

Metastasizing Primary Atrial Leiomyosarcoma Causing a Functional High-Grade Mitral Stenosis

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ABSTRACT

We report the case of a 43-year-old man who presented with a primary cardiac leiomyosarcoma and multiple metastases. Despite the severely poor prognosis, cardiac surgery was performed as part of a multidisciplinary palliative approach, which paved the way for further chemotherapy and radiation therapy.

INTRODUCTION

With a prevalence rate of 0.001% to 0.28%, primary cardiac tumors are very rare compared with cardiac metastasis [McAllister 1978; Neragi-Miandoab 2007]. Primary cardiac leiomyosarcomas are particularly rare. Approximately 25% of primary heart tumors are malignant, with 75% of such tumors being sarcomas [Vander Salm 2000]. The most common types of cardiac sarcomas are angiosarcomas (33%), rhabdomyosarcomas (21%), malignant mesotheliomas (16%), and fibrosarcomas (11%) [McAllister 1978; Vander Salm 2000]. Because of their limited anatomic structures, the required radical surgery often cannot be performed, thus leading to their poor prognosis.

CASE REPORT

A 43-year-old man was admitted to the hospital because of paravertebral back pain and recurrent exercise-related dyspnea. An echocardiography evaluation and a computed tomography scan showed a large left atrial tumor (59 × 27 × 32 mm; Figure 1, A-C) causing a high-grade functional mitral valve stenosis. An ultrasonography examination of the abdomen revealed an obscure tumor of the left kidney. An interventional biopsy was performed. A histopathologic examination of the tumor revealed a metastasis of a myogenic sarcoma. A computed tomography scan showed

multiple pulmonary, renal, pancreatic, and hepatic metastases. In addition, a bone metastasis in thoracic vertebra 10 could be demonstrated intruding into the spinal canal. Laboratory tests revealed an increased concentration of neuron-specific enolase (36 ng/mL).

Despite the poor prognosis suggested by these results, this young patient was admitted to our cardiac surgery department for evaluation of the surgical feasibilities in this

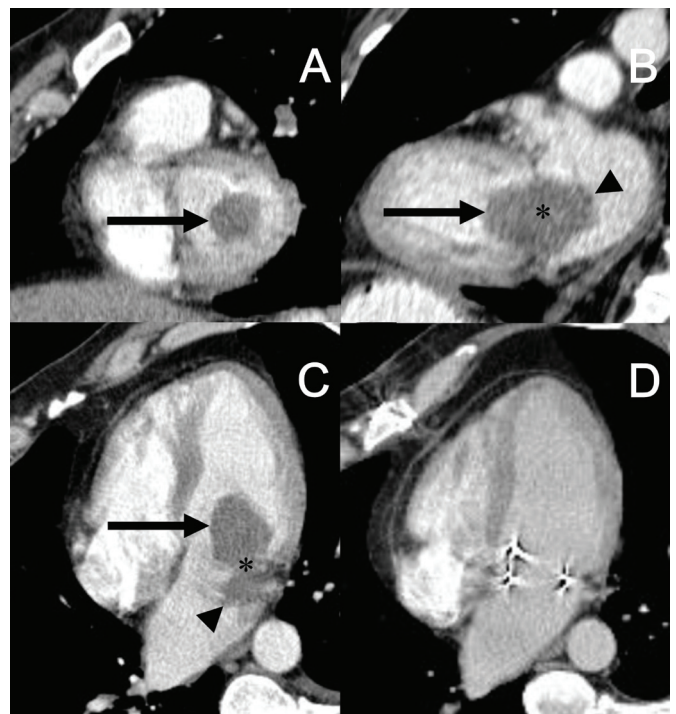


Figure 1. Computed tomography scan of the chest (A, B, C). Preoperative scan shows a large tumor originating from the posterior mitral leaflet (C) and prolapsing from the left atrium into the left ventricle (A, B). D, Postoperative control after 3 months, demonstrating a typical location for the bioprosthesis, with no signs of local relapse of the tumor (Somatom Sensation 16; Siemens, Erlangen, Germany) (axial 3-mm layer; 80 mL Solustrast 300 and 50 mL NaCl intravenously; 35-second delay; collimation, 16 × 1.5 mm; 120 kV). The left ventricular part of the tumor (arrows), the left atrial part of the tumor (arrowheads), and the mitral valve level (asterisks) are indicated.

