

Non-Hodgkins Lymphoma Involving Multiple Cardiac Chambers with Skeletal Muscle Involvement in a Hepatitis C Positive Patient

Behranwala A, Bharat Shivdasani, Nihar Mehta* and Amit Chandan

Jaslok Hospital and Research Centre, Mumbai, Maharashtra, India

*Corresponding author: Nihar Mehta, Jaslok Hospital and Research Centre, Mumbai, Maharashtra, India, Tel: 91-9819139900; Email: dr.niharmehta@gmail.com

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Introduction

Primary cardiac lymphomas are rare and involvement of multiple cardiac chambers is not common. It usually invades primarily the right atrium, followed by the right ventricle, the left ventricle, and finally the atrial septum. Skeletal muscle involvement is also rare. FDG PET Scan is useful in diagnosis and in determining the disease extent, site for biopsy and its response to treatment.

We present a rare case of a Primary Cardiac Lymphoma in a Hepatitis C positive male which developed 3 years after he underwent CABG Surgery, which involved the right atrium, inter-atrial septum and left atrium and prolapsed across the tricuspid valve into the right ventricle. Additionally there was involvement of the Internal Oblique muscle that was proven on biopsy. FDG PET CT Scan was a useful guide to determine the extent of involvement, select the site of biopsy and evaluate the response to chemotherapy.

Case Report

A 53 year old Indian male, seropositive for hepatitis C virus, with a past history of coronary artery bypass surgery 3 years back, presented with recent onset dyspnea on exertion Class II and chest discomfort in April 2014. His 2D Echocardiography and trans esophageal echocardiography showed a large mass occupying nearly all of the right atrium, attached to lateral wall and protruding into the inter-atrial septum. The mass was lobulated with a variegated appearance. The mass was prolapsing across the tricuspid valve into the right ventricle. There was an extension of the mass across the inter-atrial septum into the left atrium. Cardiac function was normal with no pericardial effusion or pulmonary hypertension (Figure 1).

CT scan of chest, abdomen and pelvis was done which confirmed the findings of a large 6.1 X 4.9 cm lobulated mass within right atrium, extending into the left atrium, inter-ventricular septum and right ventricle. It was associated with lesion in the spleen and multiple enlarged pre-aortic, pre-caval and bilateral para aortic lymph nodes. A CT guided trucut biopsy of left para-aortic lymph node showed reactive lymphoid tissue positive for CD20.

An 18-FDG PET CT Scan showed metabolically active intra-cardiac lobulated lesion in the right atrium with extension through tricuspid valve into the right ventricle and inter-ventricular septum as well as extension across the septum into the left atrium, 9.4 × 4.6 cm in size. Two lobulated lesions were noted in spleen. The left internal oblique abdominal muscle as well as cervical lymph nodes and retroperitoneal lymph nodes also show metabolically active lesions (Figure 2). An excisional biopsy of left internal oblique abdominal muscle showed a high-grade Diffuse B cell Non Hodgkins lymphoma, CD20 positive in tumor cells.

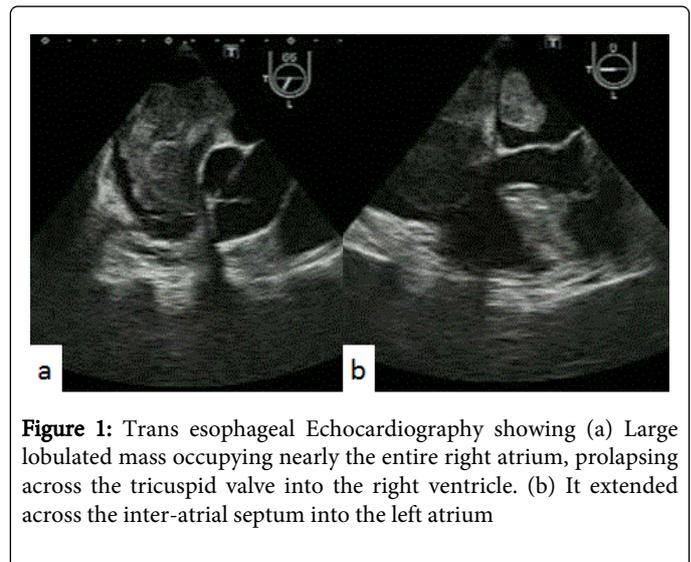


Figure 1: Trans esophageal Echocardiography showing (a) Large lobulated mass occupying nearly the entire right atrium, prolapsing across the tricuspid valve into the right ventricle. (b) It extended across the inter-atrial septum into the left atrium

Patient was given 8 cycles of R-CHOP (Rituximab + cisplatin + hydroxyurea + oncovin + prednisolone) chemotherapy regimen at the interval of 21 days. Post chemotherapy, the patient remains asymptomatic and on regular follow up with a series of echocardiographies, has shown a regular reduction in the lobulated mass, with the last echocardiography in November 2014 showing a small residual pedunculated right atrial tumour of 1.8 × 1 cm dimension and complete resolution from the left atrium and right ventricle. However, PET Scan showed no metabolic activity in the residual tissue (Figure 3).

