The Heart Surgery Forum #2013-203
16 (6), 2013 [Epub December 2013]
doi: 10.1532/HSF98.2013203

Online address: http://cardenjennings.metapress.com

Left Atrial Leiomyosarcoma Extending into the Posterior Mediastinum and Mimicking a Left Atrial Myxoma

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ABSTRACT

Background: Intracardiac malignancies are extremely rare and hard to detect or differentiate preoperatively.

Case Report: We present a 48-year-old female patient who was diagnosed primarily with left atrial myxoma and taken into emergency surgery. The tumor extended into the pulmonary veins and infiltrated the atrial endocardium, and the histopathologic diagnosis was leiomyosarcoma. The left atrial endocardium was successfully peeled off with the tumor and complete resection was achieved.

Conclusion: The possible malignant nature of intracardiac masses should be kept in mind, especially in middle-aged patients. The extent of the tumor must be determined in elective cases to establish the proper strategy for complete resection, which is the only chance of successful treatment for this lethal disease entity. Endocardial peeling is warranted for successful removal of the tumor mass in leiomyosarcoma.

INTRODUCTION

Primary cardiac tumors are rare, with an incidence of 0.0017% to 0.019% [Shanmugam 2006]. One-fourth of these tumors are malignant, and leiomyosarcomas constitute 8%-9% of all cardiac sarcomas [Malyshev 2006; Shanmugam 2006]. Although most primary sarcomas are located in the right heart chambers and major great vessels, half of all cases of leiomyosarcoma originate in the left atrium. We present a patient who was primarily diagnosed with left atrial myxoma obstructing the mitral orifice and was taken into emergency surgery. An infiltrating sarcomatous tumor in the left atrium was suspected during the operation and complete resection was achieved.

CASE REPORT

A 48-year-old female patient was admitted to our emergency clinic with acute onset of severe dyspnea and orthopnea, nonproductive cough, and palpitations. Laboratory values were essentially normal, with a sedimentation rate of 31 mm at 1 hour. Echocardiographic imaging revealed a prominent mobile left atrial mass causing obstruction at the mitral valve, which was diagnosed primarily as a left atrial myxoma (Figure 1A). The patient was taken into emergency surgery under cardiopulmonary resuscitation because of severely elevated pulmonary arterial pressure (110 mmHg) due to obstruction of the mitral orifice, which resulted in sudden cardiac arrest. The need for the patient to undergo resuscitation precluded us from further investigations before the operation.

The operation was performed through a median sternotomy using standart techniques. Upon left atriotomy, a large mass was found to be filling the entire left atrial cavity (Figure 1B). The tumor was attached to the left atrial posterior wall between the right and left superior pulmonary veins (Figure 2A), and there were multiple smaller tumoral infiltrations of the endocardial lining and the posterior leaflet of the mitral valve (Figure 3A). Endocardial infiltrations extended into the right pulmonary veins and left superior pulmonary vein, obstructing their orifices. Thickened and infiltrated left atrial endocardium was peeled off completely along with the mass on the mitral valve (Figure 3B), and the posterior atrial wall was resected at the site of tumor attacment because of the suspicion of infiltration at this site (Figure 2B). The resultant defect was repaired primarily with a 4/0 polyproplene suture. Infiltrations at the orifices of the pulmonary veins were removed successfuly. The left atrial appendix was amputated and the defect was repaired primarily. After removal of the mass, mitral valve competence was confirmed by saline injection into the left ventricle. The left atriotomy was closed primarily. Perioperative transesophageal echocardiography showed a competent, normal functioning mitral valve without any intracardiac residual mass.

Histopathologic investigation revealed spindle cells arranged in interweaving fascicles. Nuclear atypia and geographic necrosis were easily identified. The tumor had 43 mitoses per 10 high-power fields (Figure 4A). Immunohistochemical study showed focal cytoplasmic positivity for desmin and diffuse strong cytoplasmic immunoreactivity for smooth muscle actin (SMA) (Figure 4B). CD34, CD99, and

Received July 10, 2013; accepted October 8, 2013.

