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Leiomyosarcoma Protruding into the Left Ventricle during Diastole: Report of a Case

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ABSTRACT

Cardiac leiomyosarcoma is a rare tumor with poor survival prospects. Surgery prolongs survival, but the tumor often recurs early after surgery. The diagnosis is often made by transthoracic echocardiography. Magnetic resonance imaging and computed tomography are required to characterize the location and extent of cardiac masses. In this report, we present a patient with a leiomyosarcoma that was resected completely. The tumor was located in the left atrium, mimicked a myxoma, and protruded into the left ventricle during diastole.

CLINICAL SUMMARY

A 41-year-old woman was admitted to the hospital with severe dyspnea. On admission, she had a regular pulse of 110 beats/min and a blood pressure of 108/67 mm Hg. Hepatomegaly was noted. Leukocytosis was observed, but her blood chemistry results were within normal limits. A chest radiograph showed consolidation of the left middle lung field with pleural effusion. A sputum culture revealed normal flora with no malignancy. A transthoracic echocardiogram showed a large (12 × 5 cm) mobile mass within the left atrium that obstructed the mitral valve orifice during diastole and caused severe tricuspid valve insufficiency (Figure 1). Emergency surgery was performed because of the worsening dyspnea and orthopnea. Left and right atriotomies were performed with a transverse incision and the patient on cardiopulmonary bypass. The tumor appeared in the atrial incision (Figure 2). The mass obstructed the mitral valve orifice without infiltrating the leaflets. It originated from the left lateral wall between the right superior pulmonary veins. The tumor was resected along with the underlying endocardium and myocardium of the atrial wall. After resection of the tumor, a De Vega annuloplasty of the tricuspid valve was performed.

A gross examination revealed an ovoid mass measuring 13 × 8 × 5 cm and with a slightly irregular surface. A histopathologic examination showed vascular structures with oval cells and pleomorphic nuclei (Figure 3). An immunohistochemical examination showed cells positive for vimentin, desmin, and smooth muscle actin and negative for myogenin. A diagnosis of leiomyosarcoma was made. The patient was treated with anthracycline-based chemotherapy (doxorubicin, ifosfamide, and cisplatin every 15 days for 3 months). At 6 months after

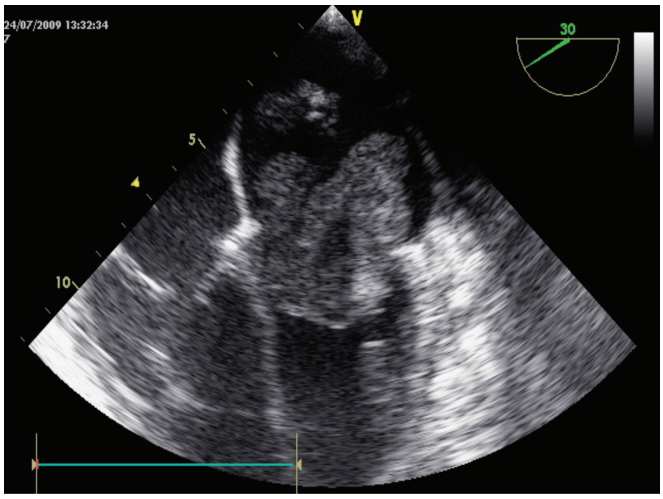

Figure 1. Transthoracic echocardiogram showing the tumor protruding into the left ventricle during diastole.

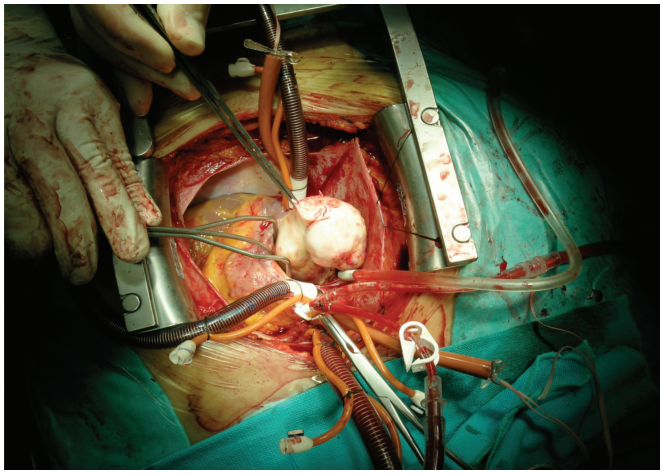

Figure 2. The large tumor appearing in the left atriotomy.

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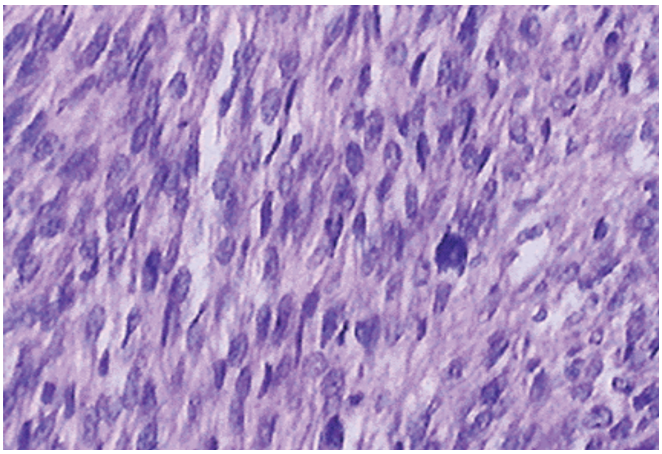

Figure 3. A histopathologic examination revealed vascular structures with oval cells and pleomorphic nuclei.

surgical resection, the patient was well with no evidence of local recurrence according to transesophageal echocardiographic and magnetic resonance imaging evaluations.

DISCUSSION

Primary cardiac neoplasms are rare, with an incidence of 0.0017% to 0.033% [Silverman 1980]. One fourth of all primary cardiac tumors are malignant, and almost all of them are sarcomas [McAllister 1978]. The outcomes for cardiac sarcomas are very poor, with a median survival time of less than 1 year; less than 15% of patients experience long-term survival [Molina 1990]. Leiomyosarcomas represent only 1% of such cardiac tumors [McAllister 1978]. Leiomyosarcomas are known to originate from the smooth muscle of the cardiac vasculature or the atrial endothelium [James 1989]. The clinical presentation is nonspecific, but the symptoms of left atrial leiomyosarcoma are generally related to obstruction of the left atrial cavity, the pulmonary veins, or the mitral valve orifice [Antunes 1991; Babatasi 1998]. The extension of the tumor to the pulmonary veins often makes pneumonectomy necessary to obtain a radical resection [Evans 2003]. In our patient, the tumor was attached to the endocardial surface without deep infiltration, thus making its complete removal feasible, as reported by Evans and Haw [2003]. Leiomyosarcomas grow rapidly and show local invasion and metastasis. Surgical resection is essentially palliative [Pessotto 1997]. Even in the case of excision of the tumor with clear margins, the prognosis is poor. Most patients die within 1 year of diagnosis [Fyfe 1991; Pessotto 1997]. Postoperative radiotherapy following complete resection has the potential to reduce the rate of local recurrence, but radical doses cannot be administered because of the risk of radiation cardiomyopathy, which can be fatal [O'Sullivan 2002].

In our patient, we believe the tumor was removed completely. There was no evidence of metastasis or recurrence in the early postoperative period. The patient was treated with anthracycline-based chemotherapy to prevent recurrence. This report supports the strategy of a multimodality treatment of these tumors to prolong survival and to achieve a good quality of life, as reported by Pessotto et al [1997].

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