Clinical and Experimental Obstetrics & Gynecology

Case Report

Uterine leiomyosarcoma presenting as septic shock in a 43-year-old woman: a case report

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Introduction: Sepsis or septic shock, a life-threatening condition, is rarely reported as the initial manifestation of uterine malignancy. *Case report*: A 43-year-old woman consulted our hospital for pyrexia, anorexia, dyspnea, and a markedly distended abdomen. Computed tomography images showed a large abdominal tumor occupying almost the entire abdominal cavity with an air-fluid level and multiple masses in both lungs. Pelvic examination revealed necrotic tissues with blood, prolapsing from the uterus, and having a putrid odor. Her vital signs indicated a systemic inflammatory response syndrome. Antibiotic, anti-shock, and anti-disseminated intravascular coagulation therapies were administrated, but the patient died of multiple organ failure at 22 hours after admission. Autopsy and histopathological examination revealed that the giant abdominal mass was a uterine leiomyosarcoma, in which the myometrium was extensively necrotic with a putrid odor. The patient's blood culture was positive for *Escherichia coli*, and sepsis and septic shock due to intrauterine infection of the uterine leiomyosarcoma was considered as the cause of death. *Conclusions*: Physicians should be aware that uterine malignancy could present as sepsis in women with an abdominal mass.

Keywords

Sepsis; Septic shock; Leiomyosarcoma; Systemic inflammatory response syndrome

1. Introduction

Uterine leiomyosarcoma is a rare disease, which accounts for approximately 1% of female genital tract cancers [1]. It has poor clinical outcomes because it is not diagnosed in the early stage, and no effective treatments have been established [2]. Patients with uterine leiomyosarcoma present with nonspecific symptoms, such as abdominal distention, abdominal pain, and atypical genital bleeding [3].

Sepsis and subsequent septic shock, which are induced by the immune response of the host to a bacterial, viral, and/or fungal infection, are life-threatening [4]. They cause systemic inflammatory response syndrome (SIRS), which could eventually lead to multiple organ dysfunction syndrome [4]. Although sepsis or septic shock has been reported to be present in patients with uterine sarcoma [5, 6], there has been hitherto no reported case, to our knowledge, in which septic shock was the initial manifestation of uterine sarcoma. The present patient did not show uterine bleeding but abruptly showed septic shock. We report herein a case of uterine leiomyosarcoma with multiple lung metastases presenting as a septic shock with a rapid deterioration in the patient's condition, leading to death.

2. Case report

A 43-year-old nulligravida woman had pyrexia, anorexia, and dyspnea for 2 weeks. Although the patient had been taking over-the-counter drugs for common colds, the symptoms did not improve. She also had noticed that her abdomen was markedly distended over the past few months. She did not undergo consultation and hardly left home for the last 2 years due to depression. She presented herself to the emergency department due to worsening dyspnea with concomitant pyrexia and anorexia. Physical examination revealed a distended abdomen with a huge mass extending from the pubic region to the xiphoid process. The abdominal circumference was 103 cm, and she had no complaints of abdominal pain or tenderness. She was referred to our gynecologist and was admitted for further examination. Her blood pressure was 138/70 mmHg, heart rate was 158 bpm, respiratory rate was 44 breaths/min, and body temperature was 39.3 °C. She had a pale face, an altered level of consciousness with a Glasgow coma scale score of 12/15, and had profuse sweating with chills. The internal pelvic examination was unremarkable due to the presence of the large pelvic mass, but it revealed necrotic tissues with blood and a putrid odor caused by a prolapsed uterus. The results of the blood examination were as follows: WBC count 8820/μL, hemoglobin 6.0 g/dL, hematocrit 20.2%, and platelet count $3.2 \times 10^4/\mu$L. Serum biochemical analysis showed that the level of C-reactive protein was 31.9 mg/dL (reference, $<$ 0.30 mg/dL), and the value of lactate dehydrogenase was 3329 U/L (reference, 115-245 U/L). Several tumor markers have been performed and their values were as followed: carbohydrate antigen 19-9, CA-125, sialyl Lewis X, and squamous cell carcinoma antigen were 144 U/mL (reference, $<$ 37.0 U/mL), 291 U/mL (reference, $<$ 35.0 U/mL), 51 U/mL (reference, $<$ 38.0 U/mL), and 4.5 ng/mL (reference, $<$ 1.5 ng/mL), respectively. Blood exam-

