

# Treatment of a primary cardiac lymphoma with orthotopic heart transplantation

Ortotopowe przeszczepienie serca w leczeniu pierwotnego chłoniaka serca

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## Abstract

We here describe the case of a patient diagnosed with primary lymphoma who underwent orthotopic heart transplantation. Postsurgically, the patient was treated with immunosuppressants and chemotherapy. Three months later, lesions were found on the right side of the neck and the inferior vena cava; 12 months later, the lesion of the neck became smaller and the lesion of the inferior vena cava was not detected by positron emission tomography/computed tomography. The problems we encountered with this rare disease are discussed and compared to previous studies.

**Key words:** primary cardiac lymphoma, orthotopic heart transplantation, R-CHOP

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## INTRODUCTION

Primary cardiac lymphoma (PCL) is a rapidly progressing and fatal primary cardiac malignant tumour [1]. Because the early diagnosis is challenging (due to the varied clinical manifestations), the prognosis is poor [1]. This report describes an orthotopic heart transplantation in a patient with PCL.

## CASE REPORT

A 53-year-old male Chinese patient with normal immune function (HIV negative) and no history of heart disease was admitted with dyspnoea of unknown cause. There was no jugular vein engorgement, breath sound was clear in both lungs, and no rales were auscultated. The heart sounds were quiet, but there were no murmurs or sounds indicative of pericardial friction. On echocardiography, a space-occupying lesion in the parenchyma of the heart blocking the tricuspid orifice was noted in addition to a small volume of pericardial effusion (Fig. 1A). A cardiac computed tomography (CT)-digital subtraction angiography revealed a large space-occupying lesion within the right atrial wall (the maximum cross-sectional area was 50 mm × 48 mm). The lesion encircled the right coronary artery and protruded into the right atrium. Fluid-density signals could be observed inside the pericardial cavity

(Fig. 2B). A CT showed no abnormal space-occupying lesions and no enlarged lymph nodes outside the heart. The patient's cardiac function was deteriorating rapidly, so emergency orthotopic heart transplantation was performed. The tumour (8 cm × 7 cm × 5 cm) protruded into the right atrium and ventricle and blocked the tricuspid orifice, but the superior and inferior vena cava were not invaded (Fig. 2A). The right coronary artery was encircled by tumour tissue.

Histology of the right ventricular myocardium determined that the tumour was a diffuse large B-cell lymphoma (Figs. 2B, C). The results of immunohistochemistry test were as follows: CD20<sup>+</sup> (FIG-2D), cyclin D1<sup>-</sup> (FIG-2E), PAX-5<sup>+</sup>, Mum-1<sup>+</sup>, CD3<sup>-</sup>, CD21<sup>-</sup>, CD10<sup>-</sup>, CD5<sup>-</sup>, Bcl-6<sup>-</sup>, and Ki67<sup>+</sup> (80%). Anti-immune rejection therapy was prescribed, and no acute rejection occurred. R-CHOP chemotherapy (i.e. rituximab, cyclophosphamide, doxorubicin hydrochloride, vincristine, prednisone) was initiated two months postsurgically.

Three months later, positron emission tomography (PET)-CT suggested abnormal hypermetabolism in the right side of the neck and local abnormal hypermetabolism in the inferior vena cava; 12 months later, the lesion of the neck became smaller and the lesion of the inferior vena cava was not detected by PET-CT (Fig. 3).

