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Case Report

Primary cardiac lymphoma: a case report and review of literature

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Abstract: Background: Primary cardiac lymphoma (PCL) is a rare cardiac neoplasm. The most common type is diffuse large B-cell lymphoma, which mainly involves the right atrium and right ventricle. Its clinical manifestations are not specific. It is usually found late and the prognosis is poor. PCL is more common in immunodeficient patients. In view of the rarity of PCL, we have made a case report and a brief literature review. Case Presentations: We documented an elderly male patient with Burkitt's lymphoma diagnosed by echocardiography and percutaneous biopsy because of chest tightness. Chemotherapy was given to the patient, and the patient's condition was alleviated without recurrence. Conclusions: Primary cardiac lymphoma is a highly malignant tumor. Early detection, diagnosis and treatment are crucial to the prognosis of PCL. At present, the most effective treatment is chemotherapy. There are more chemotherapy schemes and treatment methods worth studying.

Keywords: Cardiac neoplasms, lymphoma, non-Hodgkin's lymphoma, chemotherapy

Background

Primary cardiac lymphoma (PCL) is a rare extranodal lymphoma with a high degree of malignancy, primary to the heart and/or pericardium, mainly characterized by heart-related symptoms. Diffuse large B-cell lymphoma is the most common type in the heart [1, 2]. Burkitt lymphoma (BL) is a rare highly invasive B-cell non-Hodgkin's lymphoma (NHL), that has characteristic MYC oncogene rearrangement. There are three forms of BL: sporadic, endemic, and immunodeficiency-related. BL is currently curable, but treatment options for recurrent and refractory diseases are still limited [3]. This study reports a case of primary cardiac Burkitt lymphoma, followed by a brief literature review.

Case presentations

A 68-year-old man was admitted to the hospital with chest tightness for one week. After activity, he experienced chest area bloating, accompanied by wheezing, nausea, and occasional mild chest pain. His medical history included hypertension, lacunar cerebral infarction, and cholecystectomy. Cardiac examination revealed atrial fibrillation, heart rate 74 bpm, and no obvious pathological murmur.

Echocardiography showed dilatation of both atria, abnormal echogenic light mass (46 mm × 23 mm) in the right atrium, and a small amount of pericardial effusion (**Figure 1**). Electrocardiogram (ECG) showed atrial fibrillation and partial lead T wave abnormalities (**Figure 2**). Laboratory examination revealed NT-ProBNP 1360.0 pg/mL, hemoglobin 116.9 g/L, and lactate dehydrogenase (LDH) 342 U/L. A computed tomography (CT) scan of the chest revealed a mass in the right atrium and right diaphragm angle with a diameter of about 66 mm × 74 mm, accompanied by a filling defect in the vena cava (**Figure 3A**). Cardiac magnetic resonance imaging (MRI) indicated right atrial and right paracardial masses with a diameter of 66 mm × 71 mm and 57 mm × 65 mm, accompanied by thickening of the right ventricular wall, pericardial effusion, and right pleural effusion (**Figure 3B**). The pleural effusion examination showed exudate, of which lymphocytes accounted for 97%, suggesting that lymphoma cells might be present. CT-guided percutaneous

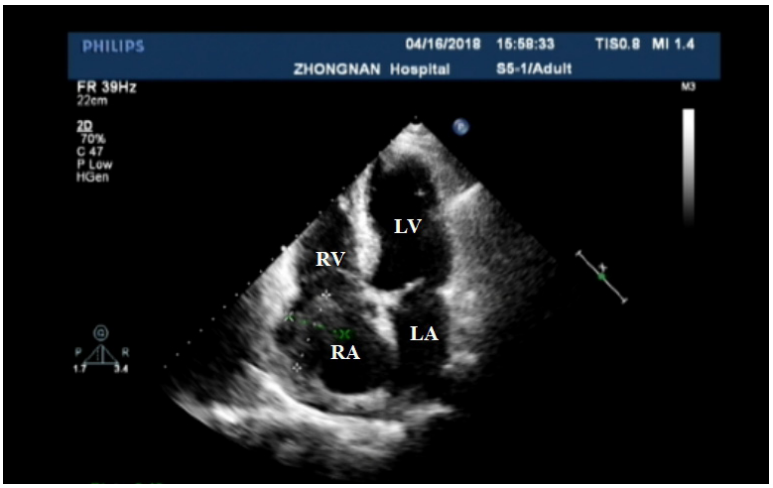

Figure 1. Echocardiography shows a 46 mm × 23 mm right atrial mass (arrow) (crosshairs represent dimension measurements). RA: right atrium; LA: left atrium; RV: right ventricle; LV: left ventricle.