ination showed a prothrombin activity of 34.5% (reference, 80%-100%) and a serum fibrinogen degradation product level of 44.0 μg/mL (reference, $<$ 4.0 μg/mL). Blood gas analysis revealed a metabolic acidosis with a pH of 7.28, $PaCO_2$ of 22 mmHg, PaO_2 of 22 mmHg, and a base excess of -14.9 mmol/L. A chest X-ray image showed multiple nodules in both the lung fields (Fig. 1). Computed tomography (CT) showed a massive uterine tumor with a maximum diameter of 27 cm with a heterogeneous surface, enhanced to the same extent as the myometrium by contrast media. The tumor had an air-fluid level content, suggesting the presence of a generated gas inside the tumor. CT also showed a liver cyst with a 10-cm diameter and a splenomegaly with a moderate amount of ascites. The physical and blood examination findings were indicative of SIRS. The diagnosis was sepsis with disseminated intravascular coagulation (DIC), probably due to an intrauterine infection of the malignant uterine tumor. She was immediately administrated antibiotic and anticoagulant therapies for sepsis and DIC. The patients' heart rate did not decrease to $<$ 100/min, and her respiratory rate was $>$ 30/min during the treatment. The blood examination results showed a decrease in the platelet count to $0.6 \times 10^4/\mu$L, suggesting progression of DIC. She died of multiple organ failure at 22 hours after admission.

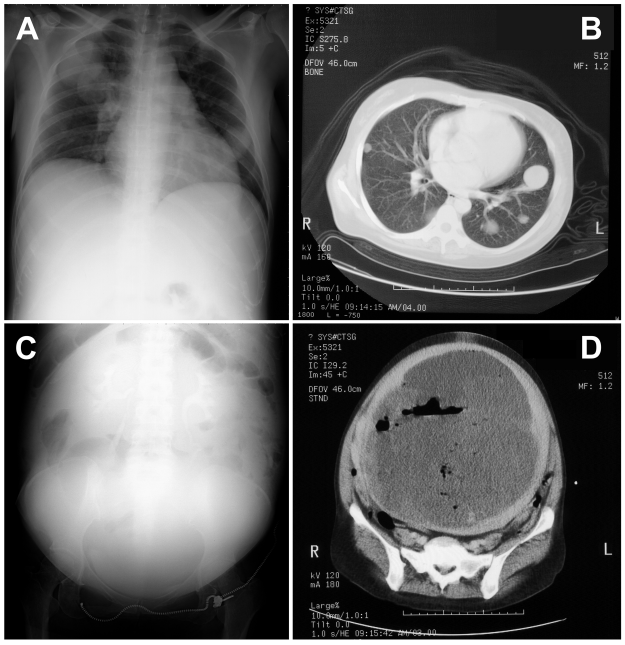

Fig. 1. Imaging findings on admission. Chest X-p (A) and computed tomography (CT) (B) images show multiple lung metastases. Abdominal X-p shows an extensive abdominal tumor (C). CT shows an intra-abdominal tumor with an air-fluid level (D).

Autopsy findings revealed that the abdominal mass was a markedly enlarged uterus weighing 8000 g (Fig. 2). There was no invasion or adhesion in the pelvic cavity. The myometrium was markedly thickened and extensively necrotic

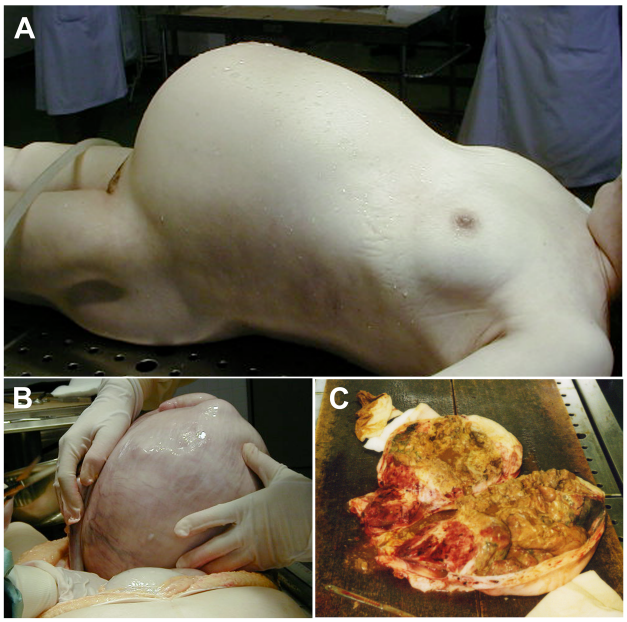

Fig. 2. Pictures of the autopsy. The patient had abdominal distension from the pubic region to the xiphoid process (A). Laparotomy findings of the enlarged uterus (B). In the extirpated uterus, a large amount of pus was found in the uterine cavity and hemorrhage and necrotic tissues were found in the myometrium (C).

with a putrid odor. The multiple lung masses could not be resected due to the family's request. Histopathological examination revealed spindle-shaped cells with nuclear pleomorphism and prominent nucleoli, which had $>$ 10 mitoses per 10 high power field. These cells stained positive for α-smooth muscle actin and HHF35, which is suggestive of a leiomyosarcoma of the uterus. Later, the blood culture was positive for *Escherichia coli*, and sepsis due to the intrauterine infection of the leiomyosarcoma was considered as the cause of death.

