

# Successful Management of Primary Cardiac Lymphoma with Minimal Debulking Surgery Combined with Adjuvant Chemotherapy

Tae Hee Hong, MD, Dong Seop Jeong, MD, PhD

Department of Thoracic and Cardiovascular Surgery, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul, Korea

## ABSTRACT

Primary cardiac lymphoma (PCL) is a rare cardiac tumor with poor prognosis. Palliative chemotherapy is still considered to be the standard management tool for PCL. A case of a 58-year-old man with a large right-heart tumor is presented. Echocardiography showed no abnormal findings except mild tricuspid stenosis with dynamic obstruction. To prevent sudden right heart failure, we pursued on-pump beating resection. After resection of the tumor near the tricuspid valve and confirmation of normalized hemodynamics by intraoperative transesophageal echocardiography, we decided not to perform a further debulking procedure, such as resection or reconstruction of the atrial/ventricular wall. The postoperative course was uneventful and the patient tolerated six cycles of adjuvant chemotherapy well. Currently, the patient visits the outpatient clinic regularly without definite evidence of lymphoma involvement on follow-up imaging studies. In cases where a rare malignant cardiac tumor is suspected, surgical resection should be considered a diagnostic tool for tissue confirmation, a therapeutic tool for hemodynamic correction, and a preventive strategy for sudden cardiac death. Additionally, a minimal debulking procedure focusing on the area of hemodynamic disturbance appears to be sufficient in PCL cases.

## INTRODUCTION

Cardiac malignancy is generally considered to be an incurable and terminal disease. Surgical resection is known to have a limited role in these settings. However, several reports regarding primary cardiac lymphoma (PCL) have suggested that debulking surgery combined with adjuvant chemotherapy is a feasible treatment option [Fujita 2009; Gyoten 2015]. On the other hand, it is questionable whether aggressive debulking procedures (e.g. free wall resection and reconstruction) are required. Since the aim of resection is mainly focused on the restoration of hemodynamics, the extent of resection could be limited to the area of hemodynamic disturbance such as the tricuspid valve. Herein, we report a case

Received July 14, 2015; accepted September 16, 2015.

Correspondence: Dong Seop Jeong, MD, PhD, Department of Thoracic and Cardiovascular Surgery, Samsung Medical Center, Sungkyunkwan University School of Medicine, 81, Irwon-ro, Gangnam-gu, Seoul, Korea; +82-2-3410-1676; fax: 82-2-3410-0489 (e-mail: [cabg.jeong@samsung.com](mailto:cabg.jeong@samsung.com)).

with one-year follow up and discuss surgical issues in cardiac malignancies, especially PCL cases.

## CASE REPORT

A 58-year-old previously healthy male was found to have a huge mass in the right heart on transthoracic echocardiography. Despite the large size of the mass (6.7 × 5.5 cm), the patient was asymptomatic and the mass was found incidentally on a routine screening test. He was referred to our hospital for evaluation and treatment of a cardiac tumor. On preoperative studies, including chest X-ray, echocardiography, and computed tomography (CT) scan of the chest and abdomen, no metastatic lesions were found in other parts of the body. Cardiac magnetic resonance imaging (MRI) and transesophageal echocardiogram (TEE) revealed a huge mass involving

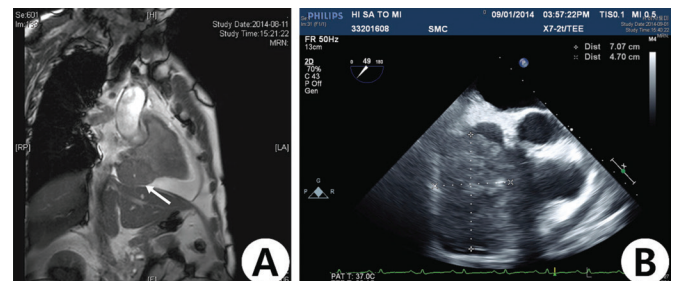


Figure 1. Preoperative imaging studies. A, Cardiac magnetic resonance imaging (MRI) revealing a huge mass involving the right atrium (RA) and right ventricle (RV) with heterogeneous enhancement and necrotic areas (white arrow). B, Transesophageal echocardiography revealing an irregularly shaped mass protruding from the RA.

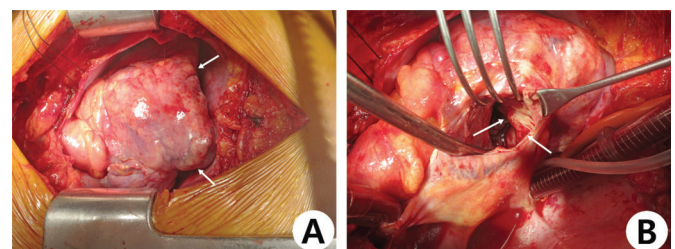


Figure 2. Operative findings. A, After median sternotomy, surface irregularity and diffuse infiltration of the RV tumor were noted (white arrow). B, After right atriotomy, a protruding mass toward the RA was confirmed (white arrow).

