

## DIFFUSE LARGE B CELL LYMPHOMA OF THE HEART: A CASE REPORT

Dr. Sura Adnan Raheem<sup>\*1</sup>, Alladin Sahham Naji<sup>2</sup>, Ghazi Farhan Haji<sup>3</sup> and Mahasen M. Abd Al-Majeed<sup>4</sup>

<sup>1</sup>Echocardiography Specialist, Department of Cardiac Cathetrization, Baghdad Teaching Hospital.

<sup>2</sup>Haematologist, Haematology Department, Baghdad Teaching Hospital.

<sup>3</sup>Assistant Professor, Cardiologist, University of Baghdad, College of Medicine, Baghdad, Iraq.

<sup>4</sup>Department of Physiology, College of Medicine, Al-Iraqia University.

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\*Corresponding author: Dr. Sura Adnan Raheem

Echocardiography Specialist, Department of Cardiac Cathetrization, Baghdad Teaching Hospital.

### ABSTRACT

Primary malignant cardiac neoplasms are enormously rare including primary cardiac lymphomas which represent only a slight entity. Though the heart in diffuse B cell malignant lymphoma is rare as a primary presentation and involvement, the early diagnosis with suitable therapy will offer a great cure. Herein we report a 39 Years old female with a history of worsening dyspnoea for three weeks. On examination, she had a normal S1, loud S2 with early diastolic murmur at the upper left sternal edge, decreased breathing sounds in the left lung base and hepatosplenomegaly with no peripheral lymphadenopathy. Blood picture revealed an increases erythrocyte sedimentation rate with neutrophilic leucocytosis with a cellular bone marrow result. Chest x-ray showed a widening mediastinum with left pleural effusion. Trans thoracic echocardiography revealed mild pulmonary hypertension, lobulated homogenous cardiac mass in the right ventricle outlet (RVOT) with an extra cardiac mass invading the wall of the RV. The histopathological result of the mediastinal mass biopsy revealed a picture of diffuse large B cell lymphoma of stage IV B. Chemotherapy was started accordingly resulting in considerable improvement of the patient general condition in addition to the dramatic regression in the cardiac mass. But as the patient neglected the treatment, she was ended with septic shock and died because of it.

**KEYWORDS:** Lymphoma, cardiac involvement, surgical biopsy.

### INTRODUCTION

Without a doubt the Cardiac involvement by malignant lymphoma represents uncommon condition.<sup>[1,2]</sup> examined a series of autopsies to state that the cardiac involvement is found in about 20% of patients with end stage malignant lymphoma. However, it is hard to some extend to give an obvious diagnosis for those patients since the difficulty of carrying out a surgical biopsy, which generally cannot be achieved because of the risk of cardiac failure. Consequently, large numbers of those patients pass away before taking chemotherapy and they are diagnosed post-mortem.<sup>[3,4]</sup> addressed that a malignant lymphoma with cardiac involvement is commonly causes focal masses, which are usually located in the cardiac wall. On the other hand, the cardiac involvement may take a diffuse pattern of involvement, which is rare condition. Besides, the classical imaging examinations, for instance, chest radiography or usual computed tomography are not sufficient for detection diffuse cardiac involvement.<sup>[4]</sup>

With reference to the transthoracic echocardiography (TTE), it is considered a sensitive technique for recognition of cardiac involvement by lymphomas, which usually appear as polypoid or nodular masses in right chambers with different levels of myocardial infiltration.<sup>[5,6]</sup> pointed to the fact that the Magnetic Resonance Imaging (MRI) represents the gold standard, where it allows to distinguish between different types of cardiac masses. Though, the MRI necessitates the patient's hemodynamic stability.

This case report details a case of non-Hodgkin lymphoma of Diffuse Large B-Cell type with diffuse cardiac involvement and illustrates the significance of prompt diagnosis and pharmacologic management of this disease.