Discussion

Primary cardiac lymphoma (PCL) is defined as non-Hodgkin's lymphoma (NHL) involving only the heart and/or the pericardium [1]. The incidence of primary cardiac tumors is approximately 0.02%, and PCLs account for approximately 1% of primary cardiac tumors and the majority is diffuse large B-cell lymphoma [2]. Secondary involvement from extra cardiac tumors is 20-40 times more common than primary cardiac tumors [3]. In our case we are unsure whether our patient had primary or secondary cardiac involvement, but the predominant tumor burden was cardiac, implying primary cardiac lymphoma. Cardiac lymphoma most commonly presents as a nodular or polypoid mass with variable myocardial infiltration [4]. Cardiac lymphoma invades primarily the right atrium, followed by the right ventricle, the left ventricle, and finally the atrial septum [5,6]. Although involvement of multiple cardiac chambers is not common, our patient had involvement of primarily the right atrium involving the right ventricle and across the septum into the left atrium (Figures

1, 2a and 2b). PCL occurs more commonly in immune compromised patients [7,8]. Our patient was hepatitis C positive but there is no correlation found between hepatitis C and cardiac lymphomas in literature except a single case report mentioning asymptomatic cardiac lymphoma in HCV positive thalassemic patient [9]. Though symptoms are non-specific and vary with the involved chamber of the hearts, the most common clinical manifestations are pericardial effusion, heart failure and AV block [10].

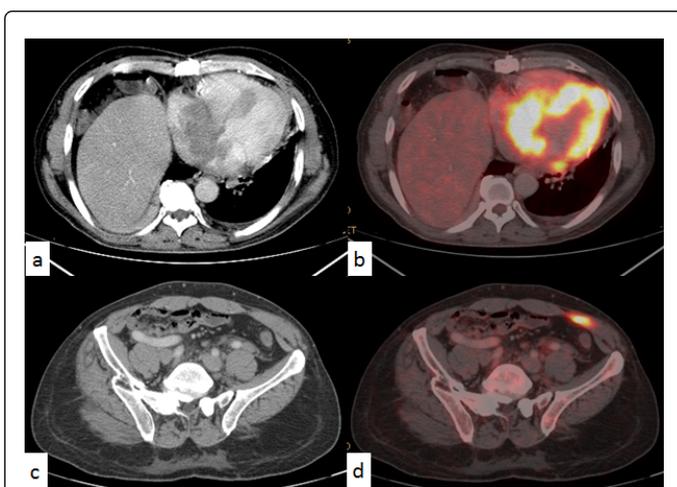


Figure 2: (a) CT Scan with contrast showing a large lobulated mass within right atrium, extending into the left atrium, inter-ventricular septum and right ventricle. (b) PET CT Scan showed metabolically active intra-cardiac lesion in right atrium with extension through tricuspid valve into the right ventricle and inter ventricular septum as well as extension across the septum into the left atrium (c and d) PET CT Scan showing metabolically active lesion in the left internal oblique muscle.

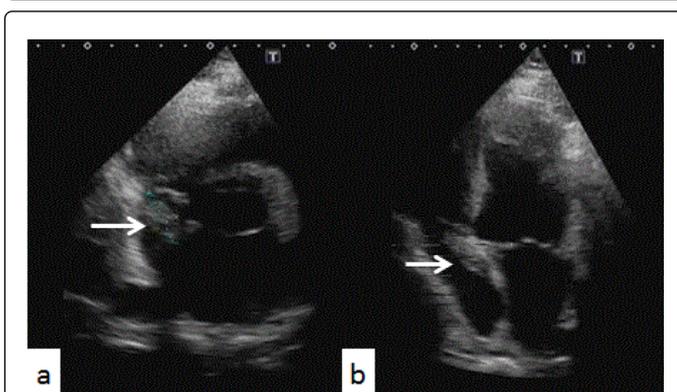


Figure 3: 2D Echocardiography on follow up showing a small residual pedunculated right atrial tumour in (a) parasternal short axis view and (b) apical four chamber view

Echocardiography is the initial diagnostic method that can detect pericardial effusion and the presence of tumors in the heart, followed by CT and MRI scans to detect the extent of involvement and the size of the tumor [11]. Serial PET has been suggested to be more accurate than MRI and echocardiography for assessing cardiac lymphoma regression [12]. Cardiac lymphomas can be distinguished from other

cardiac neoplasms on the basis of their reactivity for common leukocyte antigen and pan B-cell markers (CD 20) or pan T-cell makers (CD 3). Diffuse B-cell lymphoma accounts for about 80% of primary cardiac lymphomas in immunocompetent patients. In immunodeficiency patients, small non-cleaved or immunoblastic lymphomas are more frequent [13]. Our patient also had skeletal muscle involvement of the left internal oblique muscle, which was biopsied (Figure 2c and 2d)). Primary skeletal muscle lymphomas are very rare, accounting for approximately 1.5% of Non-Hodgkin lymphomas [14] and are associated with a poor prognosis. Mostly skeletal muscle involvement occurs secondary to hematogenous dissemination. FDG PET is useful in detecting the muscle site involved, which is usually an asymmetric focus or multiple foci of FDG uptake [15]. The site of increased uptake can be used as for biopsy, as was in our case. PCL is chemo-radiosensitive and with prompt treatment the survival of these patients can be improved. The choice of chemotherapy for PCL remains the Rituximab and CHOP regimen [5,16].

Conclusion

Primary cardiac lymphoma is a rare variety of Non-Hodgkins Lymphoma. Involvement of multiple cardiac chambers is not common. It is more common in immune compromised states but a relation to Hepatitis C seropositive state is less established. Echocardiography is used for initial diagnosis. However, FDG PET-CT Scan is the modality of choice for determining the disease extent and its response to treatment. Skeletal muscle involvement is also rare and FDG PET Scan is useful in diagnosis and in determining the site for biopsy. Chemotherapy improves the prognosis and survival of these patients.

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