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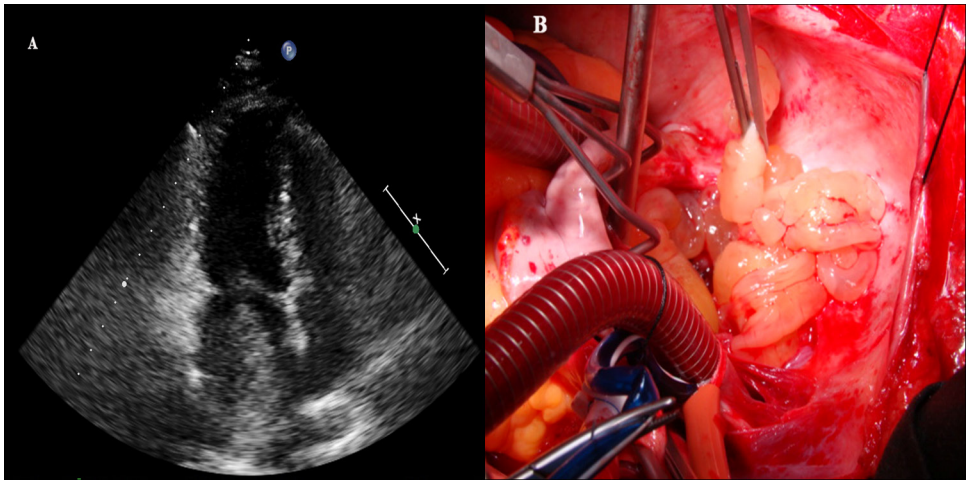

Figure 1. A, Transthoracic echocardiographic image of the patient showing a mobile giant mass in the left atrial cavity with obstruction at the mitral orifice. B, Intraoperative view presenting the emergent giant tumor mass just following the left atriotomy.

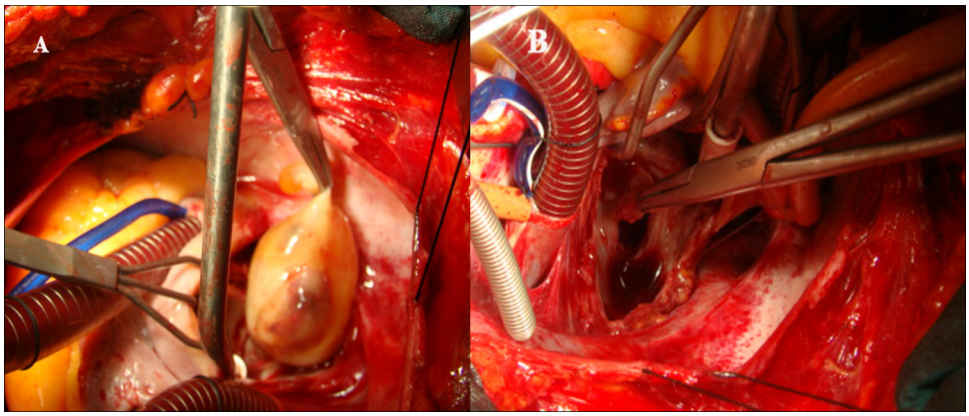

Figure 2. A, Tumoral invasive attachment to the left atrial posterior wall between the right and left superior pulmonary veins (intraoperative view). B, The resultant defect of the posterior atrial wall following resection at the site of tumor attachment (intraoperative view).

