

CASE REPORT

Three-year survival in primary cardiac angiosarcoma

Naoto Fukunaga¹, MD, Takeshi Kitai², MD, PhD, Yukihiro Imai³, MD, PhD, Yutaka Furukawa², MD, PhD, and Tadaaki Koyama¹, MD, PhD.

Department of Cardiovascular surgery¹, Cardiology² and Pathology³ Kobe City Medical Center General Hospital, 2-1-1 Minatojima-minamimachi, Chuo-ku, Kobe, 650-0047, Japan

Abstract : A healthy 18-year-old girl was referred to our hospital for further evaluations of cardiac angiosarcoma. Transthoracic echocardiography showed an immobile 4.6 cm×3.7 cm cardiac mass. The mass was occupying right atrial chamber and partially, invading into annulus of tricuspid valve on transesophageal echocardiography. At surgery, the mass was seen to be protruding from right atrial appendage and adhering to right side of pericardium. The histological findings were consistent with cardiac angiosarcoma and immunological staining was positive for CD34 and CD31. Afterward, although she received radiochemotherapy, she died of metastasis of cardiac angiosarcoma more than three years after surgical resection. *J. Med. Invest.* 64 : 181-183, February, 2017

Keywords : survival, primary cardiac angiosarcoma, surgery

INTRODUCTION

Primary cardiac angiosarcoma is a rare, but life-threatening entity because it has aggressive characteristics and the prognosis after a diagnosis made is very poor. Patients who suffered from cardiac angiosarcoma usually do not complain about any symptoms unless the disease is advanced. Therefore, most cases are diagnosed incidentally or too late, subsequently resulting in the poor outcomes (1). Eighty percent of cardiac angiosarcoma present with metastasis with median survival of only six months (2, 3). Herein, we describe a case of primary cardiac angiosarcoma with a better survival following surgical resection and radiochemotherapy.

CASE REPORT

A healthy 18-year-old girl was referred to our hospital for further evaluations of cardiac angiosarcoma, which was diagnosed by a femoral transvenous biopsy. On admission, transthoracic echocardiography showed an immobile 4.6 cm×3.7 cm cardiac mass possibly originating from the right atrial appendage and no pericardial effusion. The mass was occupying right atrial chamber and partially, invading into annulus of tricuspid valve without tricuspid valve regurgitation on transesophageal echocardiography (Figure 1 A). Computed tomography showed that the mass was in the right atrial chamber and it was near the ascending aorta without evidence of invasion (Figure 1 B and C). Further evaluations including lung perfusion scan and magnetic resonance imaging demonstrated no evidence of metastasis. Cardiac catheterization confirmed two feeding arteries from right coronary artery into the mass. At surgery, after pericardiotomy, the mass was seen to be protruding from right atrial appendage and adhering to right side of pericardium (Figure 2 A). After establishment of cardiopulmonary bypass and cardiac arrest achieved, resection of the entire mass with right atrial appendage and partially free wall of right atrium was

performed (Figure 2 B). We could identify the feeding arteries and ligated them. Cryoablation was used for edge of right atrium resection and we reconstructed it with bovine pericardial patch. Intraoperative pathological evaluation of some suspicious lesions revealed finding of invasion into superior vena cava and annulus of tricuspid valve. However, because of difficulty of radical resection with more invasiveness, surgery was over. The histological findings were consistent with cardiac angiosarcoma (Figure 3) and immunological staining was positive for CD34 and CD31 (Figure 4 A and B). She was discharged on postoperative day 13. Afterward, although she received radiochemotherapy, she died of liver metastasis of cardiac angiosarcoma more than three years after surgical resection of primary cardiac angiosarcoma. Local recurrence of cardiac angiosarcoma was absent.

