The Heart Surgery Forum #2006-1216
10 (3), 2007 [Epub April 2007]
doi: 10.1532/HSF98.20061216

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

Right Atrial Angiosarcoma: A Case Report

Adlan Olsun, MD, Cagri Duzyol, MD, Ali Kemal Gur, MD, Mehmet Kaplan, MD, Remzi Tosun, MD

Siyami Ersek Thoracic and Cardiovascular Surgery Training and Research Hospital, Department of Cardiovascular Surgery, Istanbul, Turkey

ABSTRACT

A 32-year-old female patient presented with dyspnea and palpitation, and transthoracic echocardiography revealed the presence of pericardial effusion. Pericardiosynthesis was performed for drainage. Because of the rapid accumulation of effusion and the presence of a right atrial mass on follow-up echocardiography, a computed tomography scan was done that revealed a right atrial defect and the presence of advanced pericardial effusion. The patient was prepared for an emergency operation. The mass on the right atrial wall was approached via a midsternal incision with cardiopulmonary bypass. The tumor filled the right atrial cavity, compressed vital structures, extended to the right ventricle, and had local metastases. As the tumor did not appear to be curable with surgery, a palliative approach was adopted. The right atrial free wall and tissues causing cardiac obstruction were totally removed, the tumor itself was partially excised, and local metastases were sampled. The resulting right atrial wall defect was closed with a Dacron patch. The operation ended uneventfully, and the clinical status and vital and hemodynamic findings of the patient returned to normal. The pathological diagnosis based on the samples obtained during the operation was angiosarcoma. The patient had an uneventful postoperative period and was then referred to an oncology center for clinical recovery. No findings of local recurrence or metastases were observed during the postoperative follow-up. The patient completed her combination therapy and currently is free of any clinical problems at her 13th postoperative month. We believe that advancements in radiotherapy and chemotherapy regimes combined with surgery (radical, if possible) for the treatment of cardiac angiosarcomas may provide better survival and quality-of-life results.

Received December 20, 2006; received in revised form March 4, 2007; accepted March 15, 2007.

Correspondence: Mehmet Kaplan, MD, 67. Ada, Kardelen 4-4, D: 11, Atasehir, 34758 Kadikoy, Istanbul, Turkey; 90-216-542-44-86; cellular phone: 90-532-270-36-33; fax: 90-216-360-77-40 (e-mail: mehmetkaplan@superonline.com).

INTRODUCTION

Primary cardiac tumors are quite rare, with an incidence of 0.001% to 0.03% in autopsy series. Three quarters of cardiac tumors are benign, and approximately half are myxomas. Angiosarcoma is the most common among all the primary malignant cardiac tumors, with an incidence of 37%. It is usually diagnosed at an advanced or terminal stage. However, new methods allow earlier diagnosis. The patients are usually middle-aged men and present with intracardiac obstruction, pericardial or pleural effusion, and cardiac tamponade [McFadden 1997; Centofanti 1999]. We present a case of a right atrial angiosarcoma extending toward various cardiac localizations.

CASE REPORT

A 32-year-old female patient had been admitted to another center with a history of dyspnea and palpitation over the previous 15 days. Transthoracic echocardiography had revealed the presence of pericardial effusion and cardiac tamponade, and she had been referred to our clinic for further diagnostic procedures and treatment. In our institution, 1100 cc of serohemorrhagic fluid was drained from the patient by pericardiosynthesis, and the presence of cardiac tamponade was confirmed by echocardiography (Figure 1). During follow-up, the echocardiography revealed an increase in the amount of pericardial effusion, and spiral computed tomography showed a filling defect occluding the whole right atrial cavity and advanced pericardial effusion. Therefore, the patient was prepared for an emergency operation.

Operative Technique

During the operation, the mediastinum was reached by a midsternal incision. The pericardium was distended and approximately 1000 cc of serohemorrhagic fluid was drained as soon as it was opened. Macroscopic findings were: tumor tissue infiltrating the right atrial wall, filling the right atrial cavity, compressing the caval veins, and extending to the right ventricle, the presence of tumor nodules of various sizes over the right and left ventricles, bleeding foci over the ascending aorta, and gelatinous substances around the aorta and pulmonary artery. The tumor was not suitable for curative

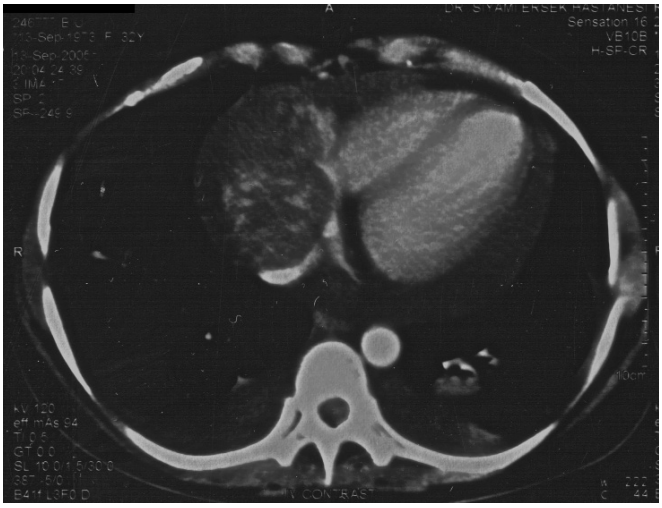

Figure 1. Preoperative computed tomography image of the right atrial mass.

surgery; restoration of the venous inflow and decompression of the atrium was planned.

