Diffuse Large B Cell Lymphoma with uncommon cardiac invasion: Case Report from Palestine

Yunis Daralammouri

An-Najah National University

Hamza Hamayel

An-Najah National University

Yahya Ismail

An-Najah National University

Sajed Majadla

An-Najah National University

Razan Odeh (✉ R.odeh@najah.edu)

An-Najah National University

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Abstract

Background: Diffuse large B cell lymphoma (DLBCL) is the most common histologic subtype of non-Hodgkin lymphoma. Secondary involvement of the heart is seen late in advanced cases and it is uncommon for DLBCL to present as a cardiac mass.

Case Presentation: A 26-year-old female patient presented with progressive shortness of breath, facial swelling, and lower limb swelling. Echocardiography showed a large right atrial mass that was obstructing the tricuspid valve. Computed topography (CT) scan confirmed the findings and a large mediastinal mass with a filling defect in the right atrium occluding the distal Superior Vena Cava was identified. Histopathology showed DLBCL. She received 6 cycles R-CHOP chemotherapy (rituximab, cyclophosphamide, epirubicin, vincristine, and prednisone) with good response and complete resolution of the mass.

Conclusions: DLBCL can present with atypical, uncommon sites as the first presentation, such as the heart. Early diagnosis and appropriate management is crucial given the poor outcome with late presentation. A high index of suspicion and the proper investigations including CT scan is recommended to allow for early intervention and favorable outcomes as what happened with the case under discussion.

Background

Diffuse large B cell lymphoma (DLBCL) accounts for approximately 25 percent of non-Hodgkin lymphoma (NHL), which is the most common histologic subtype of NHL (1). Mediastinal mass presentation accounts for 2-4 % of all NHL (2).

Despite that secondary involvement of the heart is seen in 8.7–27.2% of documented clinical cases of lymphoma (1), it occurs mostly in advanced disease or remains silent and often diagnosed on autopsy. Intra-cardiac mass is a very rare presentation of DLBL (3-6).

This is a case report of a 26-year-old immunocompetent female patient presenting with gross intracardiac mass and Superior Vena Cava syndrome.

Case Presentation

A 26 year-old immunocompetent lady with unremarkable medical history presented to our hospital with progressive shortness of breath, facial swelling, upper limb swelling, and mild lower limb swelling for 3 months. She has no history of weight loss, night sweats, or fever. The rest of the history is unremarkable.

On admission, she was hemodynamically unstable, Blood Pressure 85/60, Heart Rate 116, Temperature 36.3, Saturation 96% on room air. She was tachypnic. She had facial swelling, raised jugular venous pressure, along with upper limb swelling, and mild lower limb swelling. She had distant heart sounds, a

diastolic murmur best heard in the right lower sternal border, lung exam revealed decreased breath sounds with dullness on percussion on bilateral basal lung zones. Chest x-Ray showed bilateral pleural effusion, wide mediastinum, and bilateral hilar congestion (Figure e-1). Electrocardiogram showed sinus rhythm, Q wave in V1, and low voltage QRS complex.

Transthoracic Echocardiograph showed a right large atrial mass obstructing the tricuspid valve with moderate pericardial effusion (Figure e-2). She underwent urgent pericardiocentesis which drained around 1000 cc of serous fluid, glucose 86, negative acid fast bacilli stain, and no malignant cells were seen. After that, trans-esophageal echocardiography was done for evaluation of the mass. It confirmed the presence of a large rounded shape mass with a maximum diameter: 6.3 X 6.4 cm as seen in the attached picture obstructing the tricuspid valve (Figure e-3). Whole-body computed tomography (CT) angiograph showed a large soft tissue density filling defect almost occupying the right atrium and overriding the cardiac root, indenting the pulmonary veins and almost occluding the distal superior vena cava, with no clear cleavage line on to the adjacent mediastinal structure. It also showed a large anterior mediastinal mass 7 X 4 cm, another heterogenous mass measuring 13 X 13 X 7.5 cm was seen in the right pelvic cavity likely to be originated from the right adnexa (Figure e-4).

