Pulmonary Metastatic Leiomyosarcoma with Left Atrial Extension via the Pulmonary Vein Manifests as Acute Pulmonary Edema

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We report a case who presented to our emergency department with symptoms of congestive heart failure and acute pulmonary edema. The transthoracic echocardiography demonstrated a large tumor that occupied two-thirds of the left atrium and impeded the mitral valve flow. The patient underwent cardiac surgery with removal of the LA tumor that was intraoperatively found extending through the right inferior pulmonary vein. A big tumor mass over the right lower lobe of the lung was also disclosed. The pathological report of the left atrial tumor was metastatic leiomyosarcoma. Postoperative survey showed multiple metastatic tumors over the lung, liver, bones and mediastinal lymph nodes. The symptoms and signs of congestive heart failure improved after the surgery. The patient had history of uterine leiomyosarcoma and received debulking surgery with postoperative radiotherapy 9 years prior to this hospitalization. This is the first report of a pulmonary metastatic leiomyosarcoma reaching the left atrium by way of the pulmonary veins and manifested as acute pulmonary edema.

Key Words: Metastatic pulmonary leiomyosarcoma • Acute pulmonary edema • Tumor invasion of pulmonary veins

INTRODUCTION

Metastatic cardiac tumor with congestive heart failure is not uncommon. The origin of most metastatic cardiac tumors are melanoma, leukemia, lymphoma and carcinoma.1 Primary pulmonary malignancies with invasion of the pulmonary veins and left atrium (LA) occurred most frequently in non-small cell carcinoma of the lung.2 Primary sarcoma and malignant epitheloid leiomyoblastoma of the lung have been reported.3,4 However, metastatic pulmonary malignancies with invasion of pulmonary veins and extending into LA are rare. They have been presented in metastatic uterine carcinoma, including metastatic choriocarcinoma, cervical carcinoma, malignant fibrous histiocytoma and metastatic chondrosarcoma.5,7

Uterine leiomyosarcoma commonly spreads through hematogenous rout. The peritoneal cavity and omentum are the most frequently involved sites (59%), followed by the lungs (52%), pelvic lymph nodes (41%), paraaortic lymph nodes (38%), and liver parenchyma (34%).8 Tumor extending to the LA via pulmonary vein has never been reported in metastatic leiomyosarcoma.

CASE REPORT

A 49-year-old lady was diagnosed as uterine leiomyosarcoma 9 years prior to admission. She underwent debulking surgery with postoperative radiotherapy. She had regular follow-up till 3 years ago, and there was no local recurrence. She had been well until a week prior to
admission, when she began to experience shortness of breath on exertion. She suffered from increasing dyspnea and orthopnea in the evening before admission and was brought to another hospital, where acute pulmonary edema with pleural effusion was noted. She received diuretic treatment and the symptoms improved. Transthoracic echocardiography (TTE) disclosed a tumor mass in the LA. She was transferred to our hospital for further evaluation and management. Her past and family histories were unremarkable.

On admission, the patient showed acute ill-looking. Consciousness was alert. The physical examination revealed a temperature of 36.7 °C, blood pressure of 140/80 mmHg, pulse rate at 80 beats/min and respiratory rate at 20 min. The jugular venous pulse was 5 cm above sternal angle at 45°. There was no neck lymphadenopathy. Lung auscultation showed moist rales over bilateral bases. Cardiac examinations revealed regular heart beats and a grade 3/6 pansystolic murmur at the left lower sternal border. There was no tenderness over the abdomen or hepatosplenomegaly. No pedal edema was found.

The posterioranterior chest radiograph demonstrated cardiomegaly, pulmonary edema and pleural effusion (Figure 1). The electrocardiogram was normal. Complete blood cell count was within normal limits except hemoglobin 10.9 g/dL. Blood biochemistry showed no abnormalities. The TTE findings were dilated LA, moderate pericardial effusion, severe tricuspid regurgitation, pulmonary artery pressure 68 mmHg, a homogenous mass occupying 2/3 of the left atrium, and normal left ventricular systolic function (Figure 2). The LA mass markedly prolapsed into the left ventricular (LV) during diastole and impeded its inflow; myxoma was impressed. Because of the huge LA tumor complicating with acute pulmonary edema and pulmonary hypertension, the cardiac surgeon undertook the operation of removal of the LA tumor on the next day of the patient admission. Surprisingly, the tumor was found to extend from the right inferior pulmonary vein, and a solid lung tumor was disclosed intraoperatively. The LA tumor was excised and the pathology was metastatic leiomyosarcoma.