biopsy of mediastinal masses suggests non-Hodgkin's lymphoma (high-grade B-cell lymphoma) (**Figure 4**). Immunohistochemical analysis of neoplastic cells showed that BCL-6, CD10, CD20, EBER, and PAX5 were all positive, ki-67 positive rate was 100%, BCL-2 was suspicious for positive; and CD21, CD23, CD3, CD30, and CD5 were negative. Fluorescence in situ hybridization (FISH) showed C-MYC gene rearrangement without recombination of BCL-2 and BCL-6.

The final diagnosis was Burkitt lymphoma stage IV and IPI 4 score. Pre-chemotherapy w dexamethasone and cyclophosphamide for 3 days, followed by chemotherapy was rituximab, dexamethasone, etoposide, doxorubicin, vinpodesine, cyclophosphamide (R-EPOCH) regimen. After two cycles of chemotherapy, echocardiography showed that the right atrial mass disappeared (**Figure 5**), followed by four cycles of R-EPOCH chemotherapy. One year later, PET-CT showed that the right pericardium was thicker and no atrial mass was found (**Figure 6**). To date, there was no recurrence of lymphoma in this patient.

Discussion

Primary cardiac tumors are rare. About 90% of primary cardiac tumors are benign, including myxomas, fibromas, lipomas, hemangiomas, and rhabdomyomas. Primary cardiac lymphoma (PCL) is more rare, accounting for about 1% to 2% of the incidence of primary cardiac tumors. PCL is mostly diffuse B-cell non-Hodgkin's lymphoma [1, 4]. PCL is mainly an extranodal lymphoma that invades the cardiac cavity and/or pericardium and is characterized by cardiac symptoms, which is more common in immunocompromised populations such as those with AIDS. It often involves the right atrium and right ventricle, rarely the left atrium and left ventricle, and can also involve the superior vena cava, inferior vena cava, and pericardium [2]. Symptoms of PCL are non-specific and can be manifested as chest tightness, chest pain, dyspnea, arrhythmia, syncope, vena cava superior syndrome, heart failure and other symptoms. The electrocardiogram can show atrial fibrillation, atrioventricular block, right bundle branch block, T wave inversion, and life-threatening ventricular tachycardia [4, 5].

For patients suspected of PCL, ECG, transthoracic echocardiography (TTE), transesophageal echocardiography (TEE), cardiac CT, MRI and PET-CT should be completed. ECG and laboratory examination are not specific for PCL diagnosis. Echocardiography is a simple method to detect large cardiac tumors, pericardial effusion and pericardial thickening. TEE is more sensitive than TTE in the diagnosis of primary cardiac tumors. Cardiac MRI can clearly distinguish the relationship between tumors and surrounding tissues. PET-CT has a good assessment of the systemic invasion and metabolism of the tumor, and puncture examination of pleural effusion and pericardial effusion can lead to the diagnosis of lymphoma cells. Of course, the final diagnosis still depends on biopsy by puncture or surgical excision [5-7].

For the treatment of PCL, surgery and radiotherapy have little effect. Severe compression or obstruction can be treated surgically, but surgery can not completely remove PCL. Minimally invasive surgery can be used for early diagnosis. Systemic chemotherapy is the main treatment and should be started as soon as