Open biopsy was taken from the anterior mediastinal mass and sent for histopathology which revealed DLBCL (Figure e-5). Histopathology showed tumor cells that are diffusely positive for cluster of differentiation (CD) 20. LCA and CD 23 (focal). They are negative for CD 3, CD 30, SALL4, and PLAP immunostain. CD 15 is non-contributory. The morphologic feature and positive CD 23 are suggestive of primary mediastinal DLBCL. We defer further investigations including adnexal mass biopsy, positron emission tomography (PET) scan, and bone marrow as the patient needs cardiac stabilization before proceeding for further investigations.

The patient was started on R-CHOP chemotherapy in an urgent basis. She received the first cycle during hospitalization with a good response evident by improvement of respiratory distress, facial swelling, and upper limb swelling along with a dramatic reduction in the size of cardiac mass and adnexal mass. She received six cycles with echo follow-up between the cycles showed resolution of the cardiac mass. The last cycle of chemotherapy was in May 2019, during which she received 4 doses of intrathecal Methotrexate as prophylaxis for Central Nervous System (CNS) involvement. It was given due to high CNS-international prognostic index score with high LDH (468 U/l), multiple lymph nodes, and ovarian involvement. Follow up imaging showed a residual mass in the mediastinum and pelvis. PET scan showed no evidence of hypermetabolic lymphomatous disease. After that, she received radiotherapy for both the mediastinum and the pelvis, About 40 gray equivalents delivered for both (2 gray with 20 fractions). Currently, the patient is in complete remission, in serial follow-up every 1-2 years. Last follow-up 2 years after diagnosis with PET scan showed no evidence of any hypermetabolic active lymphomatous lesion in the body.

Discussion

This case represents a rare presentation of DLBL, as mass inside the right atrium was the first presentation. Initial clinical picture before CT scan was going with many differential diagnosis. It includes atrial myxoma as our initial thought, rhabdomyosarcoma, atrial thrombus, and lymphoma. Clinical presentations of intracardiac mass are not specific, it includes dyspnea, lower limb swelling, signs of superior vena cava syndrome, intra-cardiac thrombus, peripheral embolization, valvular heart disease, and pericardial effusion (7, 8). Other signs and symptoms suggestive of malignancy like weight loss, night sweats, and fever could be presented. However, it was absent in this case which guided us to think of a benign tumor rather than malignancy.

According to the clinical presentation and initial trans-thoracic echocardiography of pending cardiac tamponade and hemodynamic instability, our first decision was to stabilize the patient by removing pericardial and pleural fluid. Trans-esophageal echocardiography was done for better visualization, extension and attachment of the atrial mass. It's well known that transesophageal echo give better information of intracardiac masses than transthoracic echo (2). However, it failed to reveal the origin of the mass. For which CT was done to detect if there is any outside malignancies invading the heart. CT scan found to have effective modality to demonstrate the extension of the mass and the primary lesion (7). Imaging investigations confirmed primary mediastinal mass invading the heart forming intracardiac mass in the right atrium. Direct extension and infiltration of the mass from the mediastinum to the heart is a well known mechanism (9).

Tissue biopsy was the core element in the diagnosis of this patient, thoracotomy was done previously for this purpose. However, less invasive procedures like percutaneous or fluoroscopy guided biopsy are preferred for pathological examination (7). In our case, there was no accessible lymph nodes were seen by CT scan, pericardial and pleural fluid were negative for malignant cells. As the patient is hemodynamic unstable which was mainly from mass obstructing tricuspid valve. After multidisciplinary team discussion, we decided to go for open heart surgery to stabilize her hemodynamic state by relieving the obstruction and also to get biopsy. Intra-operatively, it was unresectable tumor, and biopsy was taken. Histopathology was consistent with primary mediastinal DLBL.