### CASE PRESENTATION

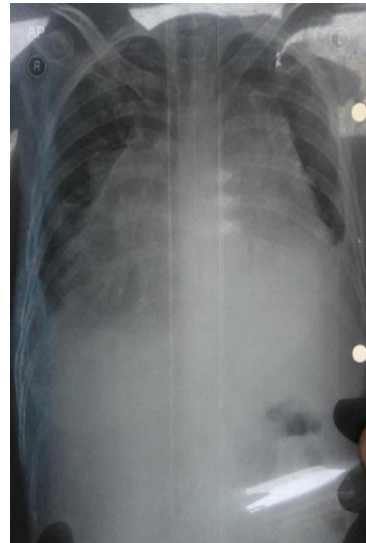
A 39 years old woman admitted to the Baghdad Teaching Hospital, cardiology department with a history of worsening dyspnea and constitutional symptoms such

as night sweat and non-significant weight loss over a period of three weeks. On admission she complained from resting shortness of breath (New York Heart Association class IV). Her blood pressure was 110/65 mmHg with pulse rate of 110 bpm and respiratory rate of 30 breaths per minute. She had normal S1 and loud S2 with early diastolic murmur at the upper left sternal edge. Additionally, pulmonary auscultation revealed decreased breath sounds in left lung base. What is more, the hepatosplenomegaly was observed with no peripheral lymphadenopathy. Finally, the patient had a negative past medical and drug history. The electrocardiography [figure 1] showed sinus tachycardia, normal axis, Q wave in the inferior leads I, III and aVF with a wide, prominent R wave in V1 (incomplete left bundle branch block). The white blood cell count for the patient was 16.800/IU, with 86.0% neutrophils, 10% lymphocytes, 3.0% monocytes, 1.0% eosinophils and 0% basophils. Additionally, the patient had thrombocytosis where the platelet count was 750.000/ IU. Furthermore, she had an elevated level of Erythrocyte Sedimentation Rate (ESR). Additionally, Virology screening was negative. Moreover, the patient had normal respiratory function test.

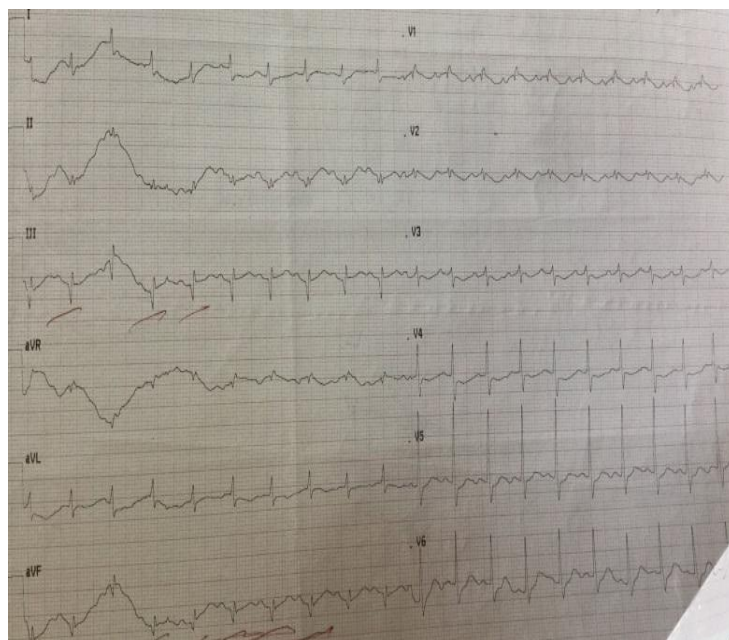
As far as chest x-ray [figure 2] is concerned, it addressed that there was an evidence of large central opacity with a lobulated contour including the heart, which was obliterating post sternal space. This was diagnosed as a large anterior mediastinal mass with left sided pleural effusion.

Regarding the Transthoracic Echocardiography scan (TTE), it showed a mildly dilated Right Ventricle (RV) and Right Atrium (RA), mild Tricuspid Regurgitation (TR) with transvalvular pressure gradient of (PG)=37

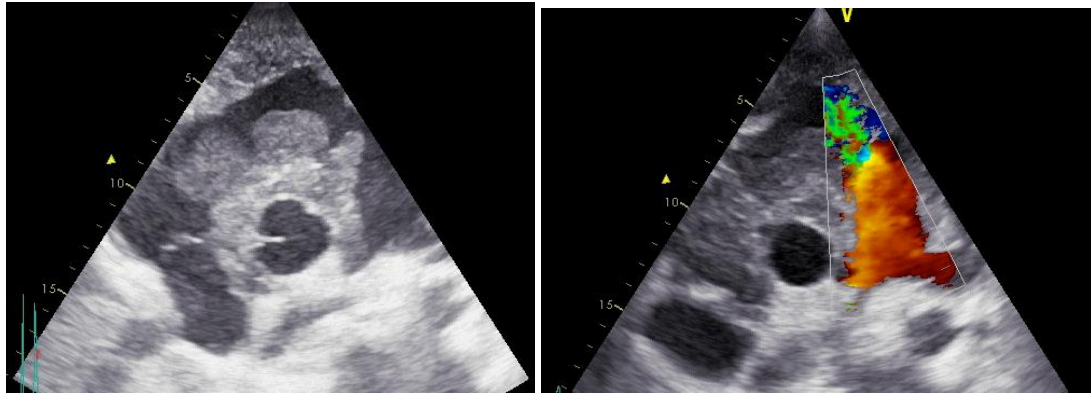
mmHg. Furthermore, the inferior vena cava (IVC) was non collapsible normal size. These finding together indicate a mild degree of pulmonary hypertension. Additionally, large extra cardiac mass invading the right ventricle (RV) wall. Severe pulmonary insufficiency (PI) was detected, with dilated pulmonary artery (pulmonary valve annulus diameter =30 mm) with large multiple homogenous lobulated mass occupying the Right Ventricular Outflow Tract (RVOT) and PV annulus, small pericardial effusion with no evidence of pericardial tamponade, grade I left ventricular (LV) diastolic dysfunction with mild mitral regurgitation (MR).