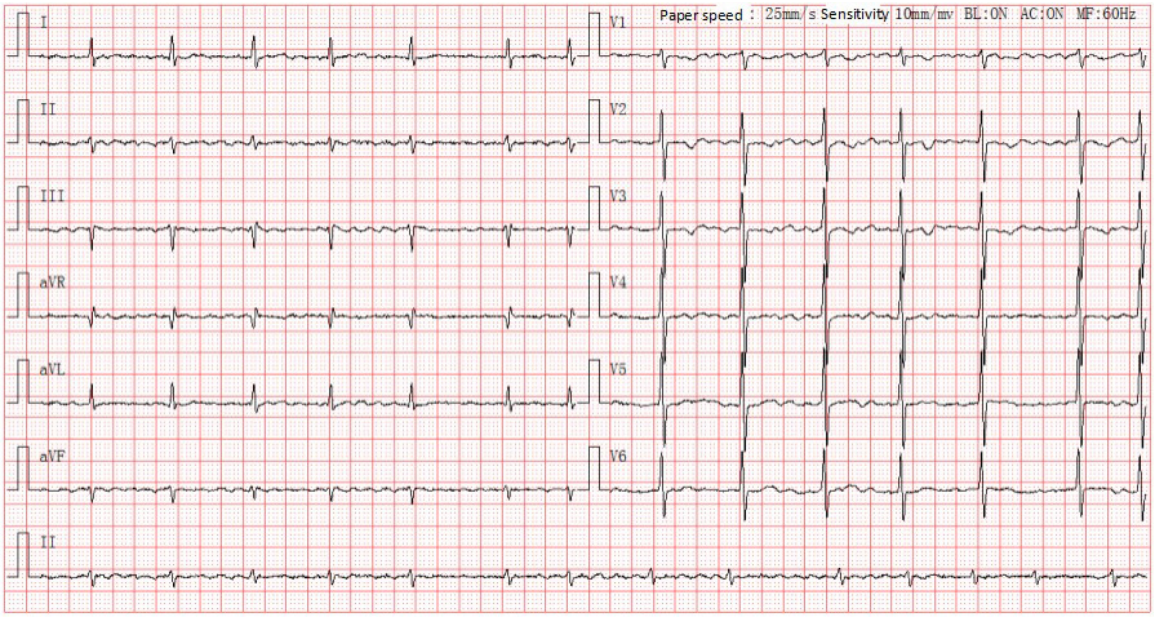

Figure 2. ECG shows atrial fibrillation and partial lead T-wave abnormalities.

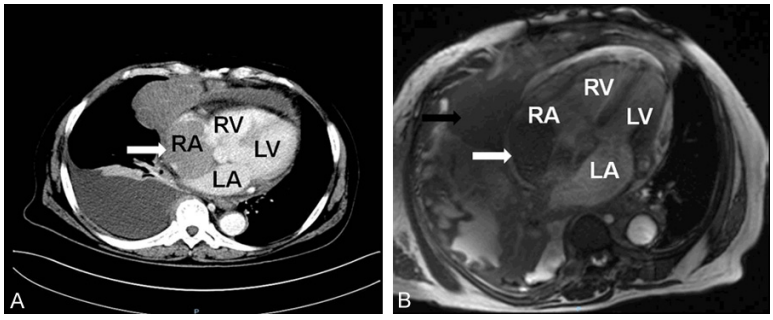

Figure 3. A. CT scan of the chest shows a 66 mm × 74 mm right atrial mass (arrow). B. Cardiac MRI shows a 66 mm × 71 mm right atrial mass (white arrow), and a right paracardial mass of 57 mm × 65 mm (black arrow), accompanied by pericardial effusion and right pleural effusion. RA: right atrium; LA: left atrium; RV: right ventricle; LV: left ventricle.

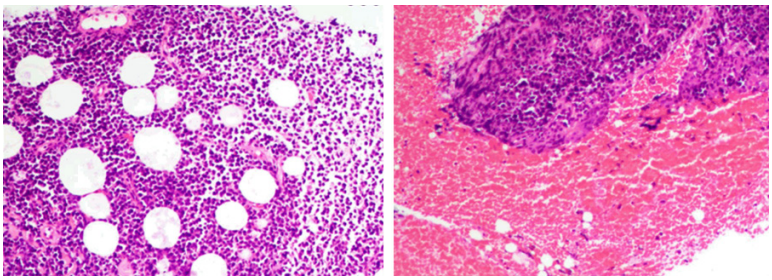

Figure 4. Biopsy and pathologic examination of mediastinal mass shows non-Hodgkin's lymphoma (high-grade B-cell lymphoma).

possible, which can significantly shrink the tumor and improve survival. The main chemotherapy regimens are CHOP (cyclophosphamide, doxorubicin, vincristine and prednisone) and R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine and prednisone) [5, 8, 9]. R-EPOCH regimen can be used in patients with recurrent or refractory diffuse large B-cell lymphoma, such as Burkitt's lymphoma. High-dose chemotherapy combined with autologous stem cell transplantation (ASCT) can also be used as a treatment for invasive lymphoma [3, 10].

