Unexpected leiomyosarcoma after total abdominal hysterectomy with indication uterine myoma in nullipara: a case report

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ABSTRACT

Introduction: Uterine sarcomas are rare tumors, with 3-7% incidence. Abnormal uterine bleeding, abdominal or pelvic masses, and pain are the most common symptoms in the patient. Non-specific clinical symptoms or preoperative diagnostic techniques to differentiate uterine myoma and uterine sarcoma lead to the general diagnosis after surgery. The incidence of uterine sarcoma established from the literature was 0.09% -0.49% in women undergoing benign hysterectomy or myomectomy. The difficulty of diagnosing preoperative leiomyosarcoma is a challenge in itself. This case report aims to provide knowledge information with characteristics of a sonographic image with a non-hypervascularity encapsulated image and intraoperative findings with a regular smooth uterine surface, which is thought to be a uterine myoma is different from the anatomical pathology results, which indicate a leiomyosarcoma malignancy.

Case description: Miss A, 47 years old with complaints of an enlarged stomach for two years, felt abdominal pain and complained about prolonged menstruation for four years. The patient was unmarried and had no previous history of surgery. Intraoperatively, there was posterior corpus adhesion with ascending colon and adhesion was performed. Large, fragile blood vessels appeared. Then it was decided to do a total abdominal hysterectomy—Intraoperative hemorrhage 1200 cc. The results of the anatomic pathology showed a leiomyosarcoma. Furthermore, the patient was consulted to the oncology subdivision for a bilateral Salpingo-Oophorectomy procedure followed by chemotherapy. MRI, 3D Doppler sonography were required to increase the preoperative diagnosis of uterine sarcoma.

Conclusion: Our case reported that a patient was diagnosed with uterine myoma and stage II hypertension. The patient showed common signs and symptoms. There was posterior corpus adhesion with ascending colon, and adhesion was performed. Theoretically, pelvic sonography is a first-line imaging study. MRI can be used to help differentiate uterine sarcomas from uterine myomas.

Keywords: leiomyosarcoma, preoperative sarcoma score, total abdominal hysterectomy, uterine sarcoma.


INTRODUCTION

Uterine cancer is an invasive neoplasm of the body of the uterus and is the fourth leading cause of malignancy among women after breast, lung and colorectal cancer.1 Incidence of uterine sarcomas 3-7% of uterine malignancies and less than 1% of female genital organ malignancies.2 Among the 52,630 cases of uterine cancer that occurred in the United States in 2014, only 5-6% were classified as uterine sarcomas.3 The 5-year survival rate ranges from 30.5 to 68%.4

According to the WHO classification, soft tissue sarcomas consist of Adenosarcoma tumors, perivascular epithelioid cell tumors, primitive neuroectodermal tumors, rhabdomyosarcoma, malignant types (Pecoma), angiosarcoma, neurogenic sarcomas, osteosarcoma, chondrosarcoma, liposarcoma, soft tissue sarcomas, and myxofibrosarcoma.2

Histologically, uterine sarcomas are grouped into endometrial stromal sarcomas or ESS (10-15%), carcinosarcoma (40%), leiomyosarcoma or LMS (40%), and undifferentiated sarcomas or USS (5-10%).5 The risk factors for the occurrence are postmenopausal women, obesity, nulliparous, and hypertension who develop endometrial carcinoma.1

Sarcoma of the uterine is a heterogeneous rare tumor. It has an aggressive clinical development and has the worst clinical outcome.6 Uterine sarcomas exist in women aged 40-60. There are several common signs and symptoms, such as abnormal uterine bleeding, abdominal or pelvic mass and pain. The most commonly reported prognostic factors, tumor stage, histological subtype, grade, lymphatic invasion, would include menopausal status and adjuvant radiation therapy.7

This case report aims to provide knowledge information with
characteristics of a sonographic image with a non-hypervascularity encapsulated image and intraoperative findings with a stable smooth uterine surface, which is thought to be a uterine myoma is different from the anatomical pathology results which indicate a leiomyosarcoma malignancy. Currently, diagnosing a leiomyosarcoma by preoperative is a challenge.

**CASE DESCRIPTION**

Miss A, 47 years old with an enlarged stomach complaining for two years, felt abdominal pain and complained about prolonged menstruation for four years. Defecate and urinate within normal limits. The patient was unmarried and had no previous surgical history. The patient has high blood pressure and has been taking the drug amlodipine since 2015. On physical examination, found a vital sign, found blood pressure 160/90 mmHg, pulse 85x / minute, respiration 20x / minute, temperature 36.4°C generalized status within normal limits. On gynecological examination of the bimanual uterus, it was found that the uterus was enlarged as high as two fingers above the center with a flat surface, solid consistency and mobile.

