Primary Cardiac Lymphoma with Complete Atrioventricular Block

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Cardiac symptoms as an initial presentation of malignant lymphoma are rarely seen. We report a 42-year-old male patient of primary cardiac lymphoma originating in the left atrium with episodes of syncope. Complete atrioventricular (AV) block was found by electrocardiography, and the patient was admitted for pacemaker implantation. Before permanent pacemaker implantation, transthoracic echocardiography (TTE) revealed a massive infiltrative thickening in the left atrial and interatrial septal walls which initially mimicked an intra-atrial thrombus. After a series of imaging studies, including TTE and transesophageal echocardiography (TEE), multislice computed tomography (MSCT), fluoroscopic-guided endomyocardial tissue biopsy, and positron electric tomography (PET), large B cell type primary cardiac lymphoma was diagnosed. The patient died two months later in spite of epicardial permanent pacemaker implantation and two courses of chemotherapy treatment.

Key Words: Complete atrioventricular block • Primary cardiac lymphoma

INTRODUCTION

Primary cardiac lymphoma is a rare disease with a high mortality rate due to diagnosis commonly being made at an advanced stage of myocardial involvement. It is defined as non-Hodgkin’s lymphoma with only heart and/or pericardium involvement. Primary cardiac lymphoma accounts for 1.3% of primary cardiac tumors and 0.5% of the extranodal lymphomas.1 On the other hand, involvement of the heart by malignant lymphomas is more common, with an incidence rate of 15%–30% of all lymphomas in various post-mortem series.2,3 The prognosis of both primary and disseminated cardiac lymphoma remains grave, chiefly due to late presentation and substantial delay in the diagnosis. There is no pathognomic picture suggestive of primary cardiac lymphoma clinically. All clinical presentations are related to the site at which the primary cardiac lymphoma is involved.

CASE REPORT

A 42-year-old male patient had a history of diabetic nephropathy in uremic stage and had been receiving hemodialysis regularly. He was admitted due to frequent episodes of syncope for one month, as well as general malaise and poor appetite. He denied chest discomfort or shortness of breath. Bradycardia with a rate of 42 bpm and blood pressure of 84/50 mmHg were noted during his hospitalization. Electrocardiography (ECG) revealed complete atrioventricular block with junctional escape rhythm.
Before permanent pacemaker implantation, transthoracic echocardiography (TTE) revealed massive infiltrative thickening of the inferior aspect of the left atrium and interatrial septum that initially mimicked an intra-atrial thrombus (Figure 1A). However, transesophageal echocardiography (TEE) showed a suspected tumor mass attached to the interatrial septum with protrusion into both atrium cavities (Figure 1B). Tumor markers (CEA, AFP, CA 19-9) were all within normal range. For fear of tumor embolization, epicardial, instead of endocardial, pacemaker lead fixation was arranged. A pericardial tissue biopsy was performed during the procedure as well, which demonstrated fibrosis only.

ECG-gated multislice computed tomography (MSCT) was performed to investigate how much the tumor mass had affected the patient’s heart. It showed a bulky infiltrative mass of about $8.62 \times 6.45 \times 9$ cm at the posterior-lateral and inferior aspects of the left atrium and involvement of the interatrial septum, tricuspid valve, part of the right atrium, adjacent pericardium, left pulmonary hilum and pleura (Figures 1C, D). This tumor had homogeneous hypo-attenuation and no obvious enhancement after the administration of intravenous contrast agent as compared with the left ventricular myocardium.

A transvenous endomyocardial tissue biopsy was done for pathologic diagnosis. Immunohistochemically, the large tumor cells were strongly and diffusely positive for CLA (common leukocyte antigen) and CD20 (marker...
for B cells), admixed with small reactive CD3+/CD5+ T cells, but negative for cytokeratin, S-100, chromogranin, CD56, desmin, FLI-1, and CD34. The pathology report was consistent with diffuse large B cell lymphoma (Figure 2). We arranged positron electric tomography (PET) to search for the origin of the tumor and tumor metastasis. The FDG-PET scan revealed remarkably increased glucose metabolism in the right atrium, interatrial septum, left atrium and in a tumor mass protruding from the posterior aspect of the pericardium. Moreover, there was no abnormal radiotracer uptake in the pleura and lymph nodes. According to these results, primary B cell cardiac lymphoma was diagnosed.