Conclusions

Primary cardiac lymphoma is a very rare disease with poor prognosis and non-specific symptoms. Early pathologic diagnosis and early treatment are of great importance, closely related to the prognosis and survival rate of patients. Currently, the most effective treatment is chemotherapy. However, there are still many side effects and complications of PCL treatment,

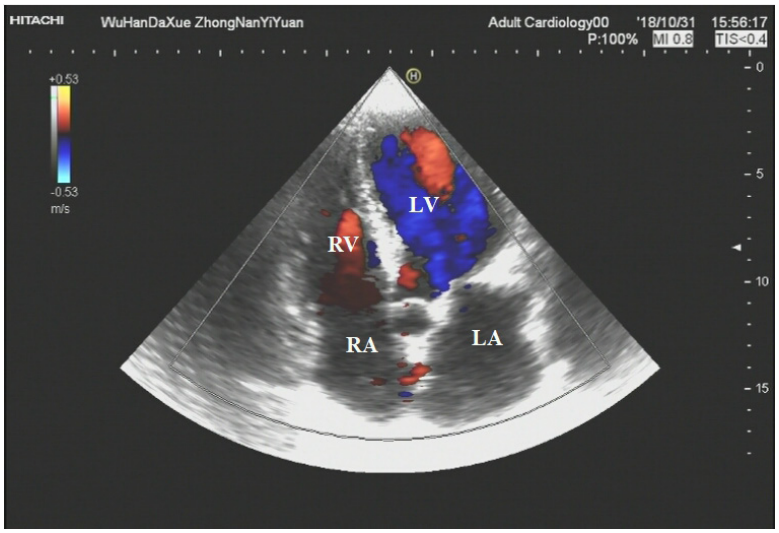

Figure 5. Echocardiography showed that the right atrial mass disappeared. LA: left atrium; RA: right atrium; RV: right ventricle; LV: left ventricle.

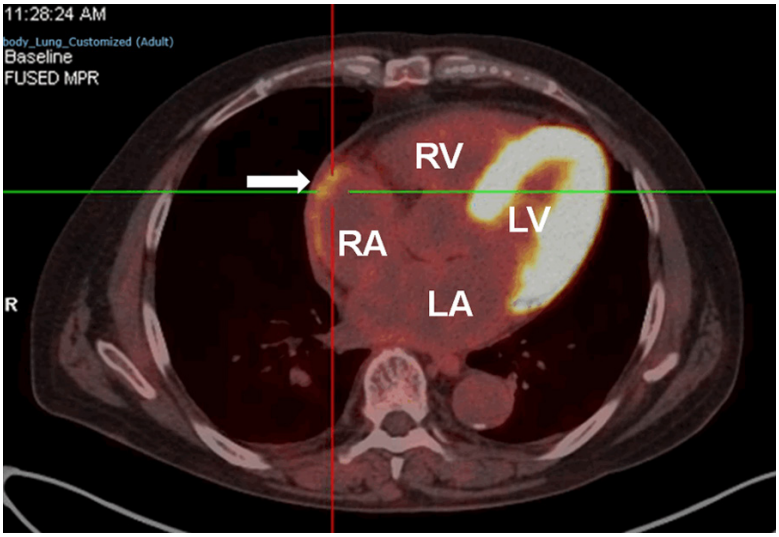

Figure 6. PET-CT showed that the right pericardium was thicker (white arrow) and no atrial mass was found. LA: left atrium; RA: right atrium; RV: right ventricle; LV: left ventricle.

and the treatment of relapse and refractory PCL is still limited. There are still great prospects for the research on the treatment of PCL and the reduction of complications and side effects.

Disclosure of conflict of interest

None.

Abbreviations

PCL, Primary cardiac lymphoma; BL, Burkitt lymphoma; NHL, non-Hodgkin's lymphoma; ECG, Electrocardiogram; BNP, brain natriuretic peptide; CT, computed tomography; MRI, magnetic resonance imaging; AIDS, acquired immune deficiency syndrome; ASCT, autologous stem cell transplantation; TTE, transthoracic echocardiography; TEE, transesophageal echocardiography; PET-CT, positron emission tomography-computed-tomography.

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