Right atrial metastasis of uterine leiomyosarcoma causing obstructive shock

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ABSTRACT

Uterine leiomyosarcomas are rare tumours, and secondary cardiac metastases are even rarer. We present the case of a 56-year-old ethnic Chinese woman who was being treated with adjuvant pelvic radiation for uterine leiomyosarcoma when she presented with signs of right heart failure and shock. She was rapidly diagnosed with a solid mass attached to the tricuspid valve. Subsequent surgical resection revealed leiomyosarcoma metastasis. Metastases of uterine leiomyosarcoma to the heart are extremely rare, but clinicians should be aware of this phenomenon. Surgical resection, when feasible, can be associated with longer survival.

KEY WORDS

Leiomyosarcoma, uterus, heart atria, neoplasm metastasis

1. INTRODUCTION

Uterine leiomyosarcomas are rare, accounting for 3% of all uterine malignancies1. Typically, early and frequent aggressive hematogenous dissemination can occur, with the lungs being the site most frequently involved, followed by the abdomen; bone and brain are less prevalent2. Symptomatic cardiac metastases from uterine leiomyosarcomas are extremely uncommon3. Here, we report the case of a woman with uterine leiomyosarcoma who developed tumour metastasis that involved the right atrium, with right-sided obstruction to flow causing obstructive shock.

2. CASE DESCRIPTION

A 56-year-old ethnic Chinese woman was referred to our institution in July 2011 for opinion on further management. She had been healthy until she experienced urinary symptoms, frequent chills, and low-grade fever in April 2011.

Pelvic ultrasonography on April 15 reported enlarged fibroids, the largest being 6.7×5.8×5.2 cm. On May 3, an endometrial biopsy reported scanty epithelium only. On May 15, computed tomography (CT) imaging of the abdomen and pelvis showed an enlarged uterus, with multiple fibroids. The patient was then booked for elective surgery. However, in June, she experienced increasing pelvic pain and an enlarging abdominal mass, and she presented herself to the emergency department. Chest radiography on June 12 was otherwise normal.

On June 14, the patient underwent a hysterectomy and bilateral salpingo-oophorectomy. A rupture of the serosal surface of the uterine fundus occurred during surgery. On the pathology report, a myometrial leiomyosarcoma, grade 3/3, measuring 15 cm, with extensive necrosis and prominent vascular invasion, was diagnosed. The tumour was also present on the serosal surface, although the surgical margins were free of tumour.

Postoperatively, the patient recovered well, and her symptoms resolved completely. Re-staging with CT of the abdomen and pelvis showed no residual malignancy. The patient consented to our recommendation of adjuvant pelvic radiation over 5 weeks, with the intent of reducing the risk of pelvic relapse. The recommended regimen was started at the end of July.

After the 6th radiation fraction, the patient was seen as part of standard medical follow-up during treatment. She described experiencing right central chest discomfort and tachycardia since the start of radiation treatment. She had not mentioned those symptoms beforehand. Otherwise, she had been feeling well.

Blood work was ordered that day (August 4). Complete blood count and liver and renal function were normal, except for D-dimer which was minimally elevated at 760 μg fibrinogen equivalent units per litre. Electrocardiography demonstrated sinus tachycardia at 113 bpm, without any other anomaly.

The next day, the patient awoke experiencing sudden dyspnea, dizziness, fever, and tachycardia.
On exam, she looked ill, diaphoretic, and mildly distressed. She was not experiencing any new pelvic symptoms or leg swelling or pain. Her vitals suggested shock: respiratory rate, 40; blood pressure, 76/60 mmHg; heart rate regular, but high at 137 bpm. In addition, jugular venous pressure was 4 cm, and peripheral pulses were weak yet palpable. Chest was clear to auscultation, and she had an abnormal S1 with a gallop rhythm and a low-pitched murmur. No abdominal findings and no clinical findings of deep venous thromboembolism were present on exam.

The patient was fluid-resuscitated, resulting in an increase in blood pressure. Urgent blood labs showed a mildly elevated white blood cell count (11.7x10^9/L), with mild neutrophilia; chemistry and renal function were normal. An urgent CT pulmonary angiogram done to rule out pulmonary embolism showed no evidence of embolism, but uncovered numerous pulmonary nodules. In addition, a 4-cm ovoid hypodense soft-tissue mass was detected interposed between the right atrium and the right ventricle (Figure 1).

