

A case of complete atrioventricular block associated with primary cardiac lymphoma reversed without cardiac pacemaker implantation

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Abstract

Primary cardiac lymphoma (PCL) is a rare malignant lymphoma that is characteristically confined to the heart and/or pericardium. Here, the case of a 70-year-old male patient with complete atrioventricular block (AVB) associated with PCL is presented. The patient had a 10-month history of palpitation and electrocardiogram (ECG) showed a complete AVB. Additionally, transthoracic echocardiography indicated pericardial effusion where atypical lymphoid cells were identified by pericardiocentesis. Subsequent mediastinal lymph node biopsy revealed non-germinal centre diffuse large B-cell lymphoma. Therefore, a diagnosis of PCL was confirmed. As the patient's vital signs were stable, he was prescribed chemotherapy without pacemaker implantation. After chemotherapy, the patient achieved remission and dynamic ECG demonstrated no recurrence of AVB. The present case demonstrates that although PCL initially manifesting as complete AVB is rare, this possibility should not be ignored when a new AVB without definite aetiology is encountered. In addition, if the vital signs of the patient are stable, pacemaker implantation may be postponed until the treatment effect of chemotherapy has been assessed.

Keywords

Primary cardiac lymphoma, atrioventricular block, chemotherapy, case report, remission, pacemaker

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Introduction

Primary cardiac tumours are particularly rare, with a prevalence of 0.02%, and primary cardiac lymphoma (PCL) accounts for only 1.3% of primary cardiac tumours and 0.5% of extra nodal lymphomas.¹ The main manifestations of PCL are cardiac symptoms caused by lymphoma infiltration of the myocardium, which may be accompanied by mediastinal lymph node enlargement, pleural exudation, pulmonary embolism and other metastatic signs.² Some patients with PCL may present with atrioventricular block (AVB) only, which may be easily misdiagnosed.³ In the present report, clinical observations are shared with the purpose of improving understanding of PCL, and providing theoretical and empirical evidence for its clinical treatment and prognosis. The study protocol was approved by the Ethics Committee of Shanghai Changzheng Hospital and written informed consent was obtained from the patient for publication of this case and associated images. The reporting of this study conforms to CARE guidelines.⁴

Case report

A 70-year-old male patient presented to the Department of Cardiology, Shanghai Changzheng Hospital (the Second Affiliated Hospital of Naval Medical University), Shanghai, China, in April 2021, with a 10-month history of dyspnoea and palpitation after exertion that had worsened over the previous 5 days. Symptoms lasted for a few minutes with each occurrence and alleviated after rest. Physical examination showed a regular heart sound with cardiac murmurs, and his heart sounds were dull and muffled suggesting an enlarged heart. Abdominal and respiratory system examination showed no obvious abnormalities. The white blood cell count was within normal limits, but

haemoglobin and haematocrit values were reduced to 105 g/L and 32%, respectively. The electrocardiogram (ECG) showed a complete AVB (Figure 1a). Echocardiography and enhanced chest computed tomography (CT) both showed a mass lying in the left and right atrium (Figure 2 and Figure 3a). Transthoracic echocardiography showed pericardial effusion, and pericardiocentesis revealed atypical lymphoid cells, suggesting B-cell lymphoma. Results from CT-guided percutaneous biopsy of the mediastinal lymph node reported a diffuse large B-cell lymphoma with non-germinal centre type (Figure 4), and subsequent immunohistochemical analyses of lymph node biopsy tissue revealed positivity for apoptosis regulator Bcl-2, B-lymphocyte antigen CD20, B-lymphocyte antigen CD19, T-cell surface glycoprotein CD5, B-cell antigen receptor complex-associated protein alpha chain (CD79a), myc proto-oncogene protein (c-Myc), interferon regulatory factor 4 (also known as multiple myeloma oncogene 1 [MUM-1]), and paired box protein Pax-5. Positron emission tomography (PET)-CT showed space-occupying lesions in the atrium, accompanied by enlargement of multiple mediastinal lymph nodes, and pericardium lesions, all of which were considered to be lymphoma. Since all imaging examinations indicated that the lesion was confined to the mediastinum without distant metastasis, and both pericardium puncture and mediastinal lymph node biopsy found diffuse large B lymphoma cells, the final diagnosis was PCL. Of note, AVB is regarded as a significant clinical manifestation of PCL.

