

Cardiac Angiosarcoma with Right Atrial Perforation and Cardiac Tamponade

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Primary cardiac angiosarcoma is a rare tumor associated with a poor prognosis. We report a case of a 59-year-old woman with right atrial angiosarcoma presenting with cardiac tamponade due to right atrial perforation. She underwent urgent surgical resection of the tumor. However, the patient died 68 days after surgery due to local recurrence. An effective treatment for cardiac angiosarcoma has not yet been established. However, more aggressive treatment with a combination of surgery, radiation, chemotherapy and IL-2 should be considered. (Ann Thorac Cardiovasc Surg 2006; 12: 145–8)

Key words: cardiac angiosarcoma, cardiac tumor, tamponade

Introduction

Primary cardiac angiosarcoma is a rare tumor that is associated with a poor prognosis.

Since angiosarcoma is essentially not responsive to current regimens of chemotherapy and irradiation, surgical resection may be the treatment of first choice. However, complete resection of cardiac angiosarcoma is often difficult due to the limited amount of myocardium and expansion of the tumor at the time of diagnosis. We report a case of right atrial angiosarcoma that presented with cardiac tamponade due to right atrial perforation.

Case Report

A 59-year-old woman was admitted to Fukuoka Tokusukai Hospital with severe chest and back pain in October 2003. On admission, the patient was alert but was in a state of shock with a systolic pressure of 80 mmHg. A chest X-ray showed mild cardiomegaly.

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An electrocardiogram showed normal sinus rhythm with a heart rate of 91/min.

Transthoracic echocardiography indicated the presence of a right atrial mass with normal left ventricular function (ejection fraction 61%), and mild pericardial effusion. Transesophageal echocardiography showed a smooth surfaced mass in the right atrium, 3×2 cm in size. Computed tomography (CT) of the chest revealed a large hematoma compressing the right atrium and ventricle (Fig. 1A), and a tumor lesion with central necrosis in the right atrium (Fig. 1B). The diagnosis was a cardiac tumor in the right atrium and cardiac tamponade due to the rupture of the tumor. An urgent operation was performed on the day after admission.

Neither direct invasion of the tumor to the pericardium nor dissemination in the pericardium were detected during the operation. Following pericardiotomy, a large hematoma was found on the right atrium and ventricle.

The tumor originated in the right atrial free wall and extended into the right atrium as a floating tumor. A perforation of the right atrial appendage was also found (Fig. 2).

Under total cardiopulmonary bypass, the tumor was resected, along with as much of the right atrial wall as possible. The right atrium was repaired with a bovine pericardial patch. The resected tumor was 6.5×4.0×2.0 cm in size. Histologic examination of a frozen section was unable to distinguish angiosarcoma because a necro-

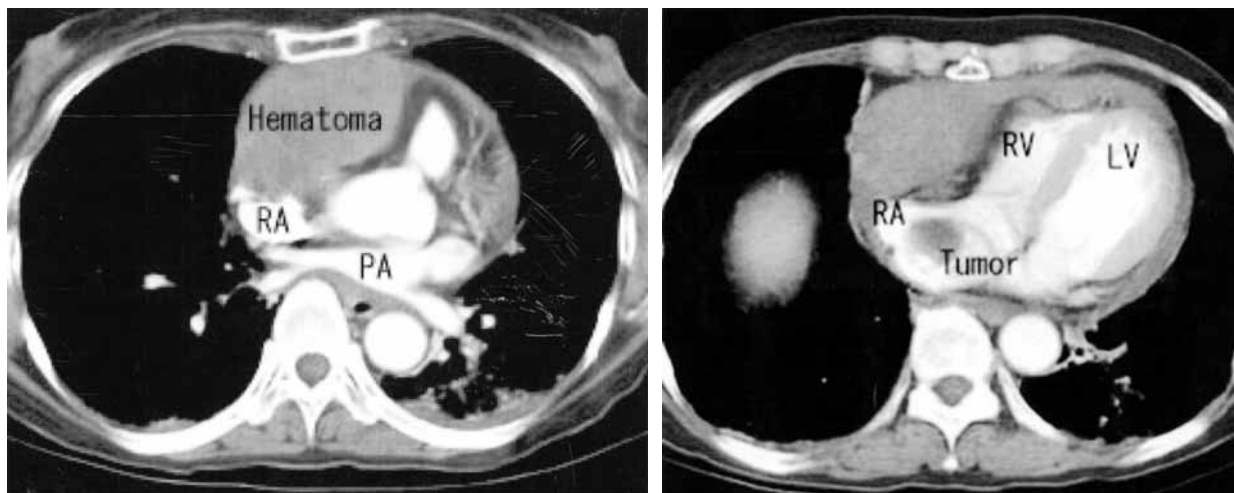


Fig. 1.
A: Chest CT presents a large hematoma compressing the right atrium and ventricle.
B: A mass lesion with central necrosis in the right atrium.
 RA, right atrium; PA, posterior-anterior; RV, right ventricle; LV, left ventricle.