The patient was admitted to hospital, and the consulting cardiologist performed an echocardiography, describing a large 4x3-cm solid mass adherent to leaflets of the tricuspid valve, completely impeding the flow across the valve. Vegetation did not seem likely on imaging. Hyperdynamic left ventricular systolic function (with an ejection fraction exceeding 75%) was present. Given the patient’s symptoms, the mass may have been secondarily infected. Blood was taken for cultures, and intravenous antibiotics for cultures, and intravenous antibiotics and full anticoagulation were ordered. (The cultures were negative after 5 days’ incubation.)

The patient underwent cardiac surgery on August 6. Operative findings were of a large tumour with a broad-based attachment to the medial wall of the right atrium. The mass was completely resected, leaving the patient with a severe tricuspid regurgitation. The right atrial wall was reconstructed using bovine pericardium. Pathology revealed the mass to be consistent with metastatic leiomyosarcoma.

The patient was discharged home on August 23. On review of systems, her only symptoms at discharge were shortness of breath while walking up an incline. On exam, her vitals were stable and signs of heart failure were no longer evident. Because of disseminated disease, radiation was cancelled, the patient having received a total of 1600 cGy in 8 fractions. She was referred to the medical oncology team to discuss palliative chemotherapy.

3. DISCUSSION

Our case reinforces the aggressive nature of uterine leiomyosarcoma. Lung, pancreatic, bone, and brain metastases have been described, and cases of thyroid, muscle, skull, head-and-neck, and breast metastasis have also been identified. Uterine leiomyosarcomas can therefore behave in an unpredictable fashion, as demonstrated in this case of a 56-year-old patient whose cardiac metastasis manifested itself during adjuvant radiation treatment.

Because of rarity, few cases of uterine leiomyosarcoma with tumour emboli to the heart have been reported. In a case series reported in 1960, only 6 of 2600 patients with metastatic cardiac tumours had those tumours traced to a uterine leiomyosarcoma primary. Several groups have published more recent reports. Calleja et al. described the case of a 48-year-old woman with a history of uterine leiomyosarcoma diagnosed with extensive cardiac metastases (in the left atrium, right and left ventricles, pulmonary artery, and pulmonary vein) accompanied by pericardial tamponade, 5 years after initial diagnosis. She required a pericardial window, but because of an increase in size of the cardiac masses 3 weeks later, with concomitant pulmonary embolism, she opted for palliative care and died within hours after hospital admission. A Taiwanese group published another case of a 49-year-old woman experiencing sudden onset of orthopnea, dyspnea on exertion, and palpitations. She had been treated 9 years earlier for a stage III uterine leiomyosarcoma. Workup, including chest radiography and echocardiography, demonstrated pulmonary edema, bilateral effusions, and a mass lesion measuring 6.65 cm occupying two thirds of the left atrium. Ejection fraction was 69%. That patient underwent complete resection with complete resolution of symptoms, and the mass showed patterns similar to the earlier leiomyosarcoma.

To the best of our knowledge, the current case is unique because in the English-language literature, cases of metastatic uterine leiomyosarcoma have predominantly described left atrial or ventricular masses, where our case presented with right atrial tumour emboli. The right atrial involvement is most likely the result of dissemination through the bloodstream, with tumour cells adhering to the tricuspid...
valve leaflets. The resulting differential included an intracardiac clot, a primary cardiac tumour mass such as a myxoma, or a leiomyosarcoma metastasis. It is hard to discern if the patient had metastatic disease at diagnosis. Preoperative thoracic imaging (CT or radiography) is recommended during initial workup. Although chest imaging was done, the sensitivity of chest CT is greater than that of chest radiography. As part of restaging after surgery, chest CT is not routine, but it might have proven insightful here.

To define the best management strategy for these patients, all case reports are important. Although rare, cardiac metastasis should be considered in a patient with uterine leiomyosarcoma who develops cardiovascular symptoms. Echocardiography is the easiest and most widely available way to evaluate cardiac chambers; however, CT angiography provides the means to consider various causes (such as pulmonary emboli, cardiac tamponade, infection, and so on). When complete resection is feasible and undertaken, patients are able to live free of disease in the heart at follow-up. Resection can be used to provide symptom palliation and revert an emergent life-threatening situation. In our patient, surgery prevented sudden death secondary to gradual complete obstruction of the right ventricular outflow tract.

Clinicians treating uterine leiomyosarcoma should be aware of the proclivity of those tumours for metastasis, sometimes to unusual sites, and of the need to perform total surgical removal of critical lesions. Complete removal is especially important in patients with pre-existing good performance status who may be able to tolerate further systemic treatments, because newer agents offer promise.

4. CONFLICT OF INTEREST DISCLOSURES

The authors have no financial conflicts of interest to declare.

5. REFERENCES


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