Laboratory finding was normal limits. Ultrasonography showed a hyperechoic mass with a size of 17x16x18 cm with positive arterial feeding suggesting myoma uterine, and both adnexes were within normal limits. The patient was diagnosed with uterine myoma and stage II hypertension. Intraoperatively, there was posterior corpus adhesion with ascending colon and adhesion was performed. Large, fragile blood vessels appeared. Then it was decided to do a total abdominal hysterectomy—intraoperative hemorrhage 1200 cc. The results of the anatomic pathology showed a leiomyosarcoma. Moreover, the patient was consulted to the oncology subdivision for a bilateral salpingo-oophorectomy using a total abdominal hysterectomy procedure (Figure 1) procedure followed by chemotherapy.

**DISCUSSION**

**Epidemiology**

Uterine sarcomas originate from the myometrium or uterine connective tissue with a 3–7% incidence of diagnosed uterine carcinoma in America. Uterine sarcoma is a rare tumor with a very malignant nature. The incidence of uterine sarcoma is more common at a younger age than the incidence of endometroid carcinoma. Several studies have shown a higher incidence of sarcoma in black women as opposed to the overall endometrial carcinoma. In a cohort study, progestin and estradiol therapy in menopause was associated with an increased risk of leiomyosarcoma and endometrial stromal sarcoma. The risk factors for diabetes and the risk of developing uterine sarcoma have been reported in one study (Brinton et al., 2005). The general pathogenesis of the uterine sarcoma is unknown. Basic and preclinical knowledge provides a better understanding of tumor biology. Leiomyosarcoma is a malignant neoplasm similar to HGGESS, mainly characterized by pleomorphic spindle cells in fascicular growth.

**Classification**

The World Health Organization (WHO) was divided uterine sarcoma into two types, such as:
(1) tumor of mesenchymal and also mixed epithelial
(2) tumors malignant mesenchymal

According to all uterine sarcoma types, the most common type of uterine sarcoma is leiomyosarcoma (60% - 70%), and the others were LG-ESS, HG-ESS and UUS.

**Clinical Symptoms**

The clinical symptoms of uterine sarcoma are similar to uterine myomas with typical uterine bleeding and lower abdominal pain. The clinical features of uterine sarcoma are different for histological subtypes: enlargement of uterine mass or abdominal pain in LMS and abnormal characteristics of a sonographic image with a non-hypervascularity encapsulated image and intraoperative findings with a stable smooth uterine surface, which is thought to be a uterine myoma is different from the anatomical pathology results which indicate a leiomyosarcoma malignancy. Currently, diagnosing a leiomyosarcoma by preoperative is a challenge.
Patients with known uterine sarcoma are advised to undergo a Thoracic Ct scan to determine metastases and further integrate disease progression.

**Magnetic resonance imaging (MRI)**
Magnetic resonance imaging could be used to differentiate uterine sarcoma from uterine myoma. A degenerative uterine myoma found on MRI may be considered a uterine sarcoma. MRI has an essential role for preoperative diagnosis and for determining appropriate management.

When pathological results are available, the patient may be subject to an MRI examination and be seen by a pathologist, but the preoperative diagnosis of MRI of the uterine sarcoma cannot be made quickly. In these patients, further management should be considered. Brohl et al. Found a variation in the risk of UUSs in several age groups, the risk of uterine sarcoma from 10.1 / 1000 in patients 75-79 years, at 30 years, the risk was 1 case per 500 patients. In another study found that UUSs frequently appeared at the age of 40-49 years. Abnormal uterine bleeding is a common symptom. The rapid growth of the tumor mass is an indication of uterine sarcoma. UUSs ultrasonography examination shows an atypical tumor size and border picture, but UUS often misdiagnoses a uterine myoma. Abundant blood flow around the pelvic mass is vital to note.

In about 40% of cases, the MRI diagnosis is degenerative uterine fibroids, which may very well indicate uterine sarcoma. Signs that exist in MRI examination could direct us to the appropriate diagnosis. When a pathology report is available, the pathologist reviews the MRI; however, it was still challenging to make a preoperative MRI diagnosis of uterine sarcoma.

