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Spontaneous Uterine Perforation Secondary to Uterine Carcinosarcoma Presenting as Acute Abdomen with Pneumoperitoneum

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Abstract

Uterine carcinosarcoma (UCS) is a rare, aggressive malignancy with high-grade histological features of both endometrial carcinoma and sarcoma. UCS accounts for less than 5% of all uterine cancers, and typically presents with postmenopausal vaginal bleeding, pelvic pain, or vaginal discharge. Spontaneous uterine perforation is even rarer. In this case, presence of fluid in the uterine cavity and the multiple leiomyomas were suggestive of a possible uterine source for the intraperitoneal air/fluid interface. Providers need to be aware that UCS can present without gynecological symptoms, as in this case.

Keywords: Leiomyosarcoma; Carcinosarcoma; Uterine perforation; Pneumoperitoneum; Acute abdomen; Uterine cancer

Abbreviations: UCS: Uterine carcinosarcoma; BP: Blood Pressure; HR: Heart Rate; RR: Respiratory Rate; T: Temperature; CT: Computed Tomography

Introduction

Uterine carcinosarcoma (UCS) is a rare, aggressive malignancy with high-grade histological features of both endometrial carcinoma and sarcoma [1]. UCS accounts for less than 5% of all uterine cancers, with reviews estimating the incidence of UCS at 1-4 per 100,000 women aged >35 in the United States [2,3]. UCS typically presents with postmenopausal vaginal bleeding, pelvic pain, or

vaginal discharge (10), with up to one-third of all women presenting with extrauterine spread [1]. Factors influencing both overall and recurrence-free survival rates include tumor stage, advanced age, and the presence of a rhabdomyosarcoma component [4].

Spontaneous uterine perforation is even more rare; risk factors include prior uterine surgery, anatomic abnormalities, and

trauma. Defined as a perforation that is not associated with labor or childbirth, spontaneous uterine perforation can be recognized by signs of internal hemorrhage or sepsis, abdominal pain, or vaginal bleeding. Pneumoperitoneum identified on imaging should be treated emergently, and quick recognition of the signs and symptoms combined with evidence from imaging can be lifesaving.

To the best of our knowledge, there are four reported cases in the literature of uterine sarcoma presenting with signs of visceral perforation in the past decade [5-7], with this being the second to present without gynecological symptoms [8].

Case Presentation

Presentation

The patient was a 69-year-old G4P3013 African-American female who presented to the emergency department with a 4-day history of progressively worsening abdominal pain associated with nausea, vomiting, and constipation. She had a past medical history that included uterine fibroids, deep vein thrombosis, hypertension, and hyperlipidemia, in addition to a past surgical history of tubal ligation and one cesarean section (Figure 1).



Figure 1: Non-contrast CT of the abdomen/pelvis showing enlarged, globular uterus with irregular structure.

A physical exam revealed a soft and distended abdomen with diffuse tenderness in all four quadrants and involuntary guarding as well as hypoactive bowel sounds. Vital signs were notable for tachycardia and hypotension. The rectal examination was unremarkable.

Two hours after her initial presentation, the patient developed diaphoresis, worsening abdominal pain, and hypotension. At that time, vital signs were as follows: blood pressure (BP): 103/56 mmHg, heart rate (HR): 98 bpm, respiratory rate (RR): 34/min, SO₂: 100%, and temperature (T): 36.7 °C.

Investigations

A non-contrast computed tomography (CT) of the chest, abdomen, and pelvis demonstrated an enlarged, fluid-filled uterus with multiple foci of free air and inflammatory stranding

within both adnexa. Multiple calcified fibroids were also noted. Additionally, multiple foci of free air were noted throughout the abdomen, which initiated concern for perforated abdominal viscus, but a precise location for the perforation was not definitively identified on imaging despite an area of suspicion at the level of the sigmoid colon (see images). Finally, multiple pulmonary nodules were identified in the left lower lobe, measuring up to 1.3cm (Figures 2-8).

Laboratory investigations showed a hemoglobin of 8.9 g/dL, a white blood cell count of 29.90 k/uL, and a lactic acid of 6.4 mmol/L. Phosphorus was increased at 6.8 mg/dL. BUN and Creatinine were increased at 37mg/dL and 3.53 mg/dL, respectively, with an eGFR of 13 mL/min. Initial blood cultures showed no growth after 5 days. Follow-up labs four hours later showed a decrease in hemoglobin to 8.3 g/dL.

