Deadly cuddle of diffuse large B-cell lymphoma around the heart: A case report

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SUMMARY

Cardiac involvement in Non-Hodgkin Lymphoma is extremely rare. These rapidly growing high grade tumours are almost exclusively Diffuse Large B-cell lymphoma (DLBCL). This aggressive tumour is unusual among immunocompetent patients. This report describes the clinical case of a 70-year-old female who presented with cardiac tamponade in shock and heart failure symptoms. Investigations confirmed lesion within anterior and left pericardium with minimal mass effect onto the adjacent right lateral ventricle. This case highlights the importance of recognising uncommon presentations of PC-DLBCL. Definite diagnosis can be achieved through multimodality diagnostic imaging and/or pathological study.

INTRODUCTION

Cardiac involvement in Non-Hodgkin Lymphoma is extremely rare. Based on the 2015 WHO Classification of Tumours of the Heart and Pericardium, primary cardiac lymphoma (PCL) represents a minority 1-2% of all primary cardiac tumours.[1] About 80% of these rapidly growing high grade tumours are Diffuse Large B-cell lymphoma (DLBCL). The remaining 8.7-27.2% originate from disseminated lymphoma infiltrating the heart which occurs in advanced disease stage or diagnosed post mortem, with an incidence of 0.05%.[2] This rarity poses significant diagnostic challenges due to nonspecific cardiac symptoms and frequently under-recognised, complicating timely intervention and contributing to poor diagnostic outcomes.2 Right atrium is the most common location of PCL, with the tumour being intrapericardial during diagnosis.3 This aggressive tumour is unusual among immunocompetent patients.[3] Main presentation of PCL with cardiac tamponade only occurs in 20% of the cases.[4]

The authors report this remarkable clinical case due to its rare entity in the absence of immunodeficiency context.

CASE PRESENTATION

Case history and physical examination

A 70-year-old immunocompetent Malay lady with underlying hypertension and dyslipidemia presented to the emergency department of Hospital Taiping for sudden onset chest discomfort, shortness of breath and dizziness. Further history revealed she had a dry cough and was unable to lie flat for a week. She denies any palpitations, fever, loss of appetite or loss of weight. There is no family history of malignancy. She is a teetotaler and non-smoker. Premorbidly, she was an active housewife able to carry out daily activities without restriction, thus her patient performance status on the scale of Eastern Cooperative Oncology Group (ECOG) was zero.

On arrival, her blood pressure was 69/49, pulse rate of 94 , respiratory rate of 38 and oxygen saturation (SPO2) 88% under room air requiring face mask oxygen supplementation and immediately triaged to red zone. After 100 ml of fluid bolus over an hour, blood pressure picked up to 97/67. Physical examination revealed muffled heart sounds without murmur. Lung auscultation was clear and there were no signs of elevated jugular vein pressure, lymphadenopathies, organomegaly, or pedal edema. She was having cardiac tamponade clinically.

Investigation and Diagnosis

Her electrocardiogram was electrical alternans whereas chest X-ray showed water bottle sign of gross globular cardiomegaly. She was admitted to the CCU for pericardiocentesis. Urgent echocardiogram showed pericardial effusion at the measuring 2.09cm anteriorly and 1.76cm posteriorly with right ventricle diastolic collapse and distended inferior vena cava, besides discovery of an apical cardiac mass of size 5cm x 2cm. The ejection fraction was 65%, indicating preserved systolic function and suggesting that the clinical presentation was not due to cardiac failure.

Patient's full blood count, renal profile, thyroid function test, troponin I and tumour markers were within normal range. Troponin I remained within normal limits despite the presence of cardiac tamponade, as there was no evidence of myocardial ischemia. Her autoimmune antibody, Human Immunodeficiency Virus test and hepatitis panel were negative. For pericardial fluid analysis, the laboratory tests concluded as exudative effusion with lactate dehydrogenase level (LDH) of 797 U/L, negative cytology for malignant cells and negative smear for Acid Fast Bacilli. Her serum LDH was 480U/L.

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