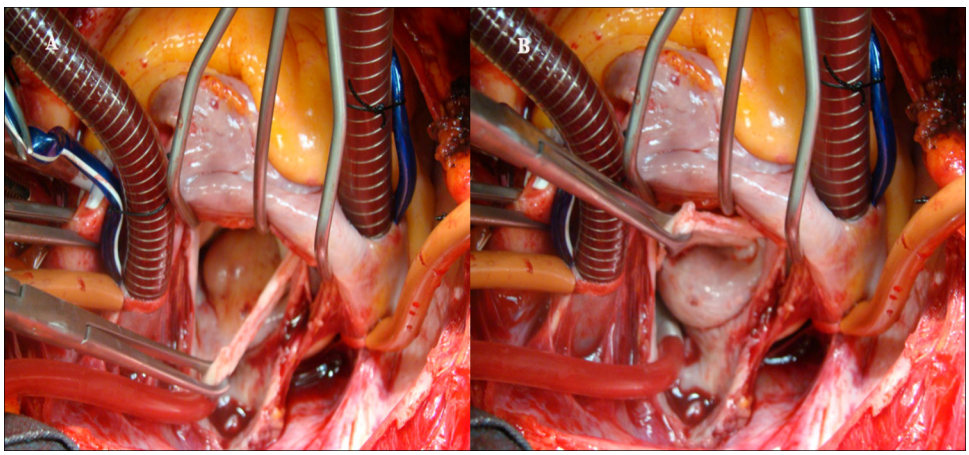

Figure 3. A, Multiple tumoral infiltrations of the endocardial lining and a bigger mass on the posterior leaflet of the mitral valve (intraoperative view). B, Thickened and infiltrated left atrial endocardium was peeled off completely along with the mass on mitral valve (intraoperative view).

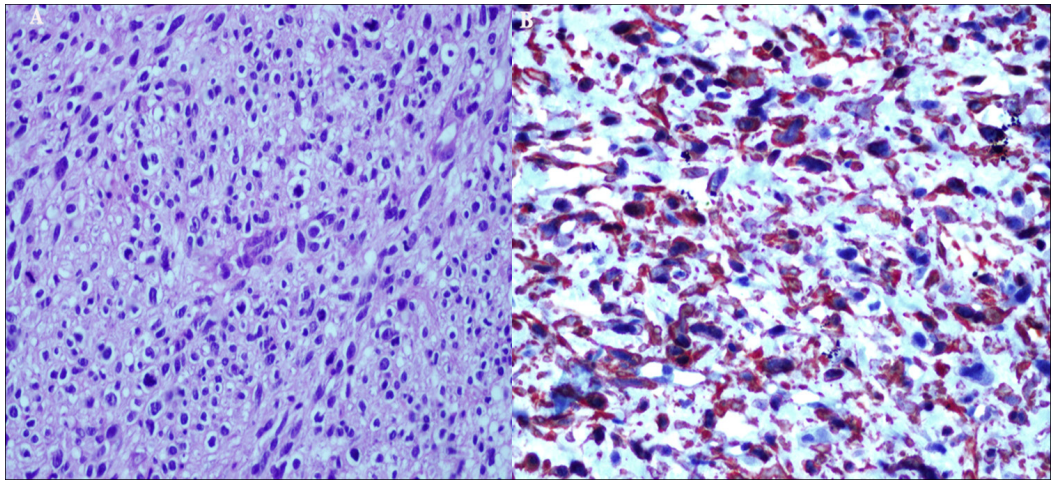

Figure 4. A, Spindle cells with abundant mitosis (hematoxylin and eosin ×200). B, Diffuse strong SMA immunopositivity (SMA ×400).

MYOD1 were negative. The patient was discharged from the hospital on the 12th postoperative day after an uneventful recovery. Further investigation with cardiac magnetic resonance imaging (MRI) showed a 7-cm × 6.5-cm mass in the posterior mediastinum with no signs of residual intracardiac tumor (Figure 5).

The patient was referred to thoracic surgery and oncology clinics for further interventions and chemotherapy. Six months later, following adjuvant chemotherapy, the mass in the posterior mediastinum was successfully removed with a second operation by the thoracic surgeon. She is now well and free of any relapse in her 3 years of follow-up.