DISCUSSION

Primary cardiac sarcomas are neoplasms deriving from mesenchymal cells and limited to the heart at the time of diagnosis. The prevalence of primary cardiac sarcomas at autopsy was 0.0017%, with an incidence from 0.001% to 0.03%. Sarcomas account for approximately 10% to 20% of total primary cardiac tumors (4). Metastatic tumors to the heart are more frequently than primary cardiac tumors.

In adults, especially in males with a mean age of 50 years (5), primary cardiac angiosarcoma is the most common type of cardiac sarcoma. Approximately seventy-five percent of primary cardiac angiosarcoma occurs in the right side of the heart, in particular right atrium.

A diagnosis of primary cardiac angiosarcoma tends to be delayed because of the absence of clinical signs or symptoms for a long time (1). Symptoms are based on the cardiac location of the tumor, its size, the degree of myocardial involvement and the presence of metastasis (5). Given the propensity of primary angiosarcoma involving the right-sided heart, patients may present right-sided heart failure, superior vena cava obstruction, pericardial effusion (6).

Because of highly aggressive and invasive propensity of cardiac angiosarcoma, pericardial constriction and cardiac tamponade are frequent, and lung, bone, liver and spleen metastasis is reported. Unfortunately, 66% to 89% of patients have evidence of metastasis

Received for publication October 12, 2016 ; accepted December 13, 2016.

Address correspondence and reprint requests to Naoto Fukunaga, MD, Department of Cardiovascular surgery, Kobe City Medical Center General Hospital, 2-1-1 Minatojima-minamimachi, Chuo-ku, Kobe, 650-0047, Japan and Fax : +81-78-302-7537.

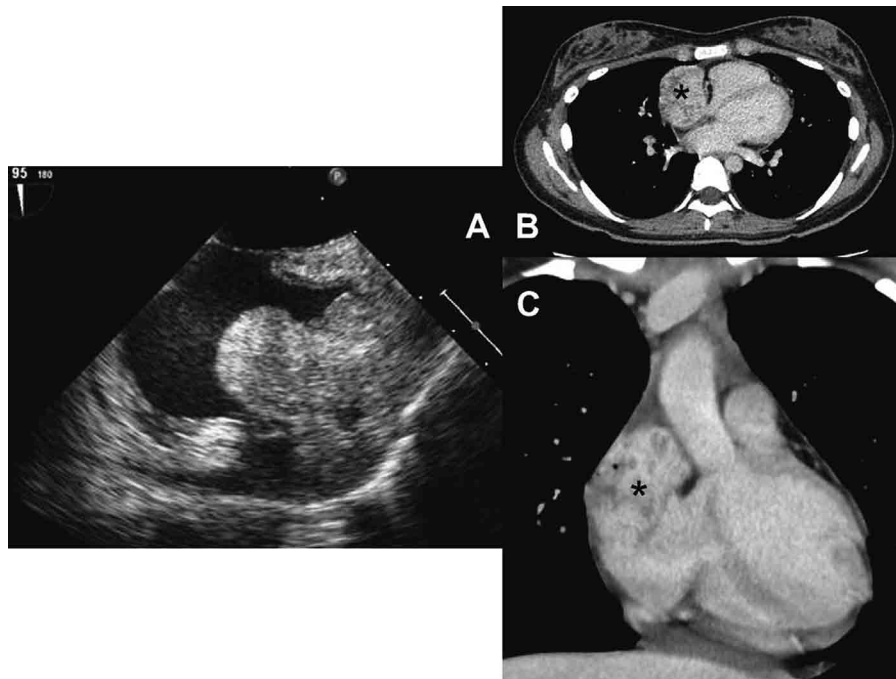


Figure 1 A. Transesophageal echocardiography shows that a mass is occupying the right atrial chamber and partially, invading into annulus of tricuspid valve.
 B. The axial view of enhanced computed tomography shows a mass is in the right atrial chamber (asterisk).
 C. The coronal view of enhanced computed tomography shows a mass is near the ascending aorta with no evidence of invasion (asterisk).

