

Diffuse Large B-Cell Cardiac Lymphoma: A Case Report

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Abstract

Cardiac lymphoma is an exceedingly rare malignancy with most cases classified as diffuse large B-cell lymphoma. Diagnosis is challenging due to nonspecific symptoms like heart failure, arrhythmias, or pericardial effusion. Imaging techniques, including echocardiography, PECT/CT, cardiac MRI, and CT, are pivotal for detecting cardiac masses, while definitive diagnosis requires histopathological confirmation. Treatment typically involves chemotherapy, with R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone) as the standard regimen.

Here we report on a 63-year-old gentleman who presented with chronic low-grade fever, night sweats and weight loss. Whole body FDG PET CT revealed FDG avid multiple enlarged lymph nodes, with FDG avid foci in the scrotum and left kidney and heterogenous foci of increased FDG uptake in the heart. Echocardiogram revealed large masses in the right and left atrium in keeping with cardiac involvement by DLBC lymphoma. The patient was started on chemotherapy and planned for six cycles (R-CHOP regimen). Follow up PET CT scan revealed complete resolution of the cardiac lesions.

Keywords: Lymphoma; Heart Neoplasms; Antineoplastic Combined Chemotherapy Protocols.

Introduction

Cardiac lymphoma is a rare form of extra nodal lymphoma, primarily classified into primary cardiac lymphoma (PCL) and secondary cardiac involvement in systemic lymphoma. PCL accounts for less than 1% of all extra nodal lymphomas and less than 2% of primary cardiac tumors and often goes undetected due to subclinical or non-specific symptoms with the highest incidence most observed in postmortem autopsy. Secondary cardiac involvement in systemic lymphoma is well-documented, with autopsies revealing cardiac infiltration in up to 30% of patients.¹

Here, we demonstrate a case of diffuse large B-cell lymphoma (DLBCL) who was incidentally found to have cardiac involvement in initial imaging, with no cardiac symptoms at presentation. Patient received the standard regimen of chemotherapy and achieved disease remission without reported complications.

Case Report

A 63-year-old male with no prior medical history presented with right testicular swelling, accompanied by low-grade fever for three months, night sweats, and unintentional weight loss of 7 kg in one month. An initial ultrasound revealed a 5.3 x 2.9 x 2.7 cm solid mass in the right testicle. He underwent a radical orchiectomy, and histopathology confirmed (DLBCL).

Staging fluorodeoxyglucose (FDG) position emission tomography/computed tomography (PET/CT) demonstrated involvement of supra- and infra-diaphragmatic lymph nodes, as well as the right scrotum and left kidney involvement. Additionally, the scan showed heterogeneous increased FDG uptake (SUVmax 14.4) in the right and left atrial regions of the heart (Figure 1). A transesophageal echocardiogram revealed a large, cauliflower-shaped mass in both the right atrium (RA) and left atrium (LA) (Figure 2). The RA mass, measuring 5 x 4 cm, originated from the inferior vena cava (IVC) and compressed the coronary sinus but spared the tricuspid and pulmonary valves. The LA mass, measuring 1.8 x 1.6 cm, was attached to the interatrial septum (IAS), with smaller masses on the aorto-mitral curtain and both atrial surfaces of the mitral valve, sparing the aortic valve. No significant regional wall motion abnormalities were noted, and the right ventricular function and left ventricular ejection fraction (LVEF) were normal at 64%.

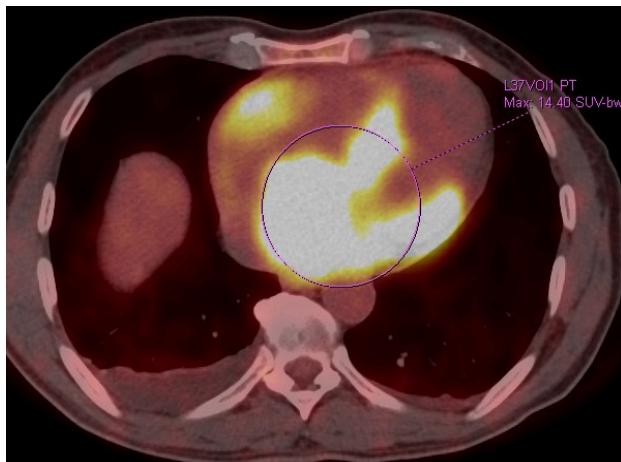


Figure 1: F-18 FDG PET/CT demonstrating intense radiotracer uptake localized to the right and left atrial regions of the heart.