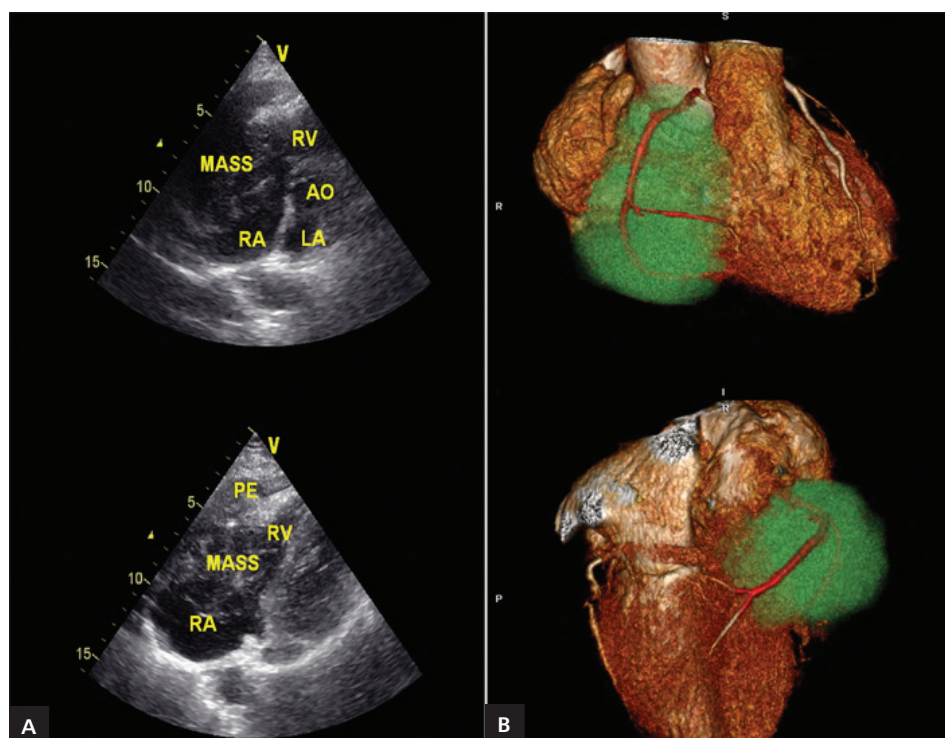
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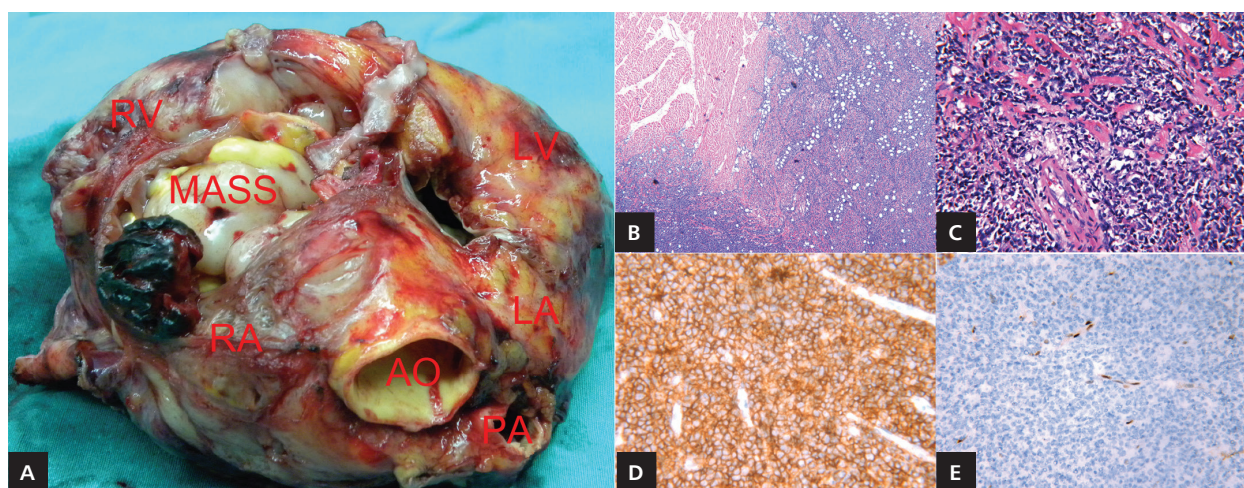
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**Figure 1.** Echocardiography and cardiac computed tomography scan from a 53-year-old man with primary cardiac diffuse large B-cell lymphoma showing; **A.** A space-occupying lesion in the parenchyma of the heart blocked the tricuspid orifice; **B.** The lesion encircled the right coronary artery and protruded into the right atrium (RA); Ao — aorta; LA — left atrium; RV — right ventricle; PE — pericardial effusion

