Multimodality Cardiac Imaging Assessment of a Large Metastatic Pericardial Leiomyosarcoma

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INTRODUCTION

Uterine leiomyosarcomas (ULMS) are rare tumors, with an incidence of 0.64 cases per 100,000 women.[1] The common sites of metastases are lung, peritoneum, and bone. Cardiac metastases are very rare. The reported sites of cardiac metastases mainly involve the right heart and associated structures. This is the first case report of a large pericardial metastatic leiomyosarcoma, with no associated lung metastasis. We illustrate how multimodality cardiovascular imaging was used for diagnosis and surgical planning.

CASE PRESENTATION

A 56-year-old woman was referred to our center for further investigation and management of a large pericardial mass. At presentation, she was clinically well. Her vital signs were stable (pulse rate 61 beats/min, regular; blood pressure 135/80 mm Hg). Her relevant history included total abdominal hysterectomy with bilateral salpingo-oophorectomy for a high-grade leiomyosarcoma (estrogen receptor–positive 20%, progesterone receptor–positive 100%, and human epidermal growth factor receptor 2/neu–negative) in August 2008. She had been maintained on anastrozole for 1.5 years before presentation. She did well until late 2016, when she developed shortness of breath, orthopnea, and lower extremity edema. Echocardiography showed bilateral pleural effusions, a large pericardial effusion with tamponade physiology, and a pericardial mass. She underwent urgent pericardial window, pericardial drainage, and pericardial biopsy. Histopathology from the pericardial biopsy demonstrated chronic inflammation with no malignancy seen, though sampling was limited. Subsequently, a localized mass was discovered in the patient's right submandibular gland. She underwent submandibular gland resection in March 2017. Histopathology demonstrated evidence of metastatic leiomyosarcoma.

Since that time, the patient has had progressive shortness of breath and palpitations, associated with weight loss. Positron emission tomography (PET) with ^{18}F-fluorodeoxyglucose (FDG) was performed and demonstrated mildly increased FDG uptake within the solid part of the large pericardial mass (Figure 1). There was an additional mildly FDG-avid paravertebral mass, with focal sternal FDG uptake, suggestive of metastatic disease. Chest and abdominal computed tomography (CT) also demonstrated a large lobulated, mixed solid-cystic pericardial mass (Figure 2A). There was a small region of possible myocardial invasion into the inferoposterior aspect of the left ventricle (Figure 2B). The left circumflex artery and its branches were "entrapped" and compressed between the mass and the left heart (Figure 3, Video 1).

Transthoracic echocardiography (TTE; Figure 4, Video 2) and TEE (Figure 5, Video 3) demonstrated a large heterogeneous mass lateral to the left ventricle. Additionally, there was a cystic component associated with the mass. There was no obvious invasion of the myocardium.

Cardiac magnetic resonance imaging (CMR) revealed a large mixed solid-cystic pericardial mass (Figures 6A–6C). The solid portion demonstrated heterogeneously hyperintense signal on black blood, white blood, and T2-weighted imaging and heterogeneous enhancement on perfusion imaging. The cystic portion demonstrated high signal intensity on T1-weighted and T2-weighted imaging without enhancement on perfusion imaging. There was uniform circumferential increased signal (suggesting fibrous encapsulation) around the lateral aspect of the mass, which was not as evident adjacent to the left ventricular lateral wall, where tissue planes were not well defined (Figure 6D). The overall features on CMR were supportive of a malignant tumor. Adherence to the anterolateral wall of the left ventricle, pericardium, and the lateral wall of the left atrium was also noted. No distinct invasion of the left ventricular myocardium was observed, although this could not be excluded, as tissue planes were not well defined. There was mild compression of the left atrium without obvious wall or chamber invasion. The mass measured approximately 12.0 × 7.0 cm in the axial plane and 9.5 × 8.7 cm in the sagittal plane (Figure 7).

Following multimodality cardiovascular imaging assessment, the pericardial mass was deemed resectable. At the time of the operation, the mass appeared to arise from a pedicle on the posterolateral aspect of the left ventricle, with a small amount of epicardial invasion into the posterolateral surface of the left ventricle (Figure 8). The mass was resected, with clear margins (Figure 9).