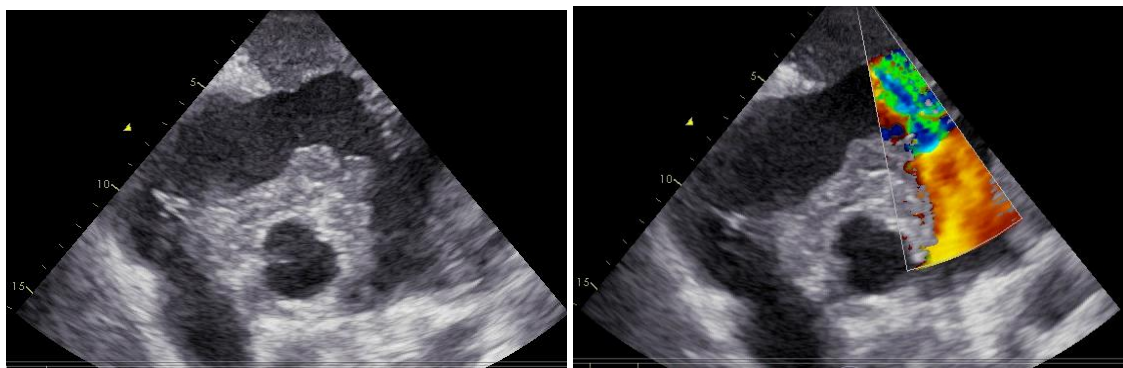
**Figure 1:** The electrocardiography showed sinus tachycardia, normal axis, Q wave in the inferior leads I, III and aVF with a wide, prominent R wave in V1 (incomplete left bundle branch block).



**Figure 2:** chest x-ray is addressed that there is an evidence of large central opacity with a lobulated contour including the heart, which was obliterating post sternal space. This was diagnosed as a large anterior mediastinal mass with left sided pleural effusion.



**Figure 3: Initial TTE Short Axis Views a. Large Lobulated Mass Occupying The Right Ventricular Outflow Tract (RVOT) and Pulmonary Valve (PV) b. Colour Doppler Across The PA With Severe Pulmonary Insufficiency (PI).**



**Figure 4: TTE Short Axis Views – After Second Dose R-CHOP regimen a. Large Lobulated Mass Occupying The Right Ventricular Outflow Tract (RVOT) and Pulmonary Valve (PV) b. Colour Doppler Across The PA With Severe Pulmonary Insufficiency (PI).**

The chest Computed Tomography (CT) scan with intravenous (IV) contrast was carried out and showed the superior and the anterior mediastinum occupied by a large lobulated mildly enhanced mass and encasing the vessel from the heart to the roof of the neck. Furthermore, the heart was enlarged with a significant evidence of large mass within the RA and the RV that can be probably diagnosed as a thrombus. Additionally, the CT scan showed a left sided pleural effusion. Besides, the mild enlargement in both of the liver and the spleen.

Pleural fluid aspiration was done with fluid analysis showed that there is a remarkable increase in lactate dehydrogenase (LDH), total protein level, and albumin giving a picture of an exudate pattern. It is interesting to mention here that the examination of bone marrow aspirate and biopsy was performed and showed a cellular marrow. Consequently, a surgical biopsy of the mediastinal mass was carried out to the patient. An initial differential diagnosis of the cardiac mass was lymphoma, thrombus, myxoma, sarcoma, metastasis and sarcoma. Over the period of waiting for the histopathological results of the biopsy, the patient was treated with corticosteroid and as the patient is considered as (class IIa) according to the ESC Guidelines on the diagnosis and management of acute pulmonary embolism. The

patient was treated with anticoagulant while waiting for histopathological results.

