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Case Report

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Multifocal Ovarian Leiomyosarcoma Presenting With Ambiguous Imaging and Rapid Progression: A Diagnostic Dilemma

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Abstract

Uterine sarcomas are rare, aggressive tumors with high recurrence rates and nonspecific presentations, making preoperative diagnosis difficult. Leiomyosarcoma with ovarian involvement is particularly uncommon and can mimic primary ovarian malignancy. A postmenopausal woman presented with progressive abdominal pain and unsteady gait. Examination revealed abdominal distension and tenderness. Imaging demonstrated a large heterogeneous pelvic mass with pelvic free fluid and bilateral inguinal lymphadenopathy, raising suspicion for ovarian malignancy. Laboratory testing revealed elevated cancer antigen 125 (CA-125). Imaging was inconclusive regarding the primary origin. The patient underwent exploratory laparotomy with total abdominal hysterectomy, bilateral salpingo-oophorectomy, tumor debulking, and cholecystectomy. Histopathology confirmed multifocal high-grade leiomyosarcoma involving both ovaries, fallopian tube, mesosalpinx, and pelvic sidewall implant, with positive margins. No uterine histological involvement was noted. Surveillance imaging revealed rapid recurrence with peritoneal and nodal metastases. The patient commenced single-agent doxorubicin chemotherapy. This case highlights the diagnostic ambiguity, aggressive course, and need for early multidisciplinary management in leiomyosarcoma.

Keywords: Uterine leiomyosarcoma; Ovarian involvement; Pelvic mass; Diagnostic ambiguity; Multidisciplinary management; Recurrence and metastasis; Exploratory laparotomy; Doxorubicin chemotherapy

Introduction

Uterine sarcoma is a rare gynecologic malignancy developed from uterine mesenchymal tissues of the myometrium and endometrial stroma [1]. It is a notorious malignancy with a high probability for recurrence, poor prognosis, and ambiguous pathway to diagnosis. Risk factors include pelvic radiation, rare cases of tamoxifen use, and hereditary cancer syndromes such as hereditary leiomyomatosis and renal cell cancer syndrome [1–3]. However, further investigation identifies obesity and diabetes as further risk factors for development [3]. Uterine sarcoma is an umbrella term that houses many clinical subtypes with an ambiguous pathway for diagnosis. The prevalent types are leiomyosarcoma, endometrial stromal sarcoma, and undifferentiated uterine sarcoma. The diagnosis is made by histopathological examination following operative management [1]. Our patient has stage IIB ovarian leiomyosarcoma. This case report offers a comprehensive analysis of the disease, emphasizing the importance of prompt detection and interdisciplinary teamwork. We highlight the clinical presentation, radiographic imaging, and management of this condition.

Case Report

Investigations

A postmenopausal woman, presented to our emergency department for abdominal pain and unsteady gait. She reported that the abdominal discomfort had progressed over several weeks, contributing to difficulty with ambulation and generalized weakness. She denied vaginal bleeding, gastrointestinal symptoms such as nausea, vomiting, or changes in bowel habits, and had no urinary complaints. Her medical history was noncontributory, and there was no known personal or family history of gynecologic malignancy. On examination, the patient was hemodynamically stable but demonstrated abdominal distension and tenderness. A large pelvic mass was suspected on palpation, prompting further workup. Laboratory evaluation was significant for elevated cancer antigen 125 (CA-125) at 453. Initial pelvic imaging revealed a large, heterogenous pelvic mass measuring 18.5 × 12.1 × 15.9 cm. The origin of the mass was unclear, with differential diagnosis including uterine leiomyosarcoma versus ovarian malignancy, as normal ovaries were not visualized. Nonspecific free fluid in the pelvis and bilateral inguinal lymphadenopathy was also noted.

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Clinical presentation and differential diagnosis

The patient initially presented with abdominal pain, unsteady gait, and a large heterogenous pelvic mass, findings commonly seen in ovarian leiomyosarcoma and other gynecologic sarcomas in postmenopausal women. Several gynecologic and non-gynecologic etiologies were considered, including: 1) primary ovarian malignancy, which was strongly suspected given the patient's postmenopausal status, elevated CA-125 level, bilateral inguinal lymphadenopathy, and inability to visualize normal ovaries on imaging; 2) uterine leiomyosarcoma, considered given the size, heterogenous nature, and rapid evolution of the mass; 3) a benign ovarian mass, which was possible but less likely due to the aggressive features and the presence of lymphadenopathy; 4) metastatic disease from another primary site, which was less suspected as no extra-pelvic primary malignancy was identified on imagining; 5) a degenerating uterine fibroid, which can present with heterogenous signals on imaging, but the patient's rapid growth and age were atypical.

Treatment

She underwent an exploratory laparotomy with total abdominal hysterectomy, bilateral salpingo-oophorectomy, tumor debulking, and cholecystectomy. Pathology confirmed leiomyosarcoma involving both ovaries, the fallopian tube, the left mesosalpinx, and pelvic sidewall implant, with tumor at the surgical margins. The gallbladder showed benign pathology.