3. Discussions

We present a case of uterine leiomyosarcoma presenting as sepsis with a rapid deterioration in the patient's condition, leading to death. We first report a case of uterine leiomyosarcoma presenting as septic shock.

Uterine sarcoma, including leiomyosarcoma, is a rare disease that accounts for 3%-5% of all cancers of the uterine corpus [3, 7]. It progresses rapidly with enlargement of the tumor and metastasis to the lymph nodes, liver, and lungs, and has a poor prognosis [8, 9]. Vaginal bleeding, lower abdominal pain, and rapid uterine enlargement are the three major signs of the disease [3]. In patients presenting with these symptoms accompanied by a palpable enlarged uterus, the clinical diagnosis is often uterine fibroids. Therefore, when a rapid uterine enlargement is observed, uterine sarcomas should be considered in the diagnosis.

Only two cases of uterine sarcoma that led to sepsis or septic shock have been reported [5, 6]. The cause in both

cases was an intrauterine endometrial infection caused by an ascending *Clostridium perfringens* infection, and the diagnosis after hysterectomy was undifferentiated uterine sarcoma in the advanced stage. These patients had genital bleeding as initial symptoms. The present case is the first report of leiomyosarcoma presenting with septic shock without genital bleeding.

Sepsis is a life-threatening organ dysfunction caused by a dysregulated host response to infection [10], and an ascending intrauterine infection causes sepsis in the pelvis. Given that the uterine cavity opens externally, an ascending infection is likely to occur, and when the infection is severe, it progresses from endometritis to myometritis. Large fibroids are prone to degeneration due to inadequate blood flow within the tumor. In addition, uterine artery embolization for the treatment of fibroids also causes necrosis of the fibroid. It has been reported that the ascending infection of these degenerated and necrotic fibroids can result in life-threatening or fatal sepsis, or even septic shock [4, 11–13]. Given that leiomyosarcomas are larger than the other uterine sarcomas, they are prone to hemorrhage and necrosis [14], which might result in endometritis-myometritis due to the ascending infection, and sepsis due to the myometritis.

Given that severe sepsis has a poor prognosis, treatment should be initiated as soon as sepsis is suspected. SIRS is a systemic inflammatory response triggered by various causes, including infection, trauma, pancreatitis, and burns. SIRS is diagnosed when ≥ 2 of the following four parameters showed abnormal findings: body temperature, heart rate, respiratory rate (or $PaCO_2$), and peripheral WBC count. Our case had abnormalities in three out of these four parameters, excluding WBC count, thereby meeting the diagnostic criteria for SIRS and suggesting increased inflammatory cytokines due to sepsis. The quick sequential (sepsis-related) Organ Failure Assessment score (qSOFA), consisting of respiratory rate, level of consciousness, and systolic blood pressure, is a useful tool for diagnosing early sepsis. A qSOFA score of ≥ 1 indicates a poor prognosis for sepsis. In our case, a slight alteration in the level of consciousness was observed with a respiratory rate of ≥ 40 breaths/min, but the systolic blood pressure was maintained at ≥ 100 mmHg, indicating a qSOFA score of 2. The diagnostic criteria of SIRS and qSOFA can help in the diagnosis of sepsis at the bedside, by simply performing blood tests; thus, treatment for sepsis can be immediately initiated.

In conclusion, in this case, the condition was very severe already at the time of admission, and thus there may have been no, or at least a few chances to save this patient's life. However, physicians should be aware that, even without genital bleeding, uterine malignancy should be listed as a differential diagnosis of unexplained septic shock: septic shock can be an initial manifestation of uterine sarcoma.

Author contributions

TT designed the study and wrote the manuscript. MM and KO collected the data. HM revised the manuscript. All authors read and approved the final manuscript.

Ethics approval and consent to participate

According to our institutional policies, this case report is exempted from obtaining an Institutional Review Board approval. We would like to thank the patient's family for providing permission to report this case.

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Conflict of interest

The authors declare no competing interests.

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