The histopathological result of the mediastinal mass biopsy revealed a picture of diffuse large B cell lymphoma of stage IV B. The initial treatment, which was corticosteroid and anticoagulant, was ended and substituted by chemotherapy R-CHOP regimen for six doses over 15 weeks. It is worth to mention here that an echocardiography scan was additionally carried out for the patient before starting the chemotherapy, the findings were completely similar to the finding of the initial echocardiography scan.

After the chemotherapy R-CHOP regimen treatment, a CT scan was ordered which shown that there wasn't any change in the size of the mediastinal mass. Because of the presence of lung metastasis, the patient was not referred to the radical radiotherapy. For this reason, the second line chemotherapy R-ICE regimen was highly recommended as the next stage for treatment.

During follow-up, the patient was examined by TTE after the second and the third dose of chemotherapy, there was a significant decrease in the cardiac mass size (about 50%) in the first TTE examination, while the second TTE presented a considerable reduction in the size of cardiac mass size reached to about 80-90%.

After the first dose of the R-ICE regimen, there was improvement in the general condition of the patient with a notable reduction in the size of mediastinal mass. Thus, the patient neglected the second dose of the R-ICE regimen and this leads to the development of neutropenic fever. As a result, the patient received empirical antibiotics and then a second dose of R-ICE regimen. After the second R-ICE regimen, the patient was referred by haematologist to take a palliative radiotherapy. However, the patient neglected the palliative radiotherapy and this leads to deterioration of her condition ending with septic shock. After few days, the patient passed.



**Figure 4: follow up TTE Short Axis Views shows noticeable regression of mass size after the third dose of chemotherapy.**

## DISCUSSION

Primary cardiac lymphomas Pharmacologic treatment leads to complete remission in more than 60% of patients, as well as surgical eradication of the tumour is associated with worse prognosis.<sup>[7,8]</sup>

However, usage of standard chemotherapy for patients with diffuse large B cell-lymphoma have a poor prognosis and the treatment should be designed according to the patient's comorbidities.<sup>[1]</sup>

Considering cardiac non-Hodgkin lymphoma, the most common histologic subtype is diffuse large B-cell lymphoma (58%), followed by (16%) T-cell lymphoma, (9%), Burkitt's lymphoma and (6%) small lymphocytic lymphoma.<sup>[9]</sup>

The most common clinical presentation is Symptomatic heart failure, as in our case report, but some patients may experience no cardiac symptoms.

However, the overall survival is generally poor, with a median survival duration of approximately 3 months, but immuno-chemotherapy strongly improves the survival.<sup>[1]</sup>

Echocardiography has been described to be a valuable examination method for assessing the cardiac involvement of malignant lymphomas; showing a mass

lesion, pericardial effusion and thickened ventricular wall.<sup>[10]</sup>

In contrast, computed tomography (CT) scanning did not offer perfect images of the inner structure of the cardiac wall. However, CT scans could distinguish mediastinal lymphadenopathy.<sup>[3]</sup>

The imaging modalities we did providing, in combination, with surgical biopsy that lead to clear evidence of cardiac involvement by diffuse B cell lymphoma.

Unlike other types of cardiac malignancies, Primary cardiac lymphoma, responds to chemotherapy. In the case described here, there was marked regression in the tumour after initiation of R-CHOP therapy without any cardiac complications.

Finally if there are clinical and radiological suspicions of primary cardiac lymphoma, aggressive diagnostic techniques should be done, and treatment should be initiated before irreversible cardiac injury happens.

## CONCLUSION

- The cardiac involvement rarely represents the initial presentation of lymphomas. Thus, the indication of cardiac involvement at initial presentation stage increases the suspension of other differential diagnosis such as thrombus.
- The cardiac involvement can be considerably detected using Echocardiography examination particularly when other diagnostic techniques are contraindicated.
- In case of existing a doubtful evidence of pulmonary embolism, the initiation of anticoagulant therapy should be established based on the factor of risk and benefit for the patient.
- Lymphoma should be taken into account in case of existing of cardiac masses.
- The surgical removal of the cardiac mass is not essential to accomplish complete remission as remission can be induced by chemotherapy.

**Conflict of interest:** None.

## Consent

Informed consent was obtained from the patient relatives for publication of this case report and any accompanying images.

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