Considering that the vital signs of the patient were stable (temperature, 36.2°C; blood pressure, 123/74 mmHg; pulse, 70 beats per min; and respiratory rate, 18 breaths per min), the decision was made to prescribe initial chemotherapy without implantation of a pacemaker, and to observe whether the complete AVB would disappear.

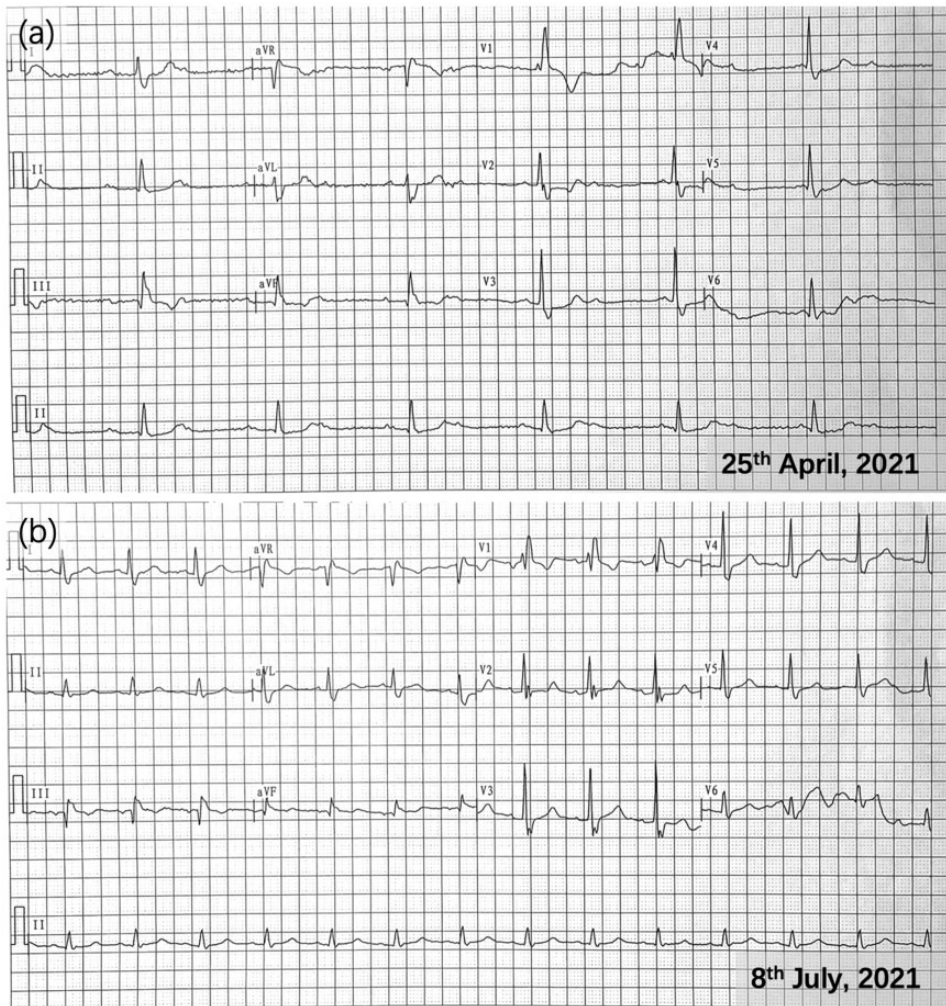


Figure 1. Representative 12-lead electrocardiograms from a 70-year-old male patient diagnosed with complete atrioventricular block associated with primary cardiac lymphoma: (a) before and (b) after treatment.

