Successful Treatment of Primary Cardiac Lymphoma With Atrioventricular Nodal Block

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SUMMARY

A 69-year-old female suffering from third-degree atrioventricular block with syncope underwent permanent pacemaker implantation. However, she developed shortness of breath 2 months after the implantation. Blood tests revealed elevated levels of LDH, CRP, BNP, and SIL-2R. Transthoracic echocardiography showed thickened left and right atrial walls with mild pericardial effusion. A diagnosis was made based on a CT scan and histology. Although most primary cardiac malignant lymphomas are associated with a poor prognosis, the patient was treated successfully with chemotherapy. (Int Heart J 2005; 46: 927-931)

Key words: Tumor, Cardiac lymphoma, Heart, Chemotherapy

PRIMARY cardiac malignant lymphomas are extremely rare,1 and almost all are aggressive B-cell lymphomas.2 They are occasionally detected together with cardiac tamponade or a conduction disturbance, but in the majority of cases are diagnosed before death.3,4 We report a case of primary cardiac lymphoma in a patient who was successfully treated with chemotherapy.

CASE REPORT

A 69-year-old female suffering from third-degree atrioventricular block with syncope underwent permanent pacemaker implantation. However, she developed shortness of breath 2 months after the implantation. She underwent a
Figure 1. Transesophageal echocardiography demonstrating a mass encircling the left atrium (left panel; marked with arrows) and the thickened interatrial septum (right panel; marked with arrows) with mild pericardial effusion.

Figure 2. Chest computed tomography on admission demonstrating abnormal swelling of para-aortic lymph node (left panel) and thickened interatrial and interventricular septum (right panel).

checkup as an out-patient and we found expansion of CTR and little pleural effusion on chest X-rays. Blood tests revealed elevated levels of LDH, 407 IU/L (120-240 IU/L), CRP, 1.2 mg/dL, and BNP, 149 pg/dL. Transthoracic echocardiography showed thickened left and right atrial walls with mild pericardial effusion (Figure 1).

A CT scan of the thorax was performed (Figure 2), which confirmed a mass encasing the heart and invading the right and left atrium and intraventricular septum. In addition, a mediastinal lymph node was swollen, which could not have been found on a CT taken 2 months earlier. No other lymph node swelling or marrow permeation was detected. SIL-2R (1169 U/mL) was also elevated. Aspiration
of the pleural effusion was performed and histopathological examination revealed a large cell type lymphoma (Figure 3) with LCA(+), L26(+), and CD79a(+) immunohistochemically, which strongly suggested B-cell non-Hodgkin's lymphoma.

She was treated with multiagent chemotherapy. Initially she was given THPCOP therapy (vincristine sulfate, cyclophosphamide, pirarubicin hydrochloride and prednisolone), followed by CHOP therapy (vincristine sulfate, cyclophosphamide, doxorubicin hydrochloride and prednisolone). Her condition improved. Imaging over the following 3 weeks revealed resolution of the pericardial effu-

![Figure 3](image_url)

**Figure 3.** Microscopic examination of Giemsa-stained specimen showing large cells with distinct nucleoli (A and B). Immunohistochemical staining was positive for LCA, L26 and CD79a, which was a B-cell marker (C, D and E).

![Figure 4](image_url)

**Figure 4.** Chest computed tomography after 5 series of chemotherapy demonstrated no swelling of lymph node (left panel) and regression of the interatrial and interventricular septum (right panel).
sion and a reduction of the tumor mass in the atrium and the intraventricular septum. Her cardiac rhythm also recovered and blood tests revealed normalized LDH (188 IU/L) and SIL-2R (432 U/mL). A CT scan after 5 series of chemotherapy is shown in Figure 4.

**DISCUSSION**

Since primary cardiac lymphomas are extremely rare and have no specific symptoms, a diagnosis is difficult ante-mortem and the prognosis is often poor. A diagnosis of cardiac lymphoma is difficult because of limitations in the imaging resolution of available techniques, which is usually conducted with the help of a CT scan as well as echocardiography which could confirm the presence of an occupied mass. Cytology is diagnostic in only two-thirds of cases. This case was diagnosed by pleural effusion and treatment could be started. If cytology is not available, a diagnosis of primary cardiac lymphoma is usually determined by cardiac tissue biopsy. Chim, et al reported that only 9 of 21 patients could be diagnosed ante-mortem by cytological examination of pericardial effusion, surgical biopsy, or pericardial and myocardial biopsy during pericardiocentesis. Others have reported a diagnosis was possible by transvenous biopsy under transesophageal echocardiographic guidance.

Recently, a monoclonal CD20 antibody (Rituximab) was found to induce regression in B-cell non-Hodgkin’s lymphomas in patients who were CD20 positive. This case was negative for CD20 and she had heart failure. We started her initial treatment with THP-COP therapy.

In spite of no signs of conduction disturbance, sudden death occurred in many cases. This suggests that the lymphoma infiltrated the cardiac conduction system silently and caused the fatal arrhythmia suddenly. Our case also had complete atrioventricular block with syncope and required pacemaker implantation, which helped her avoid sudden death. Otsuji, et al reported that complete atrioventricular block disappeared after successful chemotherapy, which was the same course of our case.

**REFERENCES**


