

## Guess the Case

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### INTRODUCTION

A 48-year-old female patient 4 months prior to admission to the Ochsner Medical Center had a laparoscopic hysterectomy with bilateral salpingo-oophorectomy, cystoscopy, and pelvic washings at an outside hospital with an incidental finding of a uterine sarcoma. The pathology report showed undifferentiated endometrial sarcoma invading into the inner third of the myometrium, with no vascular space invasion or involvement of the cervix, tubes, or ovaries, and washings were negative. A preoperative computed tomographic (CT) scan showed no evidence of metastatic disease; it was recommended that she receive adjuvant pelvic radiation to decrease her risk of local recurrence, which she completed several weeks prior to admission. Five days prior to admission, she went to an emergency department with right flank pain and was treated for a urinary tract infection. Two days prior to admission she had a routine follow-up with her gynecologist and stated she had had a temperature of 100.8°F the previous day but otherwise was feeling well. On physical examination, there was some mild right lower quadrant tenderness and vital signs were normal. On bimanual examination and rectovaginal examination, there were no masses palpated.

On the day of her admission to Ochsner Medical Center, she was seen at an outside hospital with worsening right lower quadrant pain. A CT scan was obtained (Figure), and with a diagnosis of appendicitis she was transferred to our institution for access to a general surgeon. On arrival, she reported 8 of 10 right lower quadrant pain without fever, nausea, vomiting, or diarrhea. She was taking Bactrim for her urinary tract infection. Past medical history and review of symptoms were not significant. On physical exami-

nation, she was in moderate distress and her vital signs were normal. Her abdomen was soft without distention, guarding, or rebound, but there was tenderness in the right lower quadrant. Laboratory values revealed a normal white blood cell count without shift. Her chemistry panel was normal except for an albumin of 3.3 g/dL. What is your diagnosis?

### DISCUSSION

On further evaluation of the outside CT scan, we thought that the lesion in the right lower quadrant was a heterogenous mass, not an abscess. There was no surrounding inflammatory response. In the pelvis, there was another mass associated with dilation of the small bowel just prior to its entry into the cecum. After admission, the right lower quadrant mass was biopsied and showed recurrent undifferentiated uterine sarcoma.

This case presented a lot of interesting points. Appendicitis can be a difficult diagnosis to make. The patient certainly had worsening right lower quadrant pain, which possibly could have been appendicitis partially treated by the antibiotics given to her in the early stages, allowing for the development of chronic appendicitis with perforation and localization of a periappendiceal abscess. What argued against this, however, was the relatively mild amount of right lower quadrant tenderness, normal white blood cell count, and a CT scan without inflammatory changes. In addition, we thought that the CT showed masses and not fluid collections. For that reason, other inflammatory processes such as Crohn's disease or diverticulitis were unlikely. This example shows the difficulty in diagnosing pelvic masses on physical examination. The abdominal wall musculature is thick enough that many abdominal masses are missed until they get quite large and/or become fixed.

This was an unexpectedly early recurrence of this uterine sarcoma; however, whenever a cancer patient has a new mass, recurrent cancer is always on the differential diagnosis. In this case, the white count was normal and the pain relatively indolent relative to the typical case of appendicitis, and the CT scan showed 2 new masses without identifying the appendix. With these findings, recurrent cancer became first on the list of differential diagnoses.

Uterine sarcomas are rare tumors, making up only 9% of all uterine malignancies.<sup>1</sup> The average age at

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**This is a representative view of the abnormalities in this patient from the abdominal/pelvic computed tomography scan.**

diagnosis is 60 years, with an incidence of approximately 3.6 per 100,000 women over age 35.<sup>2</sup> The classification of these tumors changed in 2009, with the most common type of uterine sarcoma, carcinosarcoma, no longer classified as a sarcoma but rather a uterine carcinoma. Low-grade and high-grade endometrial stromal sarcomas were also reclassified as endometrial stromal sarcomas and undifferentiated endometrial sarcomas, respectively. Leiomyosarcomas make up the final tumor in this group. Uterine sarcomas typically behave more aggressively and have a poorer prognosis than uterine carcinomas.

The diagnosis of a sarcoma is considered when a premenopausal woman experiences uterine bleeding or pain disproportional to the size of the uterus and/or presence of fibroids. The suggestion that “rapidly growing” fibroids are indicative of sarcomas has not been substantiated in the literature and should not be considered a risk factor for sarcomas. In postmenopausal women, uterine bleeding is the usual presenting symptom. Endometrial sampling will give the correct diagnosis in some, but not all, patients with a uterine sarcoma. Endometrial biopsy and uterine curettage sample mainly the endometrial lining where uterine carcinomas originate and not the mesenchymal component of the uterus where the malignant cells of sarcomas may arise.<sup>3,4</sup> Currently, there are no biomarkers available for the detection of uterine sarcomas.

The only treatment of curative value for uterine sarcomas is surgical excision. Surgical staging for uterine sarcomas includes an extrafascial hysterectomy with bilateral salpingo-oophorectomy. The role of lymphadenectomy for these tumors is controversial and typically not considered useful for undifferentiated endometrial sarcomas or leiomyosarcomas. Adju-

vant postoperative pelvic radiation is recommended for undifferentiated endometrial sarcomas and leiomyosarcomas to prevent local recurrence, but there is limited evidence that it provides any survival benefit. Uterine sarcomas most commonly metastasize to the lungs, abdomen, and pelvis.<sup>5</sup> Chemotherapy may be used at the time of initial diagnosis or for recurrent disease, although uterine sarcomas as a group tend not to be very chemosensitive with most responses of short interval. Because of the limited number of these tumors, there is little consensus about the use of adjuvant chemotherapy for sarcomas. Endometrial stromal sarcomas have the best prognosis of the uterine sarcomas and require only observation for stage I and II disease because of the more indolent nature of these tumors.<sup>6</sup> For more advanced stages or recurrent endometrial stromal sarcomas, adjuvant hormonal therapy is recommended. Typical regimens include progestins or aromatase inhibitors, depending on the estrogen and progesterone receptor status of the individual tumor. Uterine sarcomas carry a poorer prognosis than uterine carcinomas. Stage I uterine sarcomas have a 5-year survival rate of 76%, falling to 60%, 45%, and 29% for stage II, III, and IV disease, respectively.

Final treatment plans for this patient were for surgical debulking followed by adjuvant chemotherapy. Secondary surgery/debulking is an aggressive option for recurrent uterine sarcoma but was thought to be appropriate in this patient because of her symptoms. Statistically, the overall outcome is likely to be poor.

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