Follow-up and outcomes

Postoperative positron emission tomography (PET) scan showed no metabolically active distant diseases, and the University of Michigan tumor board concluded that there was no measurable residual or metastatic disease; however, adjuvant chemotherapy was recommended due to positive margins and suspicious pelvic fluid. Follow-up computed tomography (CT) imaging raised concerns for recurrent disease near the hysterectomy bed, and magnetic resonance imaging (MRI) confirmed progression of peritoneal and pelvic nodal metastases. Given the progression, the patient agreed to begin systemic chemotherapy with single-agent doxorubicin (60 mg/m^2 every 21 days).

Discussion

Uterine sarcomas are uncommon but highly malignant neoplasms. Due to their nonspecific symptoms and histologic differences, they often pose diagnostic and therapeutic challenges, making treatment difficult. Our patient's presentation with insidious abdominal pain and ataxic gait, without vaginal bleeding or gastrointestinal (GI)/urinary symptoms, is typical of the elusive symptomatology of these tumors. While uterine leiomyosarcoma represents about 1% of all uterine malignancies, the cancer is usually not diagnosed until after a laparoscopy or operation. This is because preoperative imaging studies and serum markers are not able to differentiate the malignancy from benign fibroids during the preoperative stage of diagnosis [2, 4].

Despite advancements in diagnostic imaging, including the MRI or PET-CT, preoperative diagnosis of uterine sarcomas continues to be sufficiently inaccurate, underscoring the duality of diagnosis in surgical intervention and confirmation by histopathology [5, 6]. Tumor markers such as CA-125 and CA19-9 may be elevated in a variety of gynecologic conditions, including benign adnexal masses and malignant tumors, thereby limiting their specificity in preoperative risk stratification. Recent evidence suggests that while these markers can support clinical suspicion when interpreted alongside imaging and symptomatology, they should not be used in isolation to distinguish benign from malignant gynecologic disease [7].

Leiomyosarcomas are well known for their hematogenous spread and high local recurrence, even when identified early in the disease course [8, 9]. The use of adjuvant chemotherapy in uterine sarcomas is controversial because of the limited randomized trial data; however, in the treatment of high-grade neoplasms or advanced-stage cancers, multimodal therapy is often pursued [10].

This case underscores the aggressive nature and diagnostic ambiguity of uterine sarcomas, particularly leiomyosarcoma with ovarian involvement. Despite surgical resection and histopathologic confirmation, the patient experienced rapid disease progression, emphasizing the tumor's resistance to conventional therapy. The decision to pursue adjuvant chemotherapy reflects the growing reliance on multimodal treatment strategies in advanced or high-risk cases.

Primary ovarian leiomyosarcomas are rare malignant neoplasms, accounting for a small fraction of ovarian tumors, with fewer than 100 cases reported in the literature to date [11]. Most reported cases occur in postmenopausal women and present with nonspecific abdominal or pelvic symptoms, including pain, distension, or a palpable mass, frequently resulting in delayed diagnosis [11]. Published case reports consistently describe tumors that are large at the time of detection, often exceeding 10 cm, and characterized histologically by high-grade spindle cell morphology, significant nuclear atypia, elevated mitotic activity, and areas of tumor necrosis [11]. These neoplasms demonstrate an aggressive biological behavior, with a strong propensity for hematogenous dissemination and early recurrence, contributing to poor overall survival outcomes [12]. Surgical resection remains the primary treatment modality across reported cases, typically involving total hysterectomy with bilateral salpingo-oophorectomy, although optimal surgical staging is not standardized due to the rarity of the disease [11, 13]. The role of adjuvant chemotherapy or radiotherapy remains controversial, with inconsistent survival benefit reported in the literature and treatment strategies often extrapolated from uterine leiomyosarcoma protocols [12]. Compared with previously published cases, the present case demonstrates a similarly aggressive clinical course, with rapid disease progression despite surgical intervention, underscoring the limited efficacy of current therapeutic options and the need for more effective systemic treatment strategies [13]. Ultimately, this report highlights the urgent need for improved

diagnostic tools, evidence-based treatment protocols, and continued surveillance to optimize outcomes in patients with uterine sarcoma.

Patient perspective

"At first it was scary to understand what was happening to me. Hearing I had a rare cancer— and that it came back so fast— was overwhelming. But my doctors took the time to make sure I understood, and that made me feel supported through the surgery and now the chemotherapy."

Learning points

Uterine leiomyosarcoma can present with nonspecific symptoms and mimic primary ovarian malignancy, especially with ovarian involvement. Preoperative imaging and serum markers often fail to distinguish uterine sarcoma from benign or other malignant pelvic masses. Even after complete macroscopic resection, high-grade leiomyosarcomas carry a high risk of rapid recurrence. Multidisciplinary management is essential for optimizing treatment strategies. Continued surveillance and research into effective systemic therapies are critical due to the tumor's aggressive biology.

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Financial Disclosure

None to declare.

Conflict of Interest

The authors report no conflicts of interest.

Informed Consent

No identifiable patient information is included in this work, so informed consent was not required.

Author Contributions

AR, MH, and DK contributed to patient management, data collection, interpretation of clinical findings, and drafting of the manuscript. AR participated in data analysis and literature review but was not directly involved in patient care. AR, MH, and DK supervised the work and critically revised the manuscript. All authors read and approved the final version and agree to be accountable for all aspects of the work.

Data Availability

The authors declare that data supporting the findings of this study are available within the article.

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