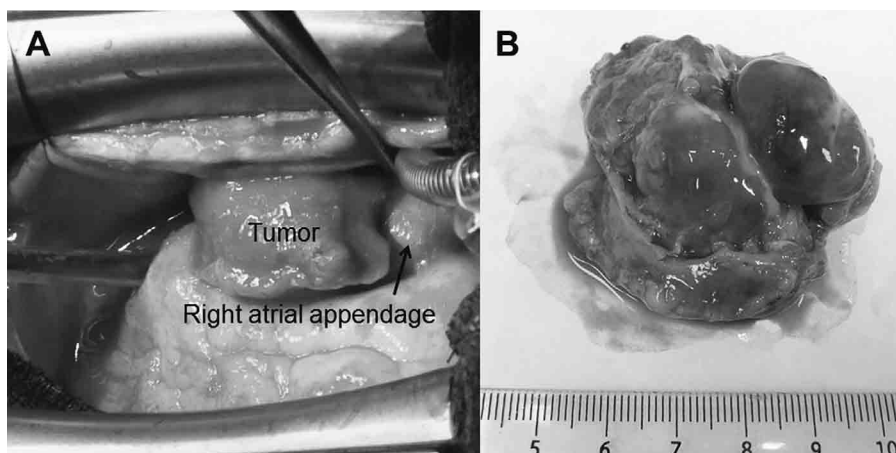


Figure 2 A. A mass is seen to be protruding from right atrial appendage and adhering to right side of pericardium.
 B. Resection of the entire mass with right atrial appendage and partially free wall of right atrium is performed.

at the time of diagnosis (6). In this situation, the prognosis is very poor with median survival of six months (2).

We applied immunostaining for CD31 and CD34 to diagnose angiosarcoma. CD31 and CD34 are useful markers for normal endothelial cells. Therefore, use of these antibodies leads to a definite diagnosis of angiosarcoma (7).

Surgery remains the mainstay of treatment in the absence of any extensive local spread and metastases. Liombart-Cussac *et al.* reported that only a surgical radical resection was found to be significantly associated with longer survival. They also found that adjuvant chemotherapy postoperatively failed to modify the natural course of this disease (8). Primary tumor resection can contribute

to improve median survival (3).

Patients with primary cardiac angiosarcoma had a shorter median survival of seven months and median disease-free progression interval of four months compared to other histological types (8). In present case, radiochemotherapy after surgical resection was effective because it inhibited aggressiveness of the angiosarcoma. We understand that radical treatment for primary angiosarcoma is difficult. Also, cardiac transplant for patients with these kinds of disease is not an option in Japan. Surgical resection and radiochemotherapy, which may be ineffective in some cases are main stay to deal with highly aggressive angiosarcoma. Surgical resection combined with radiochemotherapy was so effective to achieve relatively

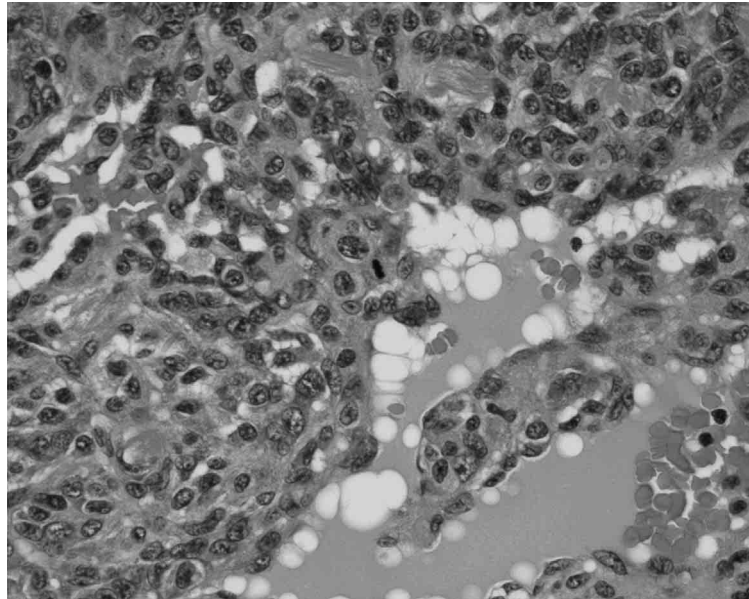


Figure 3 The histological findings are consistent with cardiac angiosarcoma. The tumor shows atypical spindle or endothelial cells which are lined around the vasoformative structures. Within those structures, erythrocytes can be recognized.