The patient was transferred to the haematology department of Shanghai Changzheng Hospital (the Second Affiliated Hospital of Naval Medical University) to receive a chemotherapy regimen of dose-adjusted (DA)-etoposide, prednisone, vincristine, cyclophosphamide, and doxorubicin (EPOCH) plus rituximab, supplemented with related supportive therapy (hydration, antacids and antiemetics).

After two cycles of chemotherapy, his symptoms had relieved and repeat ECG showed a relatively normal signal with occasional atrial or ventricular premature beats and without complete AVB (Figure 1b). After five cycles of chemotherapy, the patient achieved complete remission, and repeat enhanced chest CT indicated a much smaller mass in the pericardium than before treatment (Figure 3b).

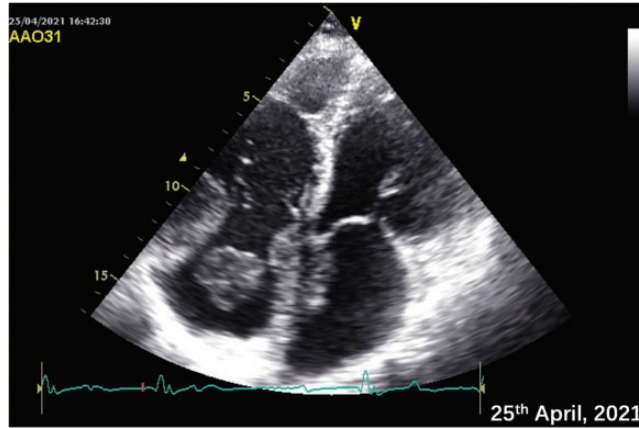


Figure 2. Representative echocardiography image from a 70-year-old male patient diagnosed with complete atrioventricular block associated with primary cardiac lymphoma, showing a mass located in the atria and atrial septum.

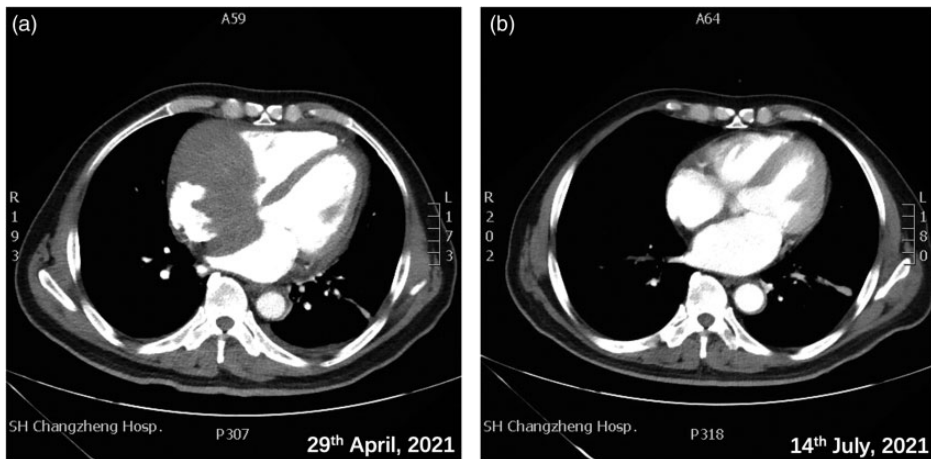


Figure 3. Enhanced chest computed tomography images from a 70-year-old male patient diagnosed with complete atrioventricular block associated with primary cardiac lymphoma: (a) before treatment, showing a mass in the atria; and (b) after treatment, showing a reduction in the size of the mass.

The patient was regularly followed-up, and at 6 months after treatment, he had no recurrence of his initial symptoms.