Partial cardiopulmonary bypass was commenced with the cannulation of the ascending aorta with an arterial cannula, direct superior caval venous cannulation with a bended cannula, and placement of a venting cannula to the pulmonary artery. The right atrium was opened during a very brief period of inferior vena caval inflow occlusion and the provision of superior vena caval venous return was managed by controlling vena cava tapes; the second venous cannula was placed into the inferior vena cava. Total cardiopulmonary bypass was then commenced.

Cardiac arrest was achieved by 28°C systemic hypothermia, aortic cross clamping, antegrade potassium crystalloid cardioplegia, and local hypothermia. The tumor was infiltrating the interventricular septum and right ventricular wall (Figure 2). The free right atrial wall and the tissues causing obstruction were completely removed up to the caval veins. The resulting defect was closed with a Dacron patch. The tumor tissue could not be totally removed.

The operation ended uneventfully. Postoperatively, blood pressure was 125/75 mmHg, heart rate was 90 beats/minute with a sinusal rhythm, and central venous pressure was 14 mmHg. This hemodynamic status was also maintained in the postoperative intensive care unit.

Pathological Examination

Pathologically, the tumor was diagnosed as an angiosarcoma. First, the samples were fixed with 10% formaldehyde and then placed in paraffin blocks and slices of 4 to 5 microns thickness were obtained. Hematoxylin-eosin stained samples were examined under a light microscope. The histopathological view led us to consider the tumor to be a high-grade malignant mesenchymal tumor. Immunohistochemical examination was used for differential diagnosis. Positive CD31 and CD34 staining were shown for tumor cells, which confirmed the diagnosis of angiosarcoma (Figure 3).

Follow-up

The postoperative course of the patient was uneventful, and she was discharged for clinical recovery and referred to an oncology clinic. No pleural effusion was evident and no space-occupying lesion was found inside the cardiac spaces on follow-up echocardiography done at the 1st, 2nd, and 13th postoperative months. Currently, the patient is in her 13th postoperative month and her radiotherapy and chemotherapy have been completed and she has Class I functional capacity.

DISCUSSION

One fourth of the primary cardiac tumors have malignant potential. Most of them (approximately 75%) are angiosarcomas. The clinical properties of primary cardiac tumors are determined by their localization, size, and metastastic potential [Centofanti 1999]. Angiosarcomas generally originate from the right side of the heart (68%), usually from the right atrium and rarely from the right ventricle. As the tumor usually involves the surrounding pericardium, the diagnosis is often confused with pericardial inflammatory diseases and correct diagnosis is usually delayed [McFadden 1997].

More than half of the patients present with precordial pain decreasing when in the upright position, fever, cough, and cachexia, thus most cases are considered to be tuberculosis or viral pericarditis. Because of the massive pericardial effusion present in our patient, although no other signs of infection were present, inflammatory pathologies were considered for the differential diagnosis. A chest x-ray generally shows diffuse cardiac enlargement as in pericardial effusion, whereas only the image of a mass that results in a prominent shape on the right heart border may be found for some patients. In our patient, the typical tent-heart appearance was seen without any mass image [Centofanti 1999].

Echocardiography aids in diagnosing cardiac tumors. Transthoracic echocardiography can be used to determine the localization, size, shape, attachment site, and mobility of the

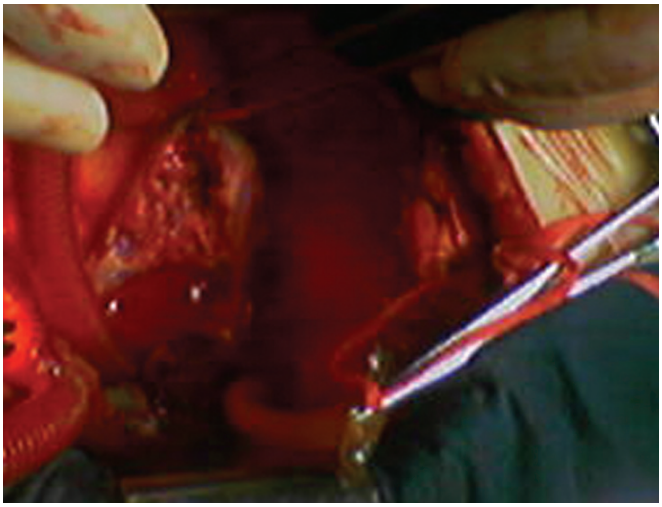

Figure 2. Intraoperative view of the tumor.