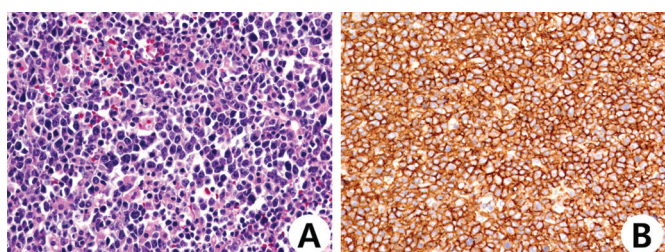


Figure 3. Histologic findings. A, Histologic examination of the biopsied specimen showing diffuse infiltration with a large number of neoplastic lymphoid cells, consistent with diffuse large-B cell lymphoma (Giemsa stain  $\times 60$ , Ki-67 stain  $\times 60$ ). B, Immunohistochemistry of the specimen revealed CD10+, CD19+ and CD20+, which strongly suggested diffuse large B-cell lymphoma.

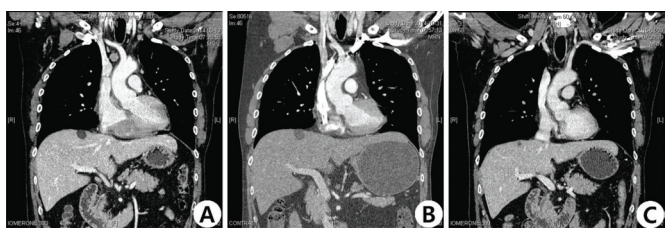


Figure 4. The postoperative computed tomography (CT) scans. A, The baseline CT image (at postoperative day 10) shows residual lymphoma involvement in the right atrium and heart base. B, After 3 cycles of chemotherapy, a follow-up coronal CT image (at postoperative month 2) reveals a noticeable decrease in the extent of lymphoma involvement. C, The latest follow-up CT image (at postoperative month 8) demonstrates no residual tumor.

the base of the right ventricle (RV) and right atrium (RA) without any hemodynamic abnormalities except for mild tricuspid stenosis with dynamic obstruction (Figure 1, A and B). Coronary angiography revealed normal results. Considering the high risk of tricuspid obstruction by the huge mass and thus the likely occurrence of sudden right heart failure, we decided to perform elective debulking surgery.

Through a median sternotomy and inverted T-shaped pericardiotomy, the basal and mid-level of the RV wall was noted to have an irregular surface due to diffuse tumor infiltration (Figure 2, A). After heparinization, the heart was cannulated in the ascending aorta, SVC, and IVC. Cardiopulmonary bypass (CPB) was initiated with a perfusion flow rate of 3.8 L/min. After snares around the SVC and the IVC were tightened, the RA was opened. The RA cavity was filled with an invasive tumor (Figure 2, B). Dynamic obstruction by the infiltrating tumor near the posterior leaflet of the tricuspid valve was noted. With the heart beating, the RA-protruding tumor near the tricuspid valve was removed. Intraoperative TEE revealed that normal tricuspid valve function was restored without residual stenosis. The tumor could not be removed completely because the lesions were diffusely distributed. In the same context, resection and reconstruction of the free wall of the RA and RV were not performed. The RA walls were closed and reinforced and the patient was weaned from CPB.

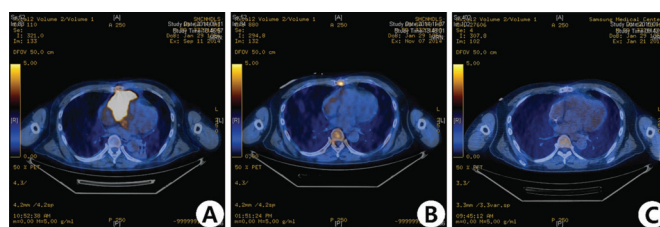


Figure 5. The postoperative positron emission tomography/computed tomography (PET/CT) scans. A, The baseline PET/CT image shows a large hypermetabolic lesion, suggestive of residual malignant tissue (SUVmax = 23.1). B, After 3 cycles of chemotherapy, a follow-up PET/CT image (at postoperative month 2) reveals markedly improved FDG uptake in the RV (SUVmax = 3.2). C, The latest follow-up PET/CT image (at postoperative month 4) demonstrates normalization of previous FDG uptake in the right heart.

The pathologic findings revealed the case to be diffuse large B-cell lymphoma (Figure 3, A and B). The patient had an uneventful postoperative course. On postoperative day 8, the patient was transferred to the oncology team of our hospital without any complications. He then underwent 6 cycles of rituximab, cyclophosphamide, adriamycin, vincristine, and prednisone (R-CHOP). Follow-up CT scans (Figure 4, A, B and C) and positron emission tomography/computed tomography (PET/CT) scans (Figure 5, A, B and C) were obtained in the immediate postoperative period, the end of the three and six cycles of chemotherapy to evaluate treatment response. Imaging studies suggested no evidence of residual tumor and normalized FDG uptake in the RV. Currently, the patient visits the outpatient clinic regularly without any complaints.