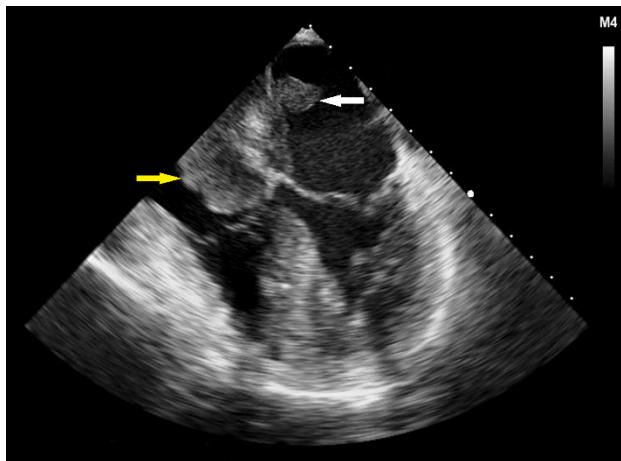


Figure 2: Transesophageal echocardiography demonstrating a large, cauliflower-shaped mass in the right atrium (yellow arrow) and a smaller mass in the left atrium (white arrow).

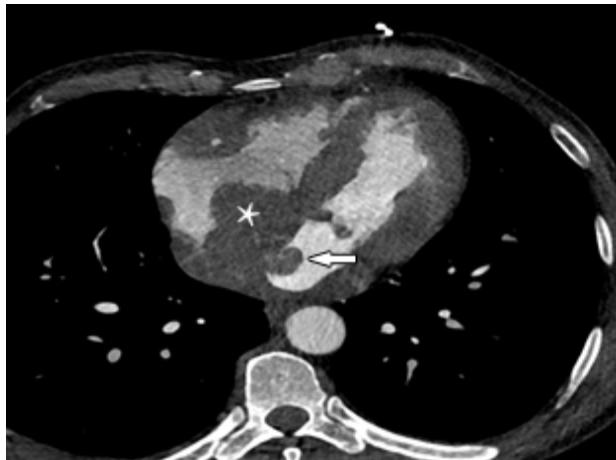


Figure 3: Contrast-enhanced cardiac CT demonstrating a broad-based right atrial mass infiltrating the interatrial septum, along with a smaller mass within the left atrium.

Contrast enhanced cardiac CT confirmed a broad-based right atrial mass (4.2 x 3.9 cm) infiltrating the interatrial septum, along with a smaller RA lesion (2.6 cm) near the anterior inferior aspect of the septum. The left atrium had a 1.6 cm mass based on the interatrial septum (Figure 3). The CT also showed thickening of the left ventricular basal lateral wall and soft tissue thickening in the right interventricular groove involving the right coronary artery (RCA) suggested infiltration by the lymphoma.

The case was discussed in a multidisciplinary team involving cardiologists, electrophysiology cardiologist, cardiothoracic surgeon and oncologist and was agreed to proceed with chemotherapy with no surgical intervention considering the maintained LVEF and absence of symptoms in terms of cardiovascular system with follow-up cardiac imaging. The patient started on six cycles of rituximab, cyclophosphamide, doxorubicin hydrochloride, vincristine sulfate and prednisone (R-CHOP) regimen. An interim PET-CT after the third cycle showed complete resolution of the previously observed heterogeneous myocardial activity, indicating a complete metabolic response (Figure 4). At the same setting, the patient was re-evaluated by cardiology, an echocardiogram at the same setting was repeated which showed a residual mass attached to the IAS in the right atrium measuring 14 x 11 mm with no other masses noted with again maintained normal ejection fraction. A cardiac magnetic resonance imaging (MRI) confirmed the disappearance of the previously visualized infiltrative soft tissue lesion in cardiac CT involving the atrioventricular groove and RCA. The patient was set to continue chemotherapy as planned with no additional intervention from cardiology or cardiovascular surgery apart from follow-up. Later, patient completed the treatment planned with six cycles of R-CHOP with no immediate complications.

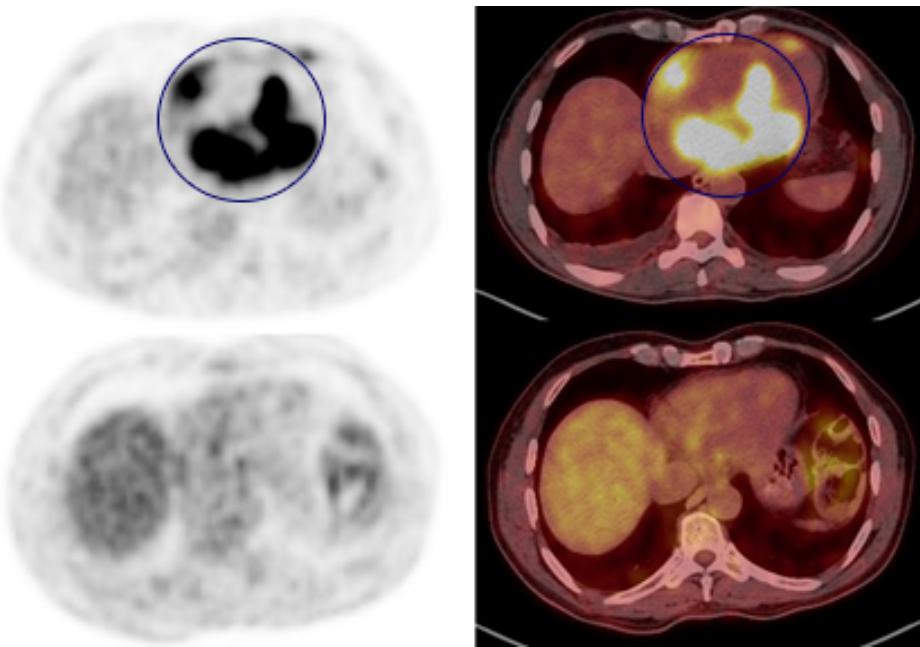


Figure 4: F18-FDG PET/CT axial images. follow-up scan after three cycles of chemotherapy demonstrating complete metabolic response.