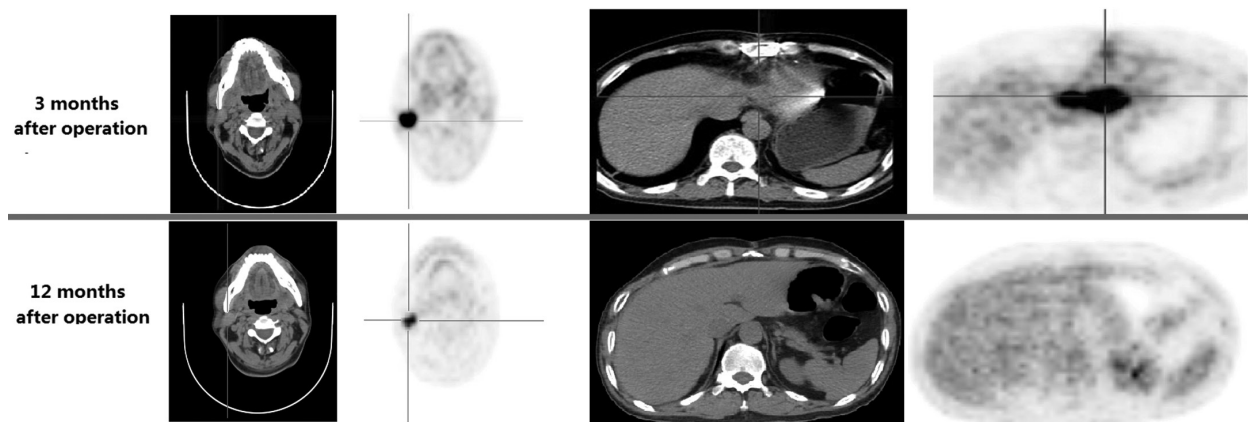


**Figure 2.** Pathological examination of the heart of a 53-year-old man with primary cardiac diffuse large B-cell lymphoma showing; **A.** The tumour (measuring 8 cm × 7 cm × 5 cm) protruded into the right atrium and ventricle and blocked the tricuspid orifice; **B, C.** Lymphoid cells (haematoxylin and eosin staining); **D.** CD20-positive tumour cells (immunohistochemical staining); **E.** Cyclin D1-negative tumour cells (immunohistochemical staining); Ao — aorta; LA — left atrium; RA — right atrium; RV — right ventricle; PA — pulmonary artery

### DISCUSSION

PCL is a non-Hodgkin's lymphoma occurring outside the lymph system, involving only the pericardium and the heart. The right atrium and right ventricle are most commonly involved

[2, 3]. Patients with PCL exhibit a paucity of specific clinical manifestations, ergo the disease is either easily overlooked or misdiagnosed [4]. Clinical manifestations vary depending on the location of tumour invasion. Some 'typical' signs include



**Figure 3.** Positron emission tomography-computed tomography scans showing: three months later, abnormal hypermetabolism in the right side of the neck and local abnormal hypermetabolism in the inferior vena cava (up); 12 months later, the lesion of the neck became smaller and the lesion of the inferior vena cava was not detected by positron emission tomography/computed tomography (down)

pericardial effusion, heart failure, and atrioventricular block [5, 6]. Echocardiography, CT, and magnetic resonance imaging can identify tumour size and range of tumour invasion as well as invasion within the pericardium, mediastinum, hilum of the lung, and diaphragm, and adjacent lymph nodes [1].

Surgery and chemotherapy are usually attempted, but complete resection of the lesion is difficult [7]. In most cases, palliative surgery improves the symptoms caused by pericardial effusion [8]. Orthotopic heart transplantation is not a routine treatment of heart tumours.

In the case described herein, the tumour significantly violated the right ventricle and atrium, blocked the tricuspid orifice, and encircled the right coronary artery. Because the disease progressed rapidly, palliative surgery could not prevent the upcoming cardiac adverse events. No metastatic lesion was identified outside the heart and a suitable heart donor was available; therefore an orthotopic heart transplantation was performed, resulting in complete removal of the primary lesion. Unfortunately, three months postsurgically, abnormal hypermetabolism in zone I on the right side of neck and local abnormal hypermetabolism in the soft tissue of the inferior vena cava was suspected. These two lesions either could have been from preoperative tiny metastatic lesions which were not found on the original CT scan, or could have been from residual tumour cells that successfully achieved immune escape.

Alternatively (or in addition), the anti-immune rejection drugs could have contributed to tumour growth. Although the primary heart lymphoma in this case was a diffuse large B-cell lymphoma which is sensitive to R-CHOP chemotherapy, as the lesion of the neck became smaller and the lesion of the inferior vena cava was not detected by PET/CT after eight treatment of R-CHOP, long-term anti-immune rejection drugs are necessary [9, 10].

## CONCLUSIONS

Although there is no experience of treating primary heart lymphoma with heart transplantation, it is undeniable that completely removing the tumour can increase survival. Further research into tumour recurrence and the use of chemotherapeutics is needed.

**Conflict of interest:** none declared

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