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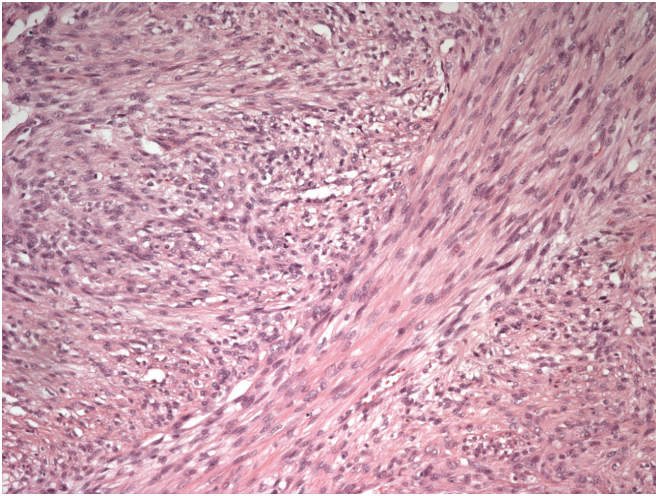


Figure 2. Histologic aspect of a high-grade leiomyosarcoma with a low grade of differentiation (hematoxylin and eosin, original magnification 10).

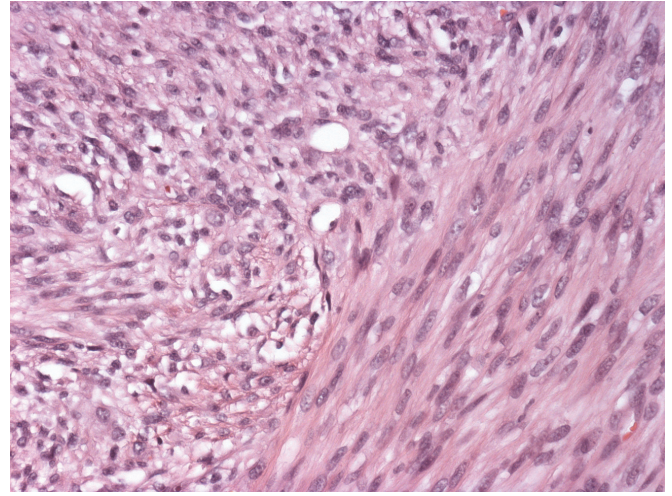


Figure 3. Histologic aspect of a high-grade leiomyosarcoma with a low grade of differentiation (hematoxylin and eosin, original magnification 40).

palliative situation. The local tumor board advised initiating a palliative chemotherapy and radiation therapy regimen only if resection of the cardiac tumor masses were possible.

Complete resection of the tumor was feasible via a left atrial approach. We supposed the adhesion point to be the posterior mitral leaflet (P3). After excision of the mitral valve, including the subvalvular apparatus, we replaced it with a biologic mitral valve (Hancock 29-mm valve; Medtronic, Minneapolis, MN, USA). The histopathologic examination of the resected tissue identified the tumor as a high-grade leiomyosarcoma with a low grade of differentiation (Figures 2 and 3). Histopathologic examination confirmed that the tumor had been resected in toto. All of the diagnostic findings and the biopsy examination of the metastases suggested that the cardiac tumor was the primary cancer.

After an uneventful intraoperative and early postoperative course, the patient developed acute and progressive paraplegia of the legs due to the vertebral metastasis. The patient was referred for emergency radiation therapy. After 3 months of follow-up, a computed tomography scan ruled out a local relapse of the tumor (Figure 1D). An echocardiography demonstrated regular regular prosthetic valve function. After approximately 9 months, the patient died from secondary metastases during the subsequent follow-up period.

DISCUSSION

In contrast to primary cardiac tumors, metastases usually involve multiple areas of the myocardium [Fine 1968; McAllister 1978]. Cardiac tumors themselves are associated with clinical symptoms in only approximately 10% of cases. Functional valve obstruction, as in the present case, is a frequent complication [Molina 1990]. Approximately 2% to 5% of solid malignant tumors present as a metastatic cancer of unknown primary site (CUP syndrome) [Moll 2005]. An intraoperative pathology evaluation in the present case and subsequently performed histopathologic examinations of

the suspected primaries and its metastases led to the diagnosis of a primary cardiac sarcoma, which is a real rarity. The present case lucidly demonstrates the palliative situation and poor prognosis of sarcomatous tumors. Despite the local extension of the primaries, cardiac surgery could be performed with good results—even during follow-up. Treatment during the subsequent clinical course of the patient was directed mainly by the clinical effects of the secondary metastases.

Patients with cardiac tumors remain a challenge for the cardiac surgeon. Because of the cardiac location, the principle of radical resection of the tumor is often not applicable, and the procedure maintains mostly a palliative character. Preoperative imaging plays an essential role in determining the surgical strategy. In the present case, cardiac surgery became an essential part of the treatment strategy of a metastatic cancer disease. A multidisciplinary approach is indispensable for gaining the best possible outcomes.

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