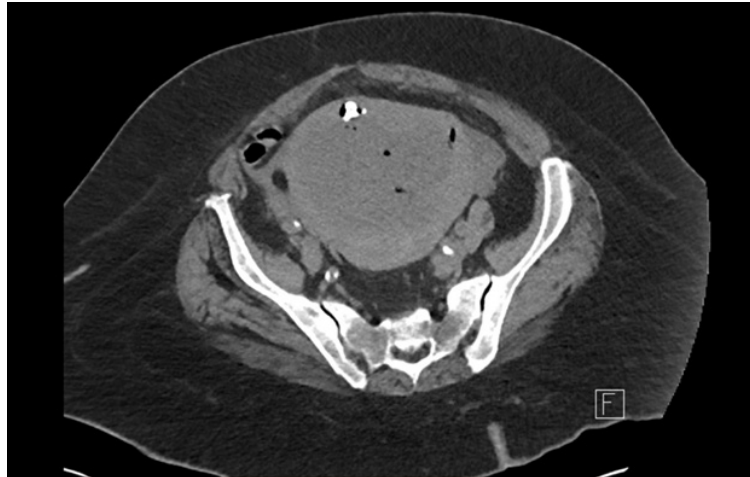


Figure 2:

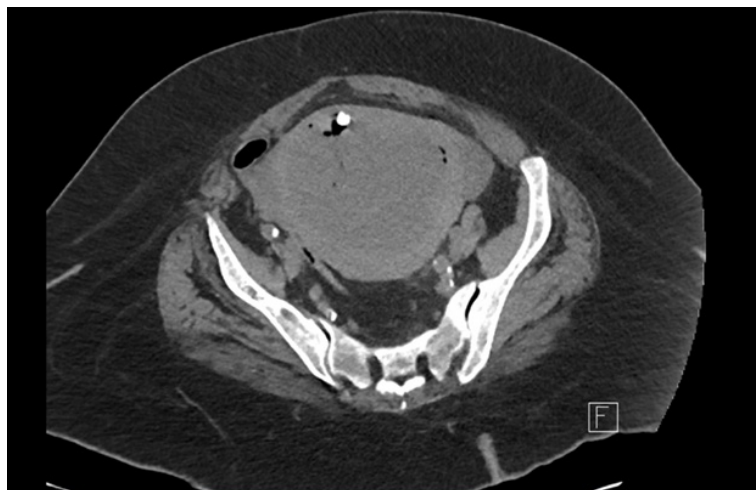


Figure 3:



Figure 4:

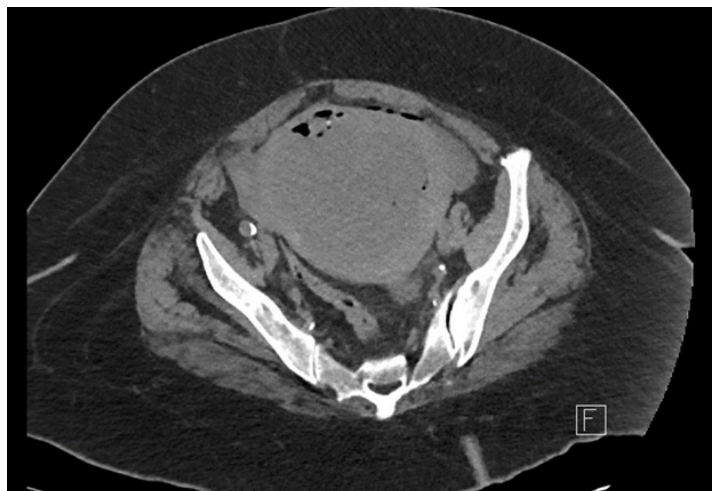


Figure 5:



Figure 6:

