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The First Case Report of a Metastatic Myxoid Liposarcoma Invading the Left Atrial Cavity and Pulmonary Vein

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ABSTRACT

Myxoid liposarcoma (MLS) is the most commonly encountered liposarcoma subgroup, accounting for about 50% of all cases. Metastatic MLS of the heart is extremely rare. Herein we describe for the first time metastasis of MLS to the left atrium and left upper pulmonary vein in a 54-year-old woman who was admitted with shortness of breath and cough persisting for 2 weeks. The patient reported that a total surgical excision of MLS of the left thigh followed by radiotherapy was performed 4 years ago. An emergency operation was performed due to rapidly progressive worsening of clinical condition and echocardiographic determination of left atrial mass protruding into the left ventricle and obstructing the mitral inflow throughout the diastole. The mass could not be totally excised because it was tightly adhered to the surrounding tissue. Postoperative magnetic resonance imaging (MRI) showed a 5 × 3 cm residual tumor deforming the posterior wall of the left atrium entirely and extending into the left upper pulmonary vein. Histopathological examination was consistent with MLS. In conclusion, considering probable cardiac metastasis in patients presenting with respiratory symptoms with medical history of soft tissue sarcomas would be life saving. The case is discussed, and a review of the literature in relation to the metastatic involvement of the heart by MLS is presented.

INTRODUCTION

Liposarcomas are among the most frequent malignant soft tissue tumors in adults. Five-year survival after resection for well-differentiated and myxoid liposarcomas is estimated to be high, but pleomorphic liposarcomas are characterized by infiltrative growth and early metastasis [Chang 1989]. Myxoid liposarcomas usually have a high predilection for extrapulmonary metastasis, but metastasis to the heart is a rare diagnosis [Estourgie 2002].

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Herein we report the first case of a metastatic myxoid liposarcoma (MLS) invading the left atrial cavity and pulmonary vein and causing congestive heart failure.

CASE REPORT

A 54-year-old woman presented with progressive shortness of breath and cough persisting for 2 weeks and made worse in supine and left lateral decubital positions. Results of a chest x-ray were consistent with pulmonary venous redistribution, a prominent pulmonary trunk, and increased interstitial signs. Laboratory tests were within normal limits. The patient was admitted to the Pulmonology Department with the preliminary diagnosis of a pulmonary disease; however, her symptoms gradually worsened within the subsequent hours. Cardiac examination revealed a high-grade, low-pitched diastolic murmur best heard at the apex and rales in the lower half of both lungs on auscultation; heart rate and blood pressure were 120 beats/minute and 80/50 mm Hg, respectively. Echocardiography showed a 7 × 4 cm homogeneous hyperechogenic mass in the left atrium that was attached to the left atrial lateral wall, protruding into the left ventricle through the mitral valve in the early diastole (Figure 1). The mitral inflow tract was almost occluded throughout the diastole, and a 29 mm Hg transvalvular peak systolic gradient was being generated. Her past medical history was remarkable for a biopsy-proven MLS of the left thigh 4 years before, which was resected and treated with subsequent chemotherapy and radiotherapy. The patient had a completely negative workup including a thorax computed tomography (CT) scan and transthoracic echocardiogram 11 and 5 months prior to the symptoms, respectively.

Because of the rapidly progressing symptoms of heart failure, the patient was promptly referred to the cardiovascular surgery department. In the emergency operation, a mass occupying almost the entire left atrial cavity and extending into the upper left pulmonary vein was encountered. Only a partial resection of the tumor could be achieved because the tumor was tightly adhered to the pericardium and myocardial layers of the left atrial wall were deeply infiltrated.