A | B

tizing lesion occupied most of the tissue. The histological diagnosis was angiosarcoma. The section showed proliferation of malignant cells with short spindle or oval nuclei, displaying very high focal cellularity and differentiation from variably sized blood vessels (Fig. 3). Cells were stained well by factor VIII. The surgical margin was positive, although we resected the tumor with as much of the right atrial wall as possible.

The postoperative course was uneventful, and she was discharged 25 days after surgery.

However, CT revealed recurrence of the tumor in the right atrial wall 3 weeks after surgery. She was readmitted due to heart failure. Neither chemotherapy nor radiotherapy could be performed because of her poor general condition. The patient died 68 days after surgery.

Discussion

A primary tumor of the heart is rare with an incidence ranging from 0.028 to 0.05% in autopsy cases.¹⁾ In adults, approximately 75% of primary cardiac tumors are benign, with myxoma accounting for up to half of the cases. The remaining 25% of cardiac tumors are malignant, and one-third of those are angiosarcoma.²⁾ Two-thirds of angiosarcomas are located in the right atrium. Angiosarcoma is 2-3 times more common in males than females.

It is well documented that angiosarcoma shows rapid growth, local invasion and distant metastases. The most frequent sites of metastases are the pericardium, lungs,

mediastinal lymph nodes, and vertebrae. Metastases are present in 66-89% of the patients at the time of diagnosis.³⁾ It is difficult to diagnose cardiac angiosarcoma because there are no specific symptoms.

Cardiac rupture due to angiosarcoma is rare. Including our own case, less than 10 cases have been reported.⁴⁻¹⁰⁾ The treatment of angiosarcoma is controversial due to its poor prognosis. Surgical resection is usually indicated when there is no evidence of metastasis and when myocardial resection is reparative. Chemotherapy or radiotherapy may be indicated as adjuvant or preferential therapies; however, their use is usually limited due to the poor physical condition of the patient. Survival ranges from 6 to 9 months regardless of the treatment.

Nakamichi et al. reported a case diagnosed with angiosarcoma at 8 years of age and, treated with an intensive and multidisciplinary approach for 2 years.¹¹⁾ The patient had an excellent quality of life 53 months after the diagnosis.

Sinatra et al. reported that the combination of surgical resection and radiation reduced the mass and eliminated symptoms despite an incomplete resection.¹²⁾

Recently, cardiac transplantation has become an alternative therapy although the outcome is not improved.

Baay et al. reported a case of successful treatment of cardiac angiosarcoma combining these therapeutic modalities.¹³⁾ In that case; the patient underwent initial chemotherapy followed by a total radiation dose of 2,600 cGy, then transplantations. Two months later the patient



Fig. 2. A perforation of the right atrial appendage was found.

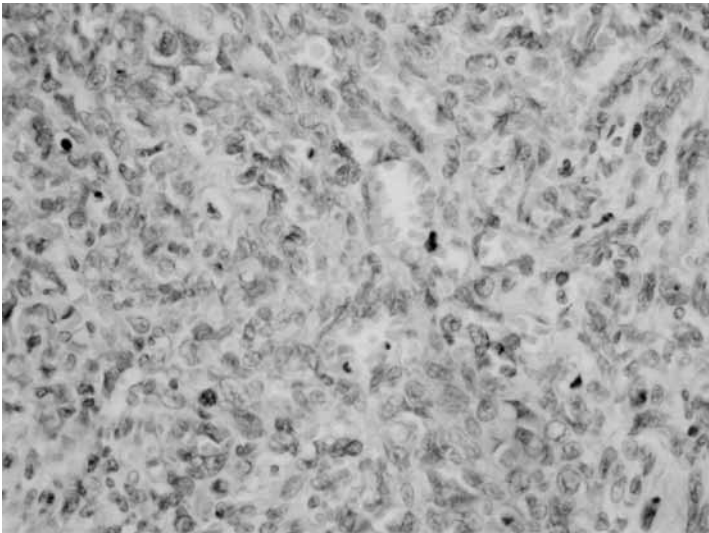


Fig. 3. A photomicrograph showed proliferation of malignant cells with short spindle or oval nuclei, displaying very high focal cellularity and differentiation from variably sized blood vessels. (HE×200)

received additional chemotherapy. The clinical outcome was favorable and metastases were not detected up to 33 months after surgery. Although successful cardiac transplantation has been reported, it is not common in Japan.

Interleukin (IL)-2, which activates T cells, lymphokine-activated cells and natural killer cells, has been reported to be effective for pulmonary angiosarcoma. Kakizaki et al. reported a case of cardiac angiosarcoma treated with a combination of chemotherapy and immunotherapy. The patient survived 30 months after surgery.¹⁴⁾

Combined treatment with surgery, radiation, chemotherapy and IL-2 may lead to long-term survival for cardiac angiosarcoma.

Conclusion

Postoperative survival does not significantly differ in patients treated with or without surgery, even when the surgical approach is transplantation. An ideal treatment for cardiac angiosarcoma has not yet been established. However, early diagnosis and more aggressive treatment with a combination of surgery, radiation, chemotherapy and IL-2 should be considered.

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