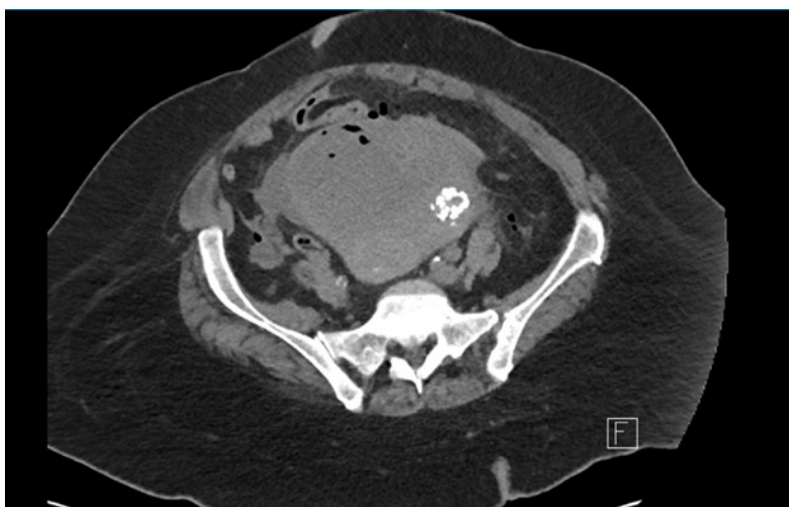


Figure 7:

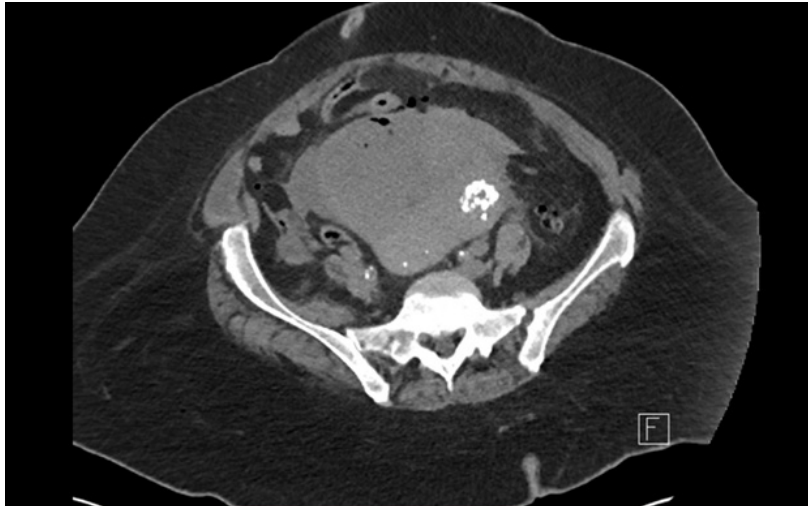


Figure 8.
Figures 2-8: Non-contrast CT of abdomen/pelvis showing necrotic uterine mass with free air traveling through fundal uterine serosa.

Treatment

In the absence of gynecological symptoms, the patient was referred to the general surgery service for evaluation of a perforated viscus, possibly secondary to a small bowel obstruction, since she had a history of abdominal surgery and symptoms consistent with obstruction. Initial treatment with intravenous fluids, antibiotics, and nasogastric suction was started before an emergent midline exploratory laparotomy was performed. The gynecologic team was called for an intraoperative consult prior to the initial incision. Frank purulence was encountered in the abdominal cavity, which was collected and sent for culture. There were no signs of perforation or ischemia throughout the small and large bowel. Examination revealed an 18-week-sized uterus with multiple leiomyomas. Two perforations were identified at the uterine fundus, secondary to a

suspected necrotic leiomyoma. A total abdominal hysterectomy, bilateral salpingo-oophorectomy, and partial omentectomy were performed by the gynecological team.

Gross examination of the uterus revealed two defects on the posterior wall and fundus measuring 2.5 x 1.9cm and 2 x 1.5cm, respectively, with necrotic tissue protruding through the defects. The anterior endometrial cavity showed an attached tan/red-green foul-smelling, necrotic, gangrenous mass measuring 11 x 8.5 x 3.5cm, protruding through the fundal defect. The posterior endometrial cavity also shows a 15 x 8.5 x 6cm yellow/red/tan/green foul-smelling, necrotic, gangrenous mass extending 100% through to the posterior serosal surface and protruding through the aforementioned defects [9,10].