## DISCUSSION

PCL is extremely rare in immunocompetent patients and accounts for 1.3% of all cardiac tumors and 0.5% of all extranodal lymphomas [Sarjeant 2003]. The clinical manifestation may vary, and may include syncope, pericardial effusion, arrhythmia, and heart failure. According to a report, the median survival time of patients with PCL is 7 months [Nonami 2007]. Unfortunately, most PCL cases require urgent or emergent surgery because of life-threatening conditions [Park 2004; Choi 2008]. To our knowledge, this is the first report of asymptomatic PCL successfully managed by a combination of elective debulking operation and adjuvant chemotherapy. Also, this is the first case of a patient surviving PCL in the Korean population.

In our institution, debulking operation in the setting of cardiac malignancy is considered when (1) tissue confirmation is needed to select a treatment plan; (2) any hemodynamic disturbances (e.g., valvular stenosis/regurgitation) are documented in the preoperative evaluation; or (3) prevention of tumor embolism (e.g. pulmonary embolism) is required due to its fragile nature. However, it is unclear whether a debulking surgery provides survival benefit in cases of PCL. According to studies on PCL, only chemotherapy improved long-term survival [Ceresoli 1997; Petrich 2011]. However, we believe that surgical resection improves clinical outcomes

in high-risk patients. In patients with extremely large tumors close to valvular structures, sudden right heart failure is a worrisome complication and a debulking procedure may provide several benefits including prevention of sudden cardiac death.

Also, we hypothesize that the extent of resection could be limited to an area of hemodynamic disturbance. This minimal debulking strategy is distinct from previously reported cases. It can be helpful to minimize pump time and the incidence of possible complications, such as postoperative bleeding, conduction block, and pericarditis. Although we left most of the tumor inside the heart, the tumor was not seen on imaging after chemotherapy. While long-term follow-up is necessary, this case provides additional evidence that some cases of PCL can be managed with a debulking operation combined with R-CHOP chemotherapy.

On-pump beating resection was also a good strategy in this case because preoperative coronary angiography was normal and the tricuspid valve was not significantly involved. If the patient requires a concomitant reconstructive procedure, such as coronary artery bypass grafting and/or valve replacement, tumor resection with cardioplegic arrest might be a better option [Mo 2008]. We did not arrest cardiac circulation and the patient remained hemodynamically stable during the procedure.

In summary, surgical resection should be considered as a diagnostic procedure in cases of unknown cardiac malignancy. After thorough preoperative evaluation and careful patient selection, surgical resection of malignant cardiac tumors may improve clinical outcomes. Furthermore, a minimal debulking operation with a beating heart appears to be an acceptable strategy in cases of PCL.

## REFERENCES

- Ceresoli GL, Ferreri AJ, Bucci E, Ripa C, Ponzoni M, Villa E. 1997. Primary cardiac lymphoma in immunocompetent patients: diagnostic and therapeutic management. *Cancer* 80:1497-506.
- Choi WS, Han IY, Jun HJ, Lee YH, Hwang YH, Cho KH. 2008. Primary malignant cardiac lymphoma in right atrium: a case report. *Korean J Thorac Cardiovasc Surg* 41:369-72.
- Fujita Y, Ikebuchi M, Tarui S, Irie H. 2009. Successful combined treatment of primary cardiac malignant lymphoma with urgent cardiac operation and chemotherapy. *Circ J* 73:967-9.
- Gyoten T, Doi T, Nagura S, et al. 2015. Primary cardiac malignant lymphoma: survival for 13 years after surgical resection and adjuvant chemotherapy. *Ann Thorac Surg* 99:1060-2.
- Mo A, Lin H, Wen Z, Lu W, Long X, Zhou Y. 2008. Efficacy and safety of on-pump beating heart surgery. *Ann Thorac Surg* 86:1914-8.
- Nonami A, Takenaka K, Kamezaki K, et al. 2007. Successful treatment of primary cardiac lymphoma by rituximab-CHOP and high-dose chemotherapy with autologous peripheral blood stem cell transplantation. *Int J Hematol* 85:264-6.
- Park KS, Ahn WS, Lee S, Kwon OC, Ko MS, Jheon SH. 2004. Primary non-hodgkin's lymphoma in right ventricle with right atrial invasion: report of 1 case. *Korean J Thorac Cardiovasc Surg* 37:376-81.
- Petrich A, Cho SI, Billett H. 2011. Primary cardiac lymphoma: an analysis of presentation, treatment, and outcome patterns. *Cancer* 117:581-9.
- Sarjeant JM, Butany J, Cusimano RJ. 2003. Cancer of the heart: epidemiology and management of primary tumors and metastases. *Am J Cardiovasc Drugs* 3:407-21.