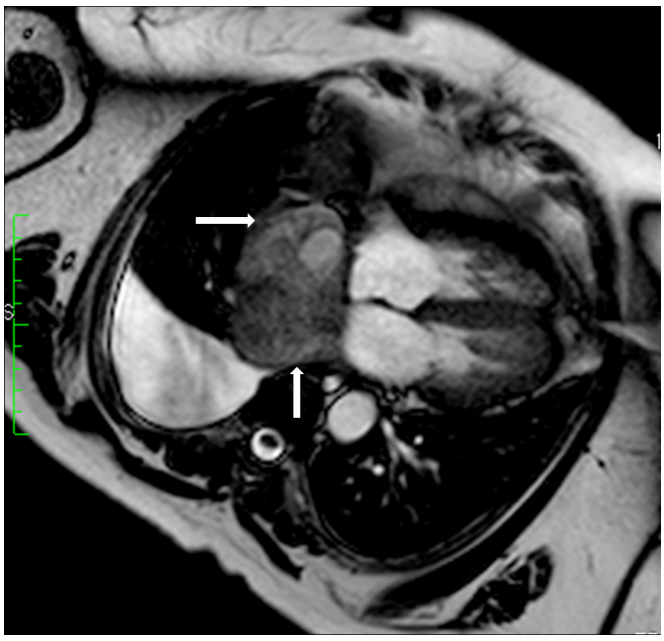

Figure 5. Postoperative cardiac MRI of the patient showing a posterior mediastinal mass (arrows) with no signs of residual intracardiac tumor.

COMMENT

Leiomyosarcoma of the left atrium and pulmonary veins is extremely rare, and 17 cases of leiomyosarcoma of the pulmonary veins and 20 cases of leiomyosarcoma of the left atrium have been reported [Malyshev 2006]. The symptoms of leiomyosarcoma of the left atrium have no specific features and are similar to those of myxoma [Malyshev 2006; Mazzola 2006] in relation to their mass effect. Echocardiographic distinction between leiomyosarcoma and myxoma is also difficult [Malyshev 2006; Mazzola 2006]. Although further investigations may help to distinguish the 2 entities, our case required emergent surgery due to unstable hemodynamics and sudden cardiac arrest related to the obstruction of the mitral orifice. Postoperative MRI control, to detect possible extention of the tumor, revealed a posterior mediastinal mass which was thought to be the primary site of origin. Combined pulmonary and cardiac involvement of leiomyosarcomas necessitates careful planning and an appropriate operative strategy to achieve complete resection, which is the best chance of cure for these patients [Korst 1999; Kono 2000].

Because the tumor had infiltrated left atrial endocardium, it was peeled off to achieve complete resection of the tumor at sites where the left atrial myocardium showed no evidence of invasion. Similar smooth dissections of the endocardium have been described previously [Mazzola 2006]. We believe that with such an extensive tumor mass filling almost the entire atrial cavity, this method ensures a complete resection and also makes it possible to differentiate sites of infiltration of the underlying myocardium. At sites of infiltration, resection of the atrial wall, if possible, will ensure complete resection.

Although it is difficult to differentiate between myxoma and a malignant mass in the left atrium, we should bear in mind the suspicion of a possible malignant nature of a cardiac tumor detected in middle-aged patients. Acute onset of symptoms, lack of systemic symptoms associated with myxoma, and extensive tumor mass may indicate the malignant nature of the tumor. Because the only acceptable treatment option in sarcoma is wide surgical resection, preoperative complete

detection of the tumor extent by means of echocardiography and MRI must be performed to develop proper surgical strategies for complete resection. Even if the diagnosis has not been made preoperatively but a malignant tumor is suspected during the operation, as in our case, every effort should be made to achieve this objective. The endocardial peeling technique allows successful and complete removal of the tumor mass in leiomyosarcoma, as in this case.

REFERENCES

Kono T, Takemura T, Hagino I, Matsumura G. 2000. Complete resection of cardiac leiomyosarcoma extending into the pulmonary trunk and right pulmonary artery. Ann Thorac Surg 70:1412-4.

Korst RJ, Rosengart TK. 1999. Operative strategies for resection of pulmonary sarcomas extending into the left atrium. Ann Thorac Surg 67:1165-7.

Malyshev M, Safuanov A, Gladyshev I, Trushyna V, Abramovskaya L, Malyshev A. 2006. Primary left atrial leiomyosarcoma: literature review and lessons of a case. Asian Cardiovasc Thorac Ann 14:435-40.

Mazzola A, Spano JP, Valente M, et al. 2006. Leiomyosarcoma of the left atrium mimicking a left atrial myxoma. J Thorac Cardiovasc Surg 131:224-6.

Shanmugam G. 2006. Primary cardiac sarcoma. Eur J Cardiothorac Surg 29:925-32.