Macroscopic examination showed a gelatinous and uncapsulated tumor measuring 7 × 7 × 1 cm with a myxoid cut surface (Figure 2). Microscopic examination was consistent with lipoblasts in different stages of differentiations in mucoid

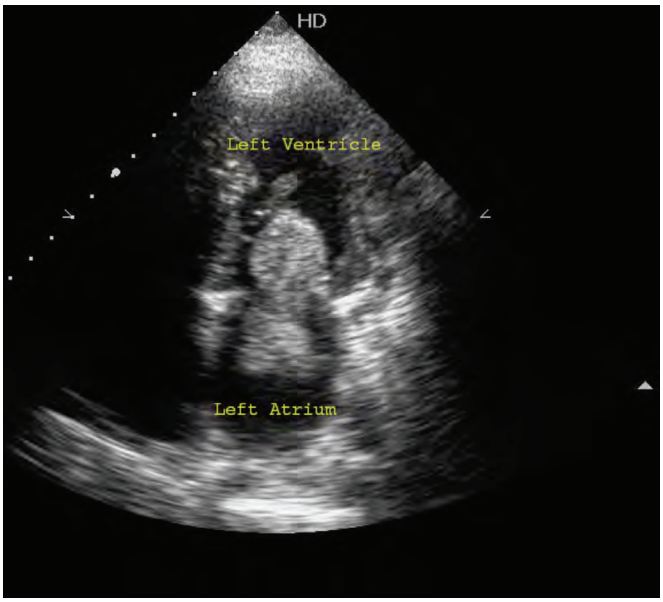

Figure 1. Echocardiographic image of the mobile mass in the left atrial cavity almost occluding the mitral inflow in the diastole.

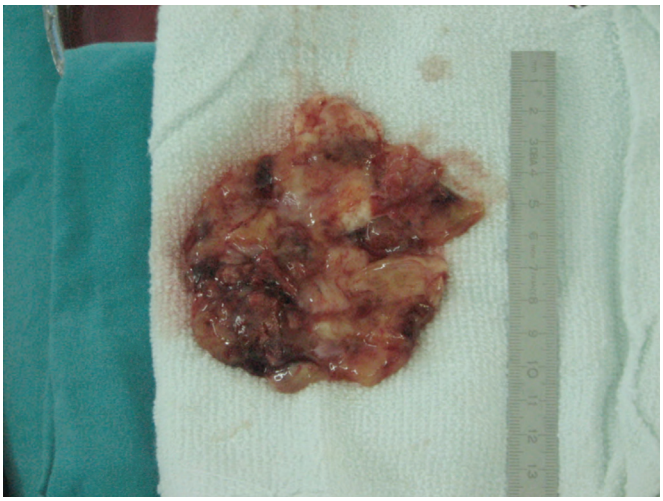

Figure 2. The appearance of the tumor after excision.

matrix. There were rare mitotic figures and tumor cells stained with S100 immunohistochemically. The tumor had infiltrated small fragments of the attached pericardium (Figure 3). The pathologic diagnosis was metastasis of a MLS.

A postoperatively performed magnetic resonance imaging (MRI) showed a 5 × 3 cm residual tumor deforming the posterior wall of the left atrium entirely and extending superiorly into the pulmonary venous confluence and left upper pulmonary vein. It was adjacent to the anterior wall of the thoracic aorta, and the pericardium of the left atrium was also involved (Figure 4). CT also identified an inoperable metastatic mass (9.5 × 8 cm) infiltrating the left lobe and caudate lobe of the liver.

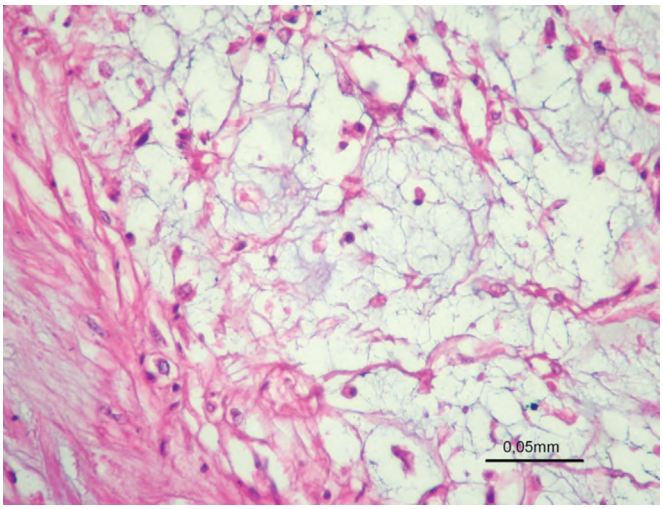

Figure 3. Atypical lipoblasts infiltrating the pericardium.