Cardiac involvement by NHL is rare, and to be presented as intracardiac mass is extremely rare. It could be a primary cardiac lymphoma or mediastinal mass with direct extension and infiltration into the right atrium giving a picture of intracardiac mass (3-6). The prognosis of both subtypes are poor due to rapid progression and invasion of the disease with detection usually in the late stage of the disease (10, 11).

The treatment of DLBL with cardiac involvement is based primarily on chemotherapy. First line treatment regimen with R-CHOP has shown a good response rate (12, 13). Other option was R-EPOCH chemotherapy (14). However, as she was symptomatic and after discussion with the patient and family about the side effects and expected outcome. The decision was to start with R-CHOP chemotherapy. Surgery is usually indicated for a patient who is hemodynamically unstable by improving blood flow to the lungs in case of right ventricle outflow obstruction as chemotherapy takes time to give benefit (15).

Regarding the pelvic mass which was seen by CT scan. Ideally, the biopsy should be taken from the mass. However, as we started the patient on urgent chemotherapy as she was severely symptomatic, we deferred further investigation until follow-up. The mass completely resolved by chemotherapy which confirms histopathology of the mediastinal mass.

DLBCL with heart involvement has a very poor prognosis, it has a 10 % survival rate of 9-12 months if not treated (8). Although it has a high mortality rate, it could be cured with intense chemotherapy (16). Post treatment Surveillance should include PET/CT to confirm response and further follow-up imaging as clinically indicated (17). Early relapse or refractory disease managed by further chemotherapy and allogeneic stem cell transplantation (2). Last guidelines recommended routine follow-up for up to 2 years after treatment. If there is no evidence of relapse, life expectancy is similar to the general population (2).

Conclusions

intracardiac mass is very rare presentation of primary mediastinal DLBCL, it's very important to make early diagnosis and starting treatment as soon as possible as it affects outcomes. Conventional chemotherapy without surgery with good follow up was effective and lead to complete response.

Abbreviations

DLBL: Diffuse large B cell lymphoma NHL: Non-Hodgkin lymphoma (NHL), R-CHOP: Rituximab, Cyclophosphamide, epirubicin, Vincristine, and Prednisone, CT: computed tomography (CT), CD: cluster of differentiation (CD), PET: positron emission tomography (PET).

Declarations

Ethics approval and consent to participate

This is a case report with no identifiable content; despite this, a written consent form was taken from the patient herself for participation and publication

Consent for publication

No images or other personal data that might compromise the anonymity of the patients.

Availability of data and materials:

All data supporting the study are presented in the manuscript or available upon request from the corresponding author of this manuscript.

Competing interest

The authors state that they do not have any conflicting interests.

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Authors' contributions

YA, YI, and SM managed her cardiac condition, pericardiocentesis, trans-thoracic and trans-esophageal echocardiography. They contributed to the diagnosis and management of this patient. RO, a hemato-oncologist who followed up her treatment protocol and put a management plan. All authors participated in the drafting of the manuscript. All authors read and approved the final manuscript.

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Figures

Figure 1

Chest X ray shows bilateral Pleural effusion obscuring heart border

Figure 2

Transthoracic Echocardiography: Transthoracic echocardiography before and after pericardiocentesis, A: showed mass in the right atrium obstructing the tricuspid valve, moderate pericardial effusion, mean gradient:10 mmHg, peak gradient:15 mmHg, RA was not dilated. B: showed the same mass with better resolution after pericardiocentesis.

Figure 3

Trans esophageal Echocardiography: It showed Presence of a large rounded shape mass with maximum diameter: 6.3 X 6.4 cm on the RA obstructing tricuspid valve.

Figure 4

CT scan: showed a large soft tissue density filling defect almost occupying the right heart and overriding the cardiac root, indenting the pulmonary veins and almost occluding the distal SVC as seen in A and B. C: About 13 x 13 x 7.5 cm heterogeneous mass lesion seen in the right pelvic cavity

Figure 5

Histopathology of the mass taken from the mediastinum: Morphologic and immune stains was consistent with primary mediastinal DLBL