Figure 1. The chest radiography showed pulmonary edema and bilateral pleural effusions.

Figure 2. Transthoracic two-dimensional echocardiography showed a large tumor mass occupying left atrium (A), and it prolapsed into left ventricle during diastole (B).
The post-operative investigations of the patient revealed multiple metastases to liver, lungs, bones and mediastinal lymph nodes.

**DISCUSSION**

Uterine leiomyosarcoma is biologically aggressiveness and has propensity for early hematogenous spread. The peritoneum, abdominal cavity and lung are the most common sites of metastasis. Cardiac metastasis is relatively rare in uterine leiomyosarcoma. Leiomyosarcoma with cardiac metastasis has been reported in several cases. However, intracardiac extension through the pulmonary vessels has never been reported in pulmonary metastatic leiomyosarcoma. Other tumors, including primary pulmonary leiomyosarcoma, metastatic cervical carcinoma, metastatic choriocarcinoma, metastatic osteosarcoma, metastatic malignant fibrous histiocytoma, and metastatic chondrosarcoma with involvement of pulmonary veins and LA have been reported. Most of those cases presented symptoms of lung metastasis (cough and hemoptysis), symptoms of left heart failure (dyspnea and impaired exercise tolerance) and symptoms of arterial embolism (cerebral infarction or acute renal failure). There were reports of leiomyosarcoma extending into the right heart through the inferior vena cava with presentation of right heart failure.

Acute pulmonary edema was noted when the patient presented to the emergency department. The invasion of pulmonary vein and the LA may have developed progressively for several months, and the patient had no obvious symptoms of heart failure until cardiac decompensation. Early diagnosis may be difficult by clinical symptoms. TTE is a very useful diagnostic tool. It provides the differential diagnosis of LA tumors including vegetation, thrombi and primary or secondary cardiac tumors. The diagnostic challenge is differentiation of myxoma and nonmyxomatous neoplasms. It is often difficult. However, some features can help us. Most atrial myxomas arise from the interatrial septum in the region of the fossa ovalis, have inhomogenous echogenecity and are highly mobile. However, 25% of myxomas may originate away from the atrial septum. Furthermore, the observed extension of tumor from pulmonary vein more effectively excludes myxoma. Most of primary and secondary cardiac neoplasms involve either the pericardium or the myocardium. Intracavity metastases can be demonstrated by TTE, as space-occupying lesions, but it is often very difficult to differentiate between primary and secondary cardiac tumors. As a general rule, nonseptal origin and pulmonary vein extension of the tumor strongly indicate the possibility of a nonmyxomatous neoplasm.

Because of the acute development of CHF with pleural effusion in our case, her lung tumor was easily overlooked on the plain chest radiograph, especially as there were no clinical symptoms of pulmonary involvement before. Subsequent studies demonstrated multiple metastases of the tumor. Tumor grade and stage appear to be valid prognostic indicators for leiomyosarcoma of the uterus. Although complete pulmonary metastasectomy can improve the survival in pulmonary metastasis, the extrapulmonary metastasis and the aggressive pattern of tumor may not improve her survival. Adjuvant therapy including radiotherapy and chemotherapy seems to play a limited role, and provides no survival benefit. Treatment for these patients should be palliative until effective therapeutic modalities prove otherwise. Our case received regular follow-up after debulking surgery of uterus till 3 years ago and then lost further surveillance. This time, multiple metastases were found through the manifestation of acute pulmonary edema. In summary, we reported a case of pulmonary metastatic leiomyosarcoma with left atrial extension via the...
pulmonary vein with presentation of acute pulmonary edema. The TTE showed a mobile intracavitary left atrial mass mimicking atrial myxoma. The LA tumor was found extending through the pulmonary vein during the operation. Careful assessment of the origin of intracardiac tumor with a TTE or transesophageal echocardiography is indispensable. More importantly, detailed history-taking and physical examinations are also essential for early diagnosis.

REFERENCES