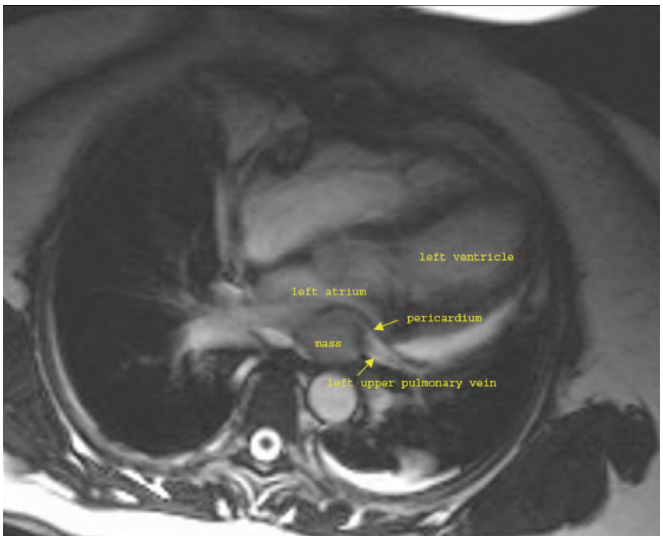

Figure 4. Axial magnetic resonance imaging (MRI) shows the residual tumor invading the pericardium and extending into the left atrial cavity and left upper pulnonary vein.

Following the surgery, the patient's symptoms were resolved completely. Radiotherapy and chemotherapy with dacarbazine and gemcitabine were performed. The patient was followed up monthly for 9 months with physical and echocardiographic examinations. She was completely asymptomatic during the follow-up period and echocardiography and MRI showed no significant change in the size of the residual tumor in the left atrium. However, abdominal CT performed 5 months after the surgery showed further enlargement of the tumor in the liver (17 × 12 cm). The patient's family reported that the patient died in the tenth month after the surgery, after 2 days of abdominal distension and widespread edema of the lower extremities.

DISCUSSION

MLS has a low rate of distant metastasis, and metastasis of MLS to the heart is a rare diagnosis [Chang 1989; Fairman 2005]. Signs and symptoms are related to the size and the location of the tumor [Veinot 1999]. Dyspnea, chest pain, and edema are among the most commonly encountered symptoms [Fairman 2005]. Experience for the treatment of metastatic MLS is limited by the rareness of the reported cases. Total excision is the choice of treatment whenever possible, but the success of the surgery is usually limited with palliative interventions [Murphy 1990; Fairman 2005; Chughtai 2007; Komoda 2009]. Most of the patients have limited life expectancies because of the widespread nature of disease or cardiac complications. Though there are no definite guidelines for these patients, based on extrapolation from studies of primary cardiac sarcomas it might be considered that chemotherapy and radiotherapy are among the treatment options for patients in whom complete resection could not be achieved [Burke 1992; Bakaeen 2009].

We present this case because in the literature there are only a limited number of cases with intracavitary cardiac metastasis, and a MLS of the left atrium invading the pulmonary venous structures has not been reported priorly. Second, in contrast to the general perception, MLS may grow fast. Our patient had a completely negative work up including a transthoracic echocardiogram 5 months prior to the symptoms. Therefore, scanning the heart for possible metastases may be indicated in cases with cardiac and pulmonary symptoms. Although the prognostic value of screening needs more conclusive evidence, transthoracic echocardiography is sensitive in detecting cardiac metastases, and it might be the investigation of choice in these patients because early diagnosis improves the surgical outcome. Third, complete resection of the tumor was not possible, but this urgent procedure was life saving, and the size of the residual tumor in the left atrium did not change throughout the echocardiographic follow-up period. Although the obstructive effect of an excessively enlarged intraabdominal metastatic mass was one of the potential causes of death, it was not possible to determine the exact etiology.

In conclusion, distant metastasis of MLS to the heart is extremely rare but must be excluded in patients presenting with cardiopulmonary symptoms. Available data are insufficient to identify the optimal treatment strategy, but emergency surgery might provide a stable state for a limited period of time.

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