The patient was thus referred to a hematologist for chemotherapy with regimens of CHOP (cyclophosphamide, doxorubicin, vincristine and prednisolone) for 3 courses, and rituximab for 2 courses. Before the first course of chemotherapy, bone marrow biopsy was performed, and no neoplastic cell was noted within the bone marrow specimen. One month later after the chemotherapy, general malaise was found during hemodialysis and also non-capture of pacing with junctional rhythm. To avoid the failure to capture, the pacemaker was adjusted by increasing the pacing output. Unfortunately, the patient died suddenly two months later after another course of chemotherapy.

DISCUSSION

Primary cardiac lymphomas are extremely rare, with a reported incidence of 0.15 to 1%.4 Diffuse B-cell lymphoma accounts for about 80% of primary cardiac lymphomas in immunocompetent patients, and small non-cleaved or immunoblastic lymphomas are more frequent in immunodeficient patients.5 Primary cardiac lymphoma predominantly invades the right atrium, followed by the right ventricle, left ventricle and interatrial septum.6 Clinical presentations are variable, including congestive heart failure, dyspnea, fainting, syncope, atrial or ventricular arrhythmia, neoplastic pulmonary embolism, pericardial effusion, pleural effusion, cardiac tamponade, superior vena cava syndrome, myocardial infarction, stroke, and sudden death. There is no specific serologic marker to identify primary cardiac lymphoma, and laboratory features only show the elevation of erythrocyte sedimentation rate, lactate dehydrogenase concentration, and interleukin-2 receptor concentration.7

Cardiac involvement as a first presentation of malignant lymphoma is rare. ECG findings include atrial flutter, atrial fibrillation, varied degree of AV block, bundle branch block, and low voltage, but these findings are nonspecific for malignant lymphoma. Chest roentgenography may just show increased heart size and pleural effusion.

TTE is a basic imaging modality for the evaluation of cardiac masses and the best imaging modality to depict small masses that arise from the cardiac valves. However, TTE for the visualization of extracardiac extensions is suboptimal for the evaluation of malignant cardiac masses. The use of TEE provides better visualization not only to differentiate tumors from thrombi but also to compare suspicious tumor masses with cardiac structure.8 Recently, MSCT has been increasingly utilized for cardiac imaging. The short image acquisition time for MSCT is advantageous in cardiac imaging as compared with the use of MRI. The use of ECG-gated MSCT provides better soft tissue contrast than echocardiography and can definitively characterize fatty content and calcifications. A wide field of view with MSCT helps to assess the extent of a cardiac malignancy and helps to detect metastatic lesions. Compared with echocardiography, MSCT imaging provides more precise assessment of morphology, composition, number, and the

Figure 2. The section shows monotonous proliferation of large centroblast-like lymphoid cells having round to irregular nuclei with vesicular chromatin and distinct one to three nucleoli (arrow), and a moderate amount of cytoplasm (H-E stain, 150X).
extent of cardiac tumors. Moreover, MSCT is superior to echocardiography in the detection of pericardium and paracardiac structures, such as the mediastinum, great vessels and lungs.\textsuperscript{9} Massive infiltration of lymphoma cells in the myocardium results in irregular thickening of atrial or ventricular walls, similar to a mural thrombus or hypertrophy. MSCT images can show cardiac lymphoma as hypo- or iso-attenuated infiltrative soft tissue mass as compared with the myocardium, and the mass demonstrates heterogeneous enhancement after the administration of a contrast agent.\textsuperscript{10}

Primary cardiac lymphomas are extremely rare among benign or malignant cardiac tumors. However, primary cardiac lymphomas have a high mortality rate. Non-specific symptoms appearing in the advanced stage could masquerade as any cardiac disease, and make primary cardiac lymphoma difficult to diagnose. Imaging features can help distinguish between benign and malignant cardiac tumors. Both TTE and TEE have limitations. The image acquisition time and better soft tissue contrast makes MSCT a powerful imaging modality to help clinicians in the diagnosis of primary cardiac lymphomas.

REFERENCES