Discussion

Primary cardiac lymphoma is relatively rare, accounting for less than 2% of cardiac tumours and 0.5% of extra nodal

lymphomas,⁵ and is more common in the elderly and in male patients. Symptoms of PCL vary, and arrhythmia, ranging from premature atrial beats to complete AVB, can act as the first manifestation. The right side of the heart seems to be more often involved than the left.⁶ Therefore, it is easily misdiagnosed and patients with suspected PCL should be examined

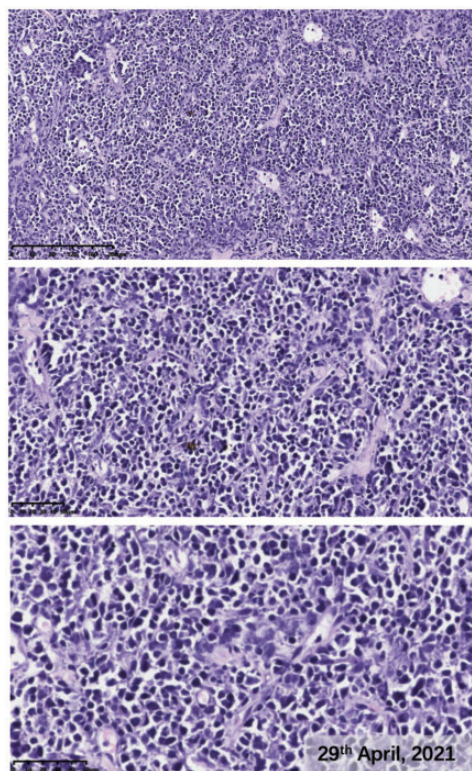


Figure 4. Representative haematoxylin and eosin-stained mediastinal lymph node tissue sections showing diffuse large B-cell lymphoma cells with non-germinal centre type (original magnification, 100 \times ; 250 \times ; 350 \times ; upper, middle and lower images, respectively).

thoroughly. The current patient presented with non-specific dyspnoea on exertion and palpitation, and echocardiography and chest CT showed a mass in the atria. Information about the location and morphology of lesions occupying the pericardium, and the relationship with surrounding structures, can be provided by echocardiography, chest CT and magnetic resonance imaging.^{7,8} A final diagnosis may only be made by histological evaluation of the tumour. In the present case, CT was used to explore the cardiac mass and mediastinal lymphadenopathy. Finally, CT-guided percutaneous biopsy of mediastinal lymph

nodes confirmed the diagnosis of lymphoma. The prognosis of patients with cardiac lymphoma is usually poor, with a median survival time of approximately 12 months.⁹ Rapid progression of cardiac involvement and generally late diagnosis are the main factors affecting prognosis. Thus, timely diagnosis and treatment is key to improving patient survival.

As previously mentioned, PCL with complete AVB as the first manifestation is extremely rare, and related treatment experience is scarce, with only a few case reports describing AVB as a major clinical presentation of PCL. Most of these patients received either a permanent or temporary pacemaker,^{10–17} but a few patients with stable haemodynamics did not receive pacemaker implantation.^{18,19} All of the patients underwent chemotherapy and prognosis varied. In the present case, although the patient had a complete AVB, his vital signs and haemodynamics were stable. Therefore, chemotherapy was selected for initial treatment, with a view of making the decision regarding whether to implant a pacemaker upon review of the post-chemotherapy ECG. Cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP) and DA-EPOCH chemotherapy regimens both represent treatment options for aggressive lymphoma. The DA-EPOCH regimen is indicated to exhibit excellent efficacy for aggressive diffuse large B-cell lymphoma,²⁰ and is expected to become the first-line treatment for high-risk diffuse large B-cell lymphoma.²¹ Hence, the patient in the present case received DA-EPOCH chemotherapy, and complete remission was eventually achieved. In addition, the dynamic ECG indicated an absence of complete AVB and, therefore, it was unnecessary to implant a pacemaker. The patient was followed-up regularly in the haematology department and his condition remained stable.

In summary, PCL should be included in the differential diagnosis when identifying the aetiology of new onset AVB. Early imaging examination and timely histopathological diagnosis are very important in diagnosing PCL. The present case describes a patient with PCL whose primary clinical presentation was complete AVB, and provides a feasible treatment modality for this type of disease.

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Author contributions

Bowen Hu and Jian Zhao collected the data and wrote the manuscript; Na Li and Xin Liang provided relative details of the case; and Changzhen Ren was involved in managing the case. All authors have read and approved the submitted manuscript.


Declaration of conflicting interest


The Authors declare that there is no conflict of interest.

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