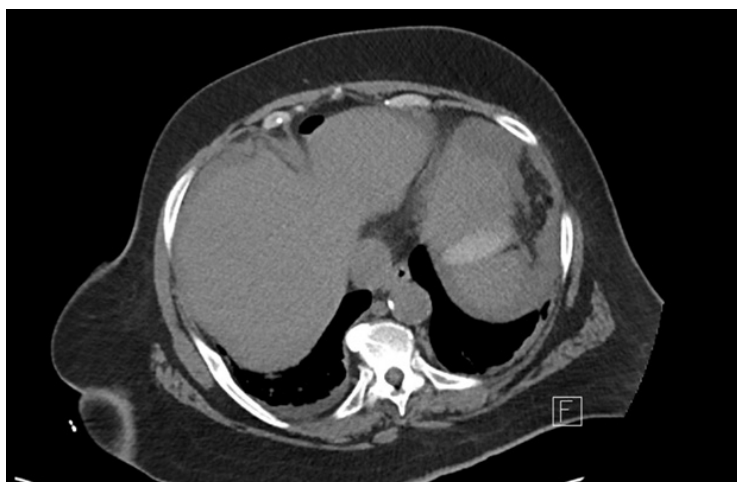


Figure 9: Non-contrast CT of abdomen/pelvis showing free air under the diaphragm.

Pathology determined the specimen to be carcinosarcoma of the uterus extending through the serosa and into the right fallopian tube. Additionally, multiple benign leiomyomas were noted. Sections from the specimen stained positive for desmin, myogenin, SMA, ER (focally), and PR (focally). Tumor cells seen in the sections were CK7-negative and exhibited a p53-null phenotype. The final, post-hysterectomy classification was FIGO stage IIIA carcinosarcoma of the uterus with extensive heterologous elements (rhabdomyosarcoma) and invasion of the myometrium. Lymphovascular invasion was extensive, but no nodes were submitted for evaluation (Figure 9).

Outcome and follow-up

The patient's recovery was complicated by a febrile urinary tract infection, positive for *Trichomonas*. She was discharged on hospital day 13 on metronidazole and nitrofurantoin with follow-up appointments for hematology/oncology, gynecology, and pulmonology [11].

Discussion

In this patient, the presence of the intraperitoneal free air in combination with the presented signs, symptoms, and laboratory findings, leads to a likely working diagnosis of gastrointestinal perforation. However, the concomitant presence of fluid in the uterine cavity and the multiple leiomyomas were suggestive of a possible uterine source for the intraperitoneal air/fluid interface. The subsequent multidisciplinary approach led to timely intervention, prompt diagnosis, and successful treatment of her emergent presentation, reinforcing the importance of interdepartmental collaboration in the management of such patients. With the gynecologic team already involved in the case and standing by in the operating theatre, the patient was able to undergo a total abdominal hysterectomy, salpingo-oophorectomy, and partial omentectomy immediately, when the need became apparent to the team. Overall, this allowed for quick identification of her UCS, allowing her to be discharged on hospital day 13 for adjuvant radiation and chemotherapy.

Many women who may have uterine cancer are not diagnosed until they present with gynecological symptoms. Abnormal vaginal bleeding is the overwhelmingly predominant symptom, being in upwards of 90% of cases [12,13]. Approximately two-thirds of uterine cancers are diagnosed at the local stage in Caucasian women, while only about half of all cases are caught in the early stages in African-American women [14]. While UCS represents less than 5% of all uterine cancers, African-American females have a two-fold higher incidence of leiomyosarcomas and carcinosarcoma than Caucasian females [15,16]. Reviewed data from 160 studies estimated the prevalence of unexpected leiomyosarcoma at the time of surgery for presumed symptomatic leiomyomas to be in the range of <1 and up to 13 per 10,000 surgeries [17].

With a median overall survival time of less than two years, UCS is associated with a poor prognosis despite improved clinical management of symptoms [18]. While this could be due to numerous factors, high-grade uterine sarcomas commonly tend

to metastasize hematogenously, most often to the lungs, leading to rapid metastatic spread before symptomatic presentation [19]. Lymph node metastases were considered uncommon in UCS, but have been increasingly common in recent years, being seen in upwards of one-quarter of cases [14]. As such, lymphadenectomy appears to be associated with an increased survival benefit [20].

Current guidelines do not recommend screening for uterine cancers [12]. However, with the rapid progression of UCS, early identification and removal of the primary tumor burden greatly improves long-term management and overall survival rate [20]. It is for this reason that providers need to be aware that UCS can present without gynecological symptoms, as in this case. Gynecologic surveillance of the postmenopausal patient with known uterine myoma should not be overlooked as sarcomatous changes can be detected by imaging techniques before the onset of clinical symptoms. Earlier recognition of our patient's disease process likely would have prevented her acute, emergent presentation.

Acknowledgement

None.

Conflict of Interest

The authors declared no conflict of interest with respect to the research, the authorship, and/or publication of this article.

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