Discussion

The presentation of cardiac lymphoma ranges widely, from being asymptomatic to causing non-specific symptoms such as dyspnea, chest pain, dysrhythmia, to causing life-threatening arrhythmias, therefore, it is frequently undetected before death. The right atrium is the most affected site in cardiac lymphoma. Pericardial thickening or effusion is often an early hallmark of the disease. Infiltration of the atrial or ventricular walls, with extension along the epicardial surfaces, is another notable characteristic. A distinctive aspect of cardiac lymphoma is its tendency to spread along the epicardial surfaces of the heart, frequently encasing nearby structures such as the coronary arteries and the aortic root. It commonly involves the right atrioventricular groove and the base of the heart.¹

PCLs were originally described as extra nodal lymphomas confined to the heart and/or pericardium; however, the definition has since been broadened to include lymphomas with cardiac involvement where most of the disease is localized to the heart. According to the World Health Organization, PCLs are characterized by lymphomas presenting with cardiac symptoms, with most of the tumors being intrapericardial at the time of presentation.² Secondary cardiac involvement in lymphoma, as in our case typically occurs as a late-stage manifestation of a widespread disease. Lymphomas spread to the heart by three pathways: (a) direct extension from mediastinal lymphoma, (b) via lymphatic vessels along coronary arteries and epicardium, and (c) hematogenous spread. However, whether the lymphoma is primary or secondary, there are no differences in imaging findings between the two.¹

Cardiac lymphoma is an exceptionally rare condition that poses significant diagnostic challenges due to its location in the heart and its often-nonspecific signs and symptoms, diagnosing cardiac lymphoma can be difficult, leading to delays in diagnosis and poor outcomes. Histopathology continues to be the gold standard for diagnosis. In terms of imaging, chest radiographs are neither highly sensitive nor specific. FCG PET/CT scan offers both anatomical and functional imaging and is frequently helpful in distinguishing DLBCL with secondary cardiac involvement from other cardiac tumor types. For assessing the extent of pericardial and myocardial involvement, cardiac MRI and contrast-enhanced CT are preferred, though MRI excels in providing better soft tissue characterization. Echocardiogram also remains a cheap, widely available, non-invasive useful imaging modality to repeatedly evaluate the tumor, cardiac function and assess response post treatment.³

No treatment guidelines exist; however, chemotherapy remains a cornerstone in the treatment of cardiac lymphoma. The standard regimen often includes anthracycline-containing chemoimmunotherapy. High-dose chemotherapy followed by autologous hematopoietic stem cell transplantation (AHSCT) has also been reported to achieve long-term remission in some cases.⁴ Surgical treatment is generally reserved for clinically unstable patients or when there is a need for immediate relief of symptoms such as superior vena cava syndrome,⁵ while it can provide immediate benefits, it does not necessarily improve long-term prognosis.⁶ A combination of therapies, including chemotherapy, surgery, and in some cases, heart transplantation, has been employed in complex cases. For instance, a case involving primary cardiac T-cell lymphoma was managed with a combination of chemotherapy, heart transplantation, and reduced immunosuppression, resulting in over two years of disease-free survival.⁷

It has been reported that primary cardiac lymphoma show an overall response rate of 79% to chemotherapy, with a complete remission rate of 59%. Comparable outcomes have also been observed in cases of secondary lymphoma. Lack of left ventricle involvement and arrhythmias are associated with improved survival,⁸ however, the outlook for cardiac lymphoma remains very poor. One study found that patients with primary cardiac B-cell NHL had better outcomes than those with secondary cardiac involvement (6 months versus 2 months),⁹ and a recent study found that DLBCL secondary cardiac lymphomas had a better median survival time compared to non-DLBCL secondary cardiac lymphomas (12 vs 2 months).¹⁰

Conclusion

Cardiac lymphoma, whether primary or secondary, remains a rare and diagnostically challenging condition with often nonspecific symptoms that can lead to delayed recognition and poor prognosis. Given its varied presentation and the absence of distinct imaging differences, histopathological confirmation is essential for diagnosis. While chemotherapy remains the mainstay of treatment, alternative regimen and multimodal approaches, including stem cell transplantation and, in rare cases, surgical intervention, may offer some benefit. However, overall survival remains limited with overall poor prognosis due to often late diagnosis consequent to non-specific symptoms.

In our case we highlight the importance of establishing early diagnosis and timely initiation of treatment as they significantly influence outcomes. In this case, conventional chemotherapy, combined with thorough follow-up and without surgical intervention, proved successful.

Patient Consent

The patient provided informed consent for publication of our case report.

Disclosure statement

The authors declare no conflicts of interest.

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