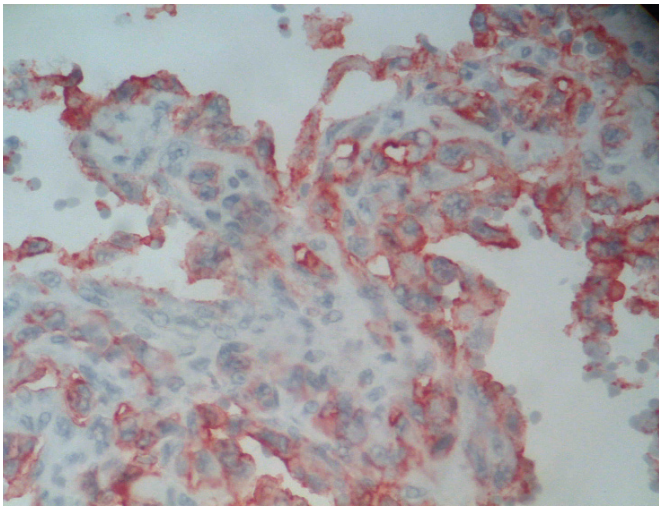

Figure 3. Microscopic view of the mass (immunohistochemical examination, positive CD31 and CD34 staining, original magnification ×400).

tumor. Transesophageal echocardiography is partially beneficial in determining the settlement site of the tumor [Centofanti 1999; Sakaguchi 2006]. In our patient, echocardiography was the main method to establish the diagnosis at an early stage. Angiography was not performed in our patient because of the absence of any previous cardiac complaints, her young age, and her emergency status.

Malignant cardiac tumors present with a rapid hemodynamic deterioration and advance progressively. The primary results of intramural enlargement are cardiac arrhythmias and conduction abnormalities. However, no rhythm or conduction disorders were detected in our patient, neither at the time of diagnosis nor during the follow-up periods. Intracavitary enlargement results in primary obstructive symptoms usually causing mechanical hemolysis and tumor embolization. Obstructions on the tricuspid valve may present, as well as serious compression on caval veins, resulting in face and upper extremity edema and serious visceral congestion [Rossi 1976; Sakaguchi 2006]. Most of the patients present with at least one component of the embolism, intracardiac obstruction, and constitutional symptoms triad [Centofanti 1999]. In our case, findings secondary to restriction of the cardiac preload were found.

The mean duration of survival is 3 to 6.6 months for primary cardiac tumors, regardless of the histological diagnosis. Our patient is still free from clinical problems at her 13th postoperative month. Sixty-six percent to 89% of tumors metastasize. However, angiosarcoma is not responsive to irradiation or chemotherapeutical agents [McFadden 1997]. For that reason, early and complete resection before the development of metastases is the main therapeutic choice for cure or palliation, as it is for all patients with cardiac neoplasia. Therapeutic regimes without the addition of surgery are generally unsuccessful. A tumor excision from the left atrium without using extracorporeal circulation has been reported and this patient lived for 3 years, representing the longest survival reported so far [Rossi 1976].

Despite aggressive interventions during preoperative and postoperative periods, most of the patients deteriorate within 9 months after surgery and long-term survival is rare. Common characteristics of patients who are symptom free for more than 34 weeks and who survive long term are: extended surgical resection together with adjuvant multidisciplinary therapy and no systemic metastases at the time of operation. The rapid growth and early metastasis potential of angiosarcomas emphasize the importance of early diagnosis and immediate application of an aggressive therapeutic strategy [Nakamichi 1997; Kurian 2006].

CONCLUSION

Although surgery is the primary therapeutic choice for primary malignant cardiac tumors, surgical resection is usually avoided during operation for patients with large tumors involving vital cardiac structures. During surgery, the usage of reconstructive techniques protecting cardiac tissues that do not have a relation with the tumor may prevent the need for prosthetic valve replacement and may result in acceptable palliation. The most suitable ones are tumors involving the right heart. Assuring the competence of the right atrioventricular valve provides circulatory balance or prevents right cardiac failure.

REFERENCES

Centofanti P, Di Rosa E, Deorsola L, et al. 1999. Primary cardiac tumors: early and late results of surgical treatment in 91 patients. Ann Thorac Surg 68:1236-41.

Kurian KC, Weisshaar D, Parekh H, Berry GJ, Reitz B. 2006. Primary cardiac angiosarcoma: case report and review of the literature. Cardiovasc Pathol 15:110-2.

McFadden PM, Ochsner JL. 1997. Atrial replacement and tricuspid valve reconstruction after angiosarcoma resection. Ann Thorac Surg 64:1164-7.

Nakamichi T, Fukuda T, Suzuki T, Kaneko T, Morikawa Y. 1997. Primary cardiac angiosarcoma: 53 months survival after multidisciplinary therapy. Ann Thorac Surg 63:1160-1.

Rossi NP, Kioschos JM, Aschenbrener CA, Ehrenhaft JL. 1976. Primary angiosarcoma of the heart. Cancer 37:891-4.

Sakaguchi M, Minato N, Katayama N, Nakashima A. 2006. Cardiac angiosarcoma with right atrial perforation and cardiac tamponade. Ann Thorac Cardiovasc Surg 12:145-8.