**Management**
Standard management for early-stage of uterine sarcoma is total abdominal hysterectomy and bilateral salpingo-oophorectomy. Primary treatment for this case, such as operative (total hysterectomy and bilateral salpingo-oophorectomy). It is a crucial factor for managing all types of uterine sarcomas; however, lymphadenectomy and surgery with the concept of cytoreductive are often done at an advanced stage.

When suspected a uterine sarcoma intraoperatively, thus a frozen section should be done. The findings at the time of surgery and the anatomic pathology of the tissue from frozen sections should be interpreted with caution, especially for women who wish to preserve fertility. Sarcoma uterus is not suitable for doing the surgery of fertility-sparing.

When adjuvant therapy is deemed necessary for high-grade ESS or undifferentiated uterine sarcoma, chemotherapy may be recommended. Doxorubicin is still the first-line treatment in uterine sarcoma. Avoid uterine morcellation in cases of suspected leiomyoma. The morcellation procedure destroys the uterus into 18 small pieces to make the patient's surgery heal faster. However, if there is an LMS (leiomyosarcoma), this procedure can spread malignant cells throughout the abdominal cavity and shorten overall survival.

An advanced condition needs a compound treatment. The palliative treatment approach depends on the presence or absence of symptoms and the potential for treatment toxicity. However, disease stabilization is an equally good goal without significant symptoms to prolong a good quality of life.

**Prognosis dan Follow-Up**
The French Federation of Anticancer Centers (FNCLCC) was already developing a scoring system to assess soft tissue sarcomas through histological evaluation of tissues such as tumor differentiation, mitotic count, and necrosis. Prognosis after primary care can be predicted with a good nomogram according to the histological finding, depth, tumor size, and the patient's age. Patients with tumor characteristics such as having a high grade (grade 3), advanced stage, high mitotic index and age above 65 years had a worse outcome. Prognosis Women with a history of metastases have a 2-year survival rate of about 30%. Unfortunately, the majority (50%) of patients with high-grade sarcoma present with distant metastases. It has 12 months median survival rate since the time the diagnosis of metastases is made. Screened is needed every three or four years.

**Ultrasound**
For women with a suspected uterine tumor, pelvic ultrasound is the most important diagnostic method used to evaluate the uterus. Trans-vaginal and doppler ultrasounds allow assessment of the uterus and endometrial vascularization. This technique evaluates the uterus, myometrium, endometrial blood vessels and the three blood vessels that supply the uterus. This technique aims to distinguish benign or malignant from endometrial disease, but several studies have yielded controversial results. We can see the uterine sarcoma with irregular walls, not encapsulated, hypo-hyperechoic features with increased peripheral and central vascular doppler.

**Computed tomography**
Computed tomography is used for imaging. This examination is suitable for determining staging and metastasis. Patients with known uterine sarcoma are advised to undergo a Thoracic Ct scan to determine metastases and further integrate disease progression.

**Diagnosis**
The preoperative diagnosis of uterine sarcoma remains very challenging. In a recent cohort study from 2000-2012 with women diagnosed with uterine leiomyosarcoma (UTLMS), 52.4% of malignancies were not diagnosed during surgery. Increasing age is a risk factor for the incidence of ULSMs (uterine leiomyosarcoma) around 9.8 / 10,000 aged 25-39 years and increased in the group of women aged 50-64 years to 33.4 / 10,000 who previously underwent laparoscopic hysterectomy with suspected uterine myoma.

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months in the first two to three years according to ESMO guidelines for patients with moderate/high-grade tumors, after that twice a year until the fifth year. And once a year after that. Another recommendation is that patients with low-grade tumors are followed up every 4-6 months for 3-5 years, then annually after that, for at least ten years. In patients with low-grade sarcomas in which the risk of local recurrence is the primary reason for follow-up.3,8

CONCLUSION
Our case reported that a patient was diagnosed with uterine myoma and stage II hypertension. The patient showed common signs and symptoms. There was posterior corpus adhesion with ascending colon, and adhesion was performed. According to the patient's condition, we decided to do a total abdominal hysterectomy. Theoretically, pelvic sonography is a first-line imaging study. MRI can be beneficial to differentiate uterine sarcomas from uterine myomas.

DISCLOSURE
Conflict Of Interest
There is no conflict of interest.

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Author Contribution
As a specialist in obstetrics and gynecology consultant, CMY is the woman in charge of Investigating the draft case report and final editing in drafting the manuscript. CRM as an obstetrician and gynecologist is also a candidate the consultant prepares the design, edits and reviews the manuscript. HS is a resident doctor in obstetrics and gynecology, preparing literature, clinical studies and editing manuscripts. All authors discussed the results and commented on the manuscript.

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