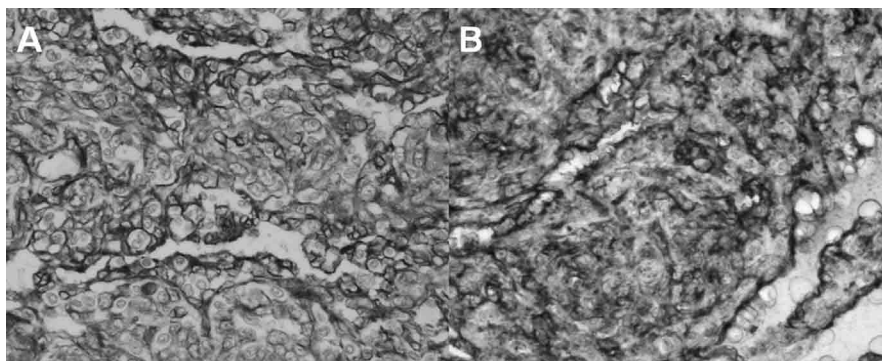


Figure 4 Immunohistochemical staining for CD31 (A) and CD34 (B) shows the focal endothelial differentiation. The positive for these immunohistochemical staining is brownish.

better survival.

CONFLICTS OF INTEREST DISCLOSURE

The authors have no financial or institutional interest in this study.

REFERENCES

- Bellitti R, Buonocore M, De Rosa N, Covino FE, Casale B, Sabte P : Primary cardiac angiosarcoma in a 25-year-old man : excision adjuvant chemotherapy, and multikinase inhibitor therapy. *Tex Heart Inst J* 40 : 186-188, 2013
- Prabu MPR, Thulkar S, Ray R, Bakhshi S : Primary cardiac angiosarcoma with good response to paclitaxel. *J Thorac Oncol* 10 : 1778-1779, 2011
- Look Hong NJ, Pandalai PK, Hornick JL, Shekar PS, Harmon DC, Chen YL, Bustrynski JE, Baldini EH, Raut CP : Cardiac angiosarcoma management and outcomes : 20-year single-institution experience. *Ann Surg Oncol* 19 : 2707-2715, 2012
- Orlandi A, Ferlosio A, Roselli M, Chiariello L, Spagnoli LG : Cardiac sarcomas : an update. *J Thorac Oncol* 5 : 1483-9, 2010
- Herrmann MA, Shankerman RA, Edwards WD, Shub C, Schaff HV : Primary cardiac angiosarcoma : a clinicopathologic study of six cases. *J Thorac Cardiovasc Surg* 103 : 655-64, 1992
- Bouma W, Lexis CPH, Willems T, Suurmeijer AJH, van der Horst ICC, Ebels T, Mariani MA : Successful surgical excision of primary right atrial angiosarcoma. *J Cardiothorac Surg* 6 : 47, 2011
- Furihata M, Takeuchi T, Iwata J, Sonobe H, Ohtsuki Y, Wakatsuki A, Morioka N, Sagara Y : Primary ovarian angiosarcoma : a case report and literature review. *Pathology International* 48 : 967-973, 1998
- Liombart-Cussac A, Pivot X, Contesso G, Rhor-Alvarado A, Delord JP, Spielmann M, Tursz T, Le Cesne A : Adjuvant chemotherapy for primary cardiac sarcomas : the IGR experience. *Br J Cancer* 78 : 1624-8, 1998