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Cardiac Angiosarcoma with Reconstruction of the Right Atrium

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ABSTRACT

We describe a case of a 65-year-old female patient who presented with shortness of breath when bending forward. A tumor highly suspicious for an angiosarcoma was identified by computed tomography and was suggestive of infiltration into the superior vena cava (SVC) and the majority of the right atrium. Surgical intervention on such an infiltrative tumor would not have been indicated. Magnetic resonance imaging, however, demonstrated that the tumor was not infiltrating the SVC and that its extension into the right atrium caused a low flow phenomenon. Surgical removal of the tumor and reconstruction of the right atrium with bovine pericardium was performed successfully. Adjuvant treatment with chemotherapy and radiation was performed afterward. The patient is free from tumor recurrence 3 months postoperatively.

INTRODUCTION

Primary cardiac tumors are rare: the autopsy incidence is 0.0001% to 0.0003%. Incidental cardiac tumors are found in approximately 1 out of 500 cardiac surgical cases. Only 25% of primary cardiac tumors are malignant, of which three-quarters are sarcomas [Bakaeen 2003; Blackmon 2009a].

Right-heart sarcomas especially tend to be asymptomatic until a very late stage of the disease. These tumors are often bulky, infiltrative, and metastasize early. Their bulk often extends to the outside of the heart, and it is uncommon for patients to develop right-heart failure. The prognosis of patients with cardiac angiosarcomas is poor. Ninety percent of patients described in studies involving medical therapy alone were dead within 9 to 24 months. Surgical series of primary cardiac sarcoma are rather small. These series describe median survival times of 12 to 17 months in cases of complete resection [Simpson 2008; Blackmon 2009a].

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CASE REPORT

We describe a 65-year-old female patient who presented with shortness of breath and globus pharynges that were exacerbated by bending forward. Routine physical evaluation and ECG did not reveal any abnormalities. A chest x-ray revealed a massively enlarged cardiac silhouette, caused by a pericardial effusion, demonstrated on echocardiography.

A computed tomography (CT) scan was performed, and a large tumor of the right atrium was found, which was highly suspicious for an angiosarcoma. According to this CT scan, the tumor occupied the majority of the right atrium and appeared to extend 5 cm into the superior vena cava (SVC) (Figure 1). Such an advanced stage of the tumor with infiltration into the SVC would not have been deemed an indication for surgery. Nevertheless, magnetic resonance imaging (MRI) was performed, demonstrating that the tumor was not, in fact, infiltrating the SVC, but rather that the volume of the tumor in the right atrium caused a low flow phenomenon of the blood entering the SVC (Figure 2).

Because the patient was highly symptomatic, the decision was made to operate immediately. After median sternotomy and bicaval cannulation for extracorporeal circulation,

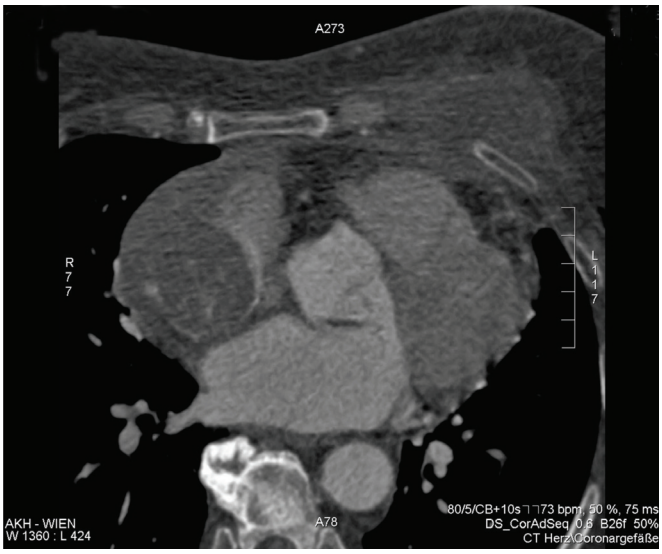

Figure 1. A computed tomography (CT) scan revealed a large tumor of the right atrium, which was highly suspicious for an angiosarcoma.

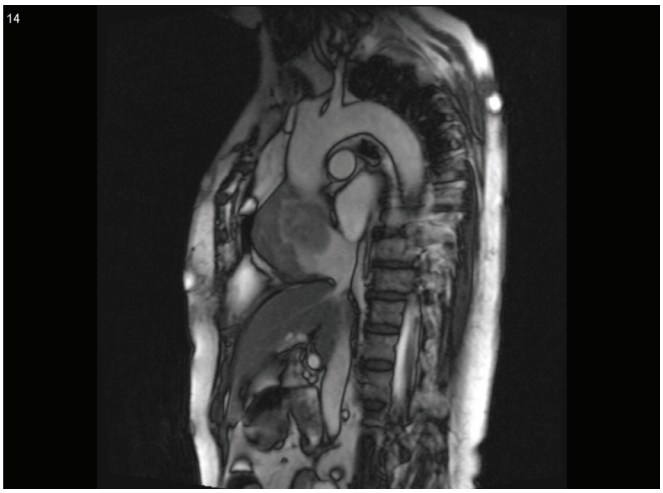

Figure 2. Magnetic resonance imaging (MRI) demonstrated that the tumor was not infiltrating the superior vena cava (SVC), but that the volume of the tumor in the right atrium caused a low flow phenomenon of the blood entering the SVC.

the right atrium was opened, and a huge tumor on the lateral wall of the right atrium was found infiltrating the fatty tissue close to the right coronary artery. Complete resection of the tumor was performed. Because of the large extent of the tumor, nearly the entire right atrium had to be resected with the atrial resection line close to the right coronary artery (RCA) and about 0.5 cm away from the insertion of the tricuspid valve. Bovine pericardium was used to reconstruct the right atrium. Intraoperative histological evaluation confirmed the diagnosis of an angiosarcoma. The tumor was completely resected macroscopically.

The patient was stable while in the intensive care unit (ICU) and was transferred to the cardiac surgical ward on the first postoperative day. On day 7 she was discharged from the hospital without any symptoms. In addition to surgical resection, the patient received adjuvant chemotherapy and radiation. Three months after the surgery, the patient remains asymptomatic and shows no signs of a tumor relapse.

DISCUSSION

Herein we describe a rare case of a cardiac angiosarcoma. The decision to proceed with surgical treatment of the tumor was made because the patient was highly symptomatic. Although the literature suggests that the prognosis of patients with angiosarcomas is poor, the operation was necessary to relieve the patient from those symptoms.

From the findings of the preoperative radiologic examinations, it seems to be important to perform not only a CT, but also MRI in cases of a cardiac mass. The fact that the tumor did not infiltrate the SVC but caused a low flow phenomenon due to its extension in the right atrium triggered the decision to perform the surgery.

The outcome of patients with angiosarcoma is generally poor, whether treated medically or surgically. Nevertheless, the patient with the longest reported survival after surgical removal of a cardiac angiosarcoma is alive 9.5 years after the intervention. In the literature, some authors recommend neoadjuvant chemotherapy for the treatment of cardiac angiosarcoma, but clear evidence is scarce [Bakaeen 2009; Blackmon 2009b].

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