Histopathologic examination of the mass demonstrated a proliferation of elongated spindle cells with eosinophilic cytoplasm and

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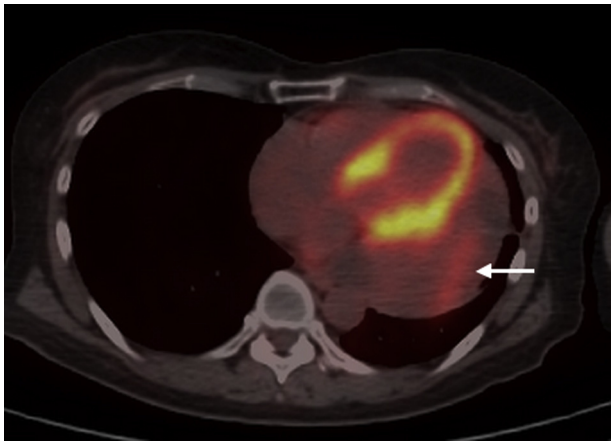

Figure 1 FDG PET/CT demonstrating radiotracer uptake in the solid part of the pericardial mass (*white arrow*). Maximum standardized uptake value is 3.48.

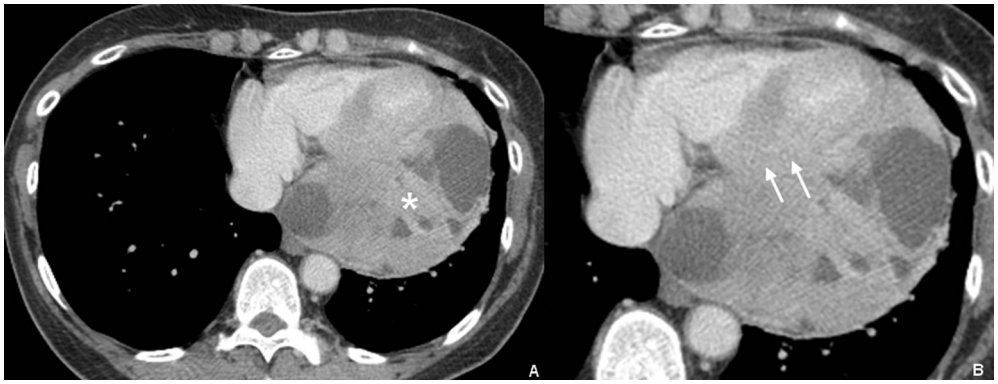

Figure 2 **(A)** Contrast-enhanced axial computed tomographic scan, demonstrating a large mixed solid-cystic pericardial mass. The solid component shows heterogeneous enhancement (*asterisk*). **(B)** Contrast-enhanced axial computed tomographic scan (zoomed-in image) demonstrating no distinction in the fat plane separating the mass from the myocardium at the inferoposterior aspect of the left ventricle (*white arrows*).

oblong blunt-ended nuclei arranged in intersecting fascicles (Figure 10A). Numerous mitotic figures were identified (14/10 high-power fields). Immunohistochemical stains were positive for smooth muscle actin and desmin (Figure 10B). Pathologic findings confirmed with metastatic pericardial leiomyosarcoma. Five days after surgery, the patient was discharged. This patient was last seen at our center when she came for postoperative FDG PET/CT (1 month after surgery). Postoperative FDG PET/CT at 1 month showed no definite residual FDG-avid disease in the resection bed. She is now followed up locally, and we do not have any further information.

DISCUSSION

Uterine sarcomas are rare neoplasms that can be classified into three types by composition: smooth muscle tumors, endometrial stromal tumors, and tumors with both smooth muscle and epithelial components.[2] Leiomyosarcoma is a malignant neoplasm with smooth muscle differentiation and the second most common subtype of uterine sarcoma.[2] In most cases, the diagnosis of ULMS is made following hysterectomy or myomectomy for presumed benign uterine leiomyomas. The standard management of a localized ULMS is surgical and includes total abdominal hysterectomy with or without salpingo-oophorectomy. In patients with extrauterine disease spread, additional surgical resection of metastatic disease or systemic chemotherapy is recommended, depending on the staging of the disease. ULMS is biologically aggressive and has a propensity for early hematogenous spread.[3] Local recurrence and metastatic disease predict poor outcomes. In a recent study of 113 patients, the most common sites of metastases from uterine sarcoma were lung (74%), peritoneum (41%), bone (33%), and liver (27%).[4] Metastasis to the heart is rare. The reported sites of cardiac metastases mainly involve the right heart and associated structures, particularly the right atrium and the