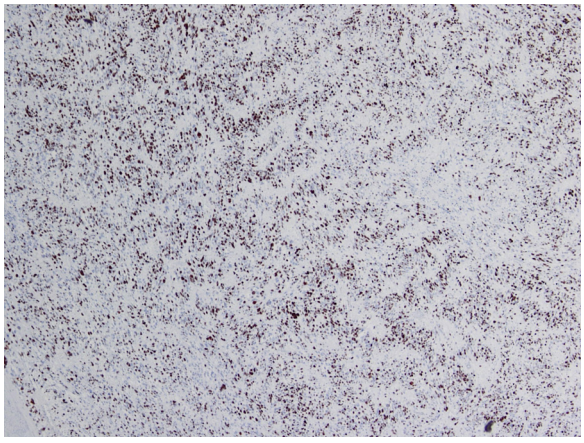

Fig. 4. Majority of the tumor cell nuclei stained with Ki67 proliferation marker (immunohistochemistry) × 100.

Postoperative histopathological evaluation showed large areas of necrosis and increased mitotic activity. Tumor cells were spindle shaped, pleomorphic and had moderate to severe atypia. There was no evidence of lymph node metastasis and peritoneal involvement. Immunohistochemical study with antibodies against p16, p53 and Ki-67 were performed and 64% of cells were positive for Ki-67 (Fig. 4).

Chemotherapy was recommended to patient, but she refused the treatment. Metastatic tumors appeared on intestinal serosa and abdominal wall after surgery. Her condition deteriorated rapidly and patient died from recurrence of LMS at 4th postoperative month.

Written informed consent was obtained from the patient shortly after the surgery for publication of this case report.

3. Discussion

Uterine LMSs are rare uterine malignancies [1]. However, the incidence of sarcoma is 1–2% in postmenopausal women [8]. LMS is an aggressive tumor associated with a high risk of recurrence and death, regardless of stage at presentation and differ

from other types of endometrial cancer [9]. Currently, there is no reliable method to differentiate uterine leiomyosarcoma from benign leiomyomas preoperatively. The diagnosis of leiomyosarcoma should be suspected when severe pelvic pain accompanies a pelvic tumor, in a postmenopausal woman particularly [9].

Uterine leiomyosarcomas may follow a rapid clinical course with a doubling time of four weeks [8]. Our patient had noticed the growth of abdomen which had been carrying 57 kg of sarcoma, just 3 months before the surgery.

LMS, like all other endometrial cancers, is surgically staged. Surgical staging should include a hysterectomy and a BSO with the resection of any visible metastatic disease. 60% of the women with LMS present with the disease limited to the uterus upon first diagnosis [5]. Cure rates of these patients range from 20 to 60%, depending on the success of the primary resection [7]. Several case series support the role of primary surgery [10,11]. Surgical cytoreduction is associated with progression-free survival (PFS); however, it is not associated with overall survival (OS). As such, the morbidity of surgery must be weighed against the improvement in PFS [12]. In the largest series involving 46 patients with LMS, a complete cytoreduction was significantly associated with disease-free survival ($p = 0.03$) [11]. Ovarian preservation can be considered in premenopausal patients with early-stage LMS of the uterus. In a study of 341 women less than 50 years old who were stage I or II LMS at diagnosis, no difference was found in the five-year disease-free survival between those who did and did not undergo a BSO [13]. We performed a BSO in this case as the patient had postmenapausal status.

However, sarcomas are aggressive tumors with a high risk of local and distant relapse even in completely resected tumours [2,14]. Patients with even International Federation of Gynecology and Obstetrics (FIGO) stages I and II LMS have a very high risk of recurrence. Survival after recurrence is poor. In one study, the 5-year survival rate for women with 1988 FIGO stage I LMS (tumour limited to the uterus) was only 51%, and for patients with stage II LMS (tumour in uterus and cervix), the 5-year survival rate was 25% [15]. The relapse rate is approximately 70% for stages I and II. The site of metastasis or recurrence is often distant due to the haematogenous spread into the lungs or liver [5].

Radiation therapy appears to have little benefit in the treatment of early-stage LMS. A retrospective review from the Surveillance, Epidemiology and End Results database of women with stage I/II LMS demonstrated no survival benefit from adjuvant radiation therapy [16]. There are few prospective data on the utility of chemotherapy for stage I/II LMS. A prospective study has demonstrated thet the combination chemotherapy of gemcitabine and docetaxel followed by doxorubicin offers a survival benefit to uterine leiomyosarcoma patients [17]. Our patient chose to reject chemotherapy despite our recommendation.

To the best of our knowledge, no case describing a similar size of leiomyosarcoma has been described in the English literature.

Conflict of interest

We declare that we have no conflict of interest.

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None.

Consent

Written informed consent form was obtained from the patient for publication of this case report.

Authors contribution

TS: Data collection, literature search.I.K.: Writing the paper, data analysis.B.M.: Data collection.I.Y.: Study concept and review.

Guarantor

Ilker Kahramanoglu.

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