Successful Combined Treatment of Primary Cardiac Malignant Lymphoma With Urgent Cardiac Operation and Chemotherapy

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A 56-year-old man, suffering from rapidly worsening general fatigue, dyspnea on exercise, and epigastralgia, was referred for evaluation and treatment of a cardiac tumor. Transthoracic echocardiography showed a 6-cm large mass occupying both the right ventricle and atrium. Gallium scintigraphy showed high uptake in the tumor site. Lymphoma was highly suggested. Urgent operation was performed and as much tumor and thrombus were removed as possible. The postoperative course was good, with cessation of right heart failure. Pathological examination suggested malignant lymphoma, diffuse large B-cell type. The patient was treated with rituximab, cyclophosphamide, Adriamycin, vincristine, and prednisone (CHOP-R) postoperatively and has survived for 2 years without signs of recurrence. (Circ J 2009; 73: 967–969)

Key Words: Chemotherapy; Primary cardiac lymphoma; Surgical resection

P rimary cardiac lymphomas are a rare malignancy and most are B-cell type. The prognosis is poor, because of the advanced stage of myocardial involvement at the time of presentation. Surgical resection is rarely performed.

We report a case of primary cardiac lymphoma in a patient who was successfully treated with surgical resection and chemotherapy.

Case Report

A 56-year-old man, suffering from rapidly worsening general fatigue, dyspnea on exercise, and epigastralgia for 1 month, visited a regional hospital. Ultrasound sonography and abdominal computed tomography (CT) scan showed pericardial effusion and ascites. Chest CT scan revealed a huge tumor in the right ventricle (RV, Figure 1). He was referred in June 2005.

He developed tachycardia of 100 beats/min, the jugular vein was dilated, and the general fatigue and appetite loss had been worsening daily since admission. Transthoracic echocardiography showed right-sided heart failure because of a 6-cm large mass occupying almost all the cavities of both the RV and right atrium (RA). Coronary angiography on day 4 after admission showed no stenotic lesion. He had a family history of gastric cancer in his father, brother, and sister.

Gallium scintigram showed high uptake in the tumor site (Figure 2). Lymphoma was highly suspected. General fatigue became marked on day 7 after admission, so he was transferred to the intensive care unit where a central venous line was inserted, the pressure of which was 22 mmHg. Although his systolic blood pressure was 130–140 mmHg, urine volume decreased. Because his right-heart failure had deteriorated rapidly after admission, we performed lifesaving urgent operation on day 8.

A median sternotomy was made and the heart was exposed. There was a serous pericardial effusion of 150 mL. The anterior and inferior walls of the RV were very stiff and immobile. Tumor was exposed to the epicardium (Figure 3) and involved the distal right coronary artery (RCA). The main trunk of the RCA was not involved, so coronary arterial bypass grafting was thought not to be needed. The lateral wall of the RA was also rigid. Echocardiography...
showed that the tumor had not invaded either the superior vena cava (SVC) or the inferior vena cava (IVC). After heparinization, the heart was cannulated in the ascending aorta, the SVC and the IVC. Cardiopulmonary bypass (CPB) was commenced to cool the body to 32°C. The perfusion flow rate was 2.4L·min⁻¹·m⁻². The ascending aorta was cross-clamped and cold blood cardioplegia was infused into the aortic root. After snares around the SVC and the IVC had been tightened, the RA was opened in an oblique fashion; 50% of the cavity was filled with thrombi and tumor. The right atrial appendage was filled with old blood clot. The tricuspid valve was not visible.

The intracardiac tumor was very soft and fragile like jelly. It was removed to expose the tricuspid valve and then a small incision was made in the outflow of the RV along the interventricular septum. Tumor was removed as much...
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as possible with pickups. Finally, a 27-mm sizer was passed through the tricuspid valve, but part of the tumor could not be removed. The tricuspid valve was well visible from both sides. Both the RV and the RA walls were closed and the patient was rewarmed. The ascending aorta was unclamped. CPB was terminated easily. Transesophageal echocardiography revealed that the right heart blood flow tract was preserved well and that the motion of the tricuspid valve was good without regurgitation. All wounds were closed in the usual manner.

The postoperative course was good. His right-sided heart failure and ascites disappeared. CT scan on postoperative day 6 showed that the tumor was decreased in size (Figure 4). Pathological examination showed diffuse proliferation of atypical large lymphoid cells with CD10+ and CD20+ immunohistochemistry, which strongly suggested malignant lymphoma of diffuse large B-cell type. The patient was then referred to another hospital on postoperative day 11 for chemotherapy. He was treated with rituximab, cyclophosphamide, Adriamycin, vincristine, and prednisone (CHOP-R).

After clinical remission, another additional course was administered. He was discharged after a 1-month admission and additional chemotherapy was performed in the outpatient clinic. The patient has been alive for 2 years without signs of recurrence.

Discussion

Because primary cardiac lymphomas are a rare malignancy, the diagnosis is difficult and the prognosis is often poor. The definition has been debated, but currently includes non-Hodgkin’s lymphoma involving only the heart or with the bulk of the tumor located in the heart. Most are B-cell lymphomas, and their treatment has not been established. Several articles report successful treatment of primary cardiac lymphoma with chemotherapy, while others report successful treatment by surgical resection and chemotherapy. However, Ceresoli et al reported that there is no evidence of survival improvement with surgery.

In the present case, right heart failure deteriorated rapidly after referral. Almost all the cavities of the RV and RA were occluded with the tumor, which impaired RV function. Lifesaving urgent operation was needed. Although the tumor was not resected completely, the mass was reduced enough to improve RV function. Even in such palliative cases, chemotherapy after operation can be successful.

The patient has been alive for 2 years without signs of recurrence. We consider that volume reduction of the tumor improved the efficacy of chemotherapy.

Chemotherapy alone might have been effective in this case, but we considered that we had no time before the tumor occluded the RV. Therefore we have no idea which is better, chemotherapy alone or a combination of chemotherapy and surgery.

In conclusion, we report a case of primary cardiac lymphoma in a patient who was successfully treated with surgical resection and chemotherapy. Because malignant lymphoma may be curable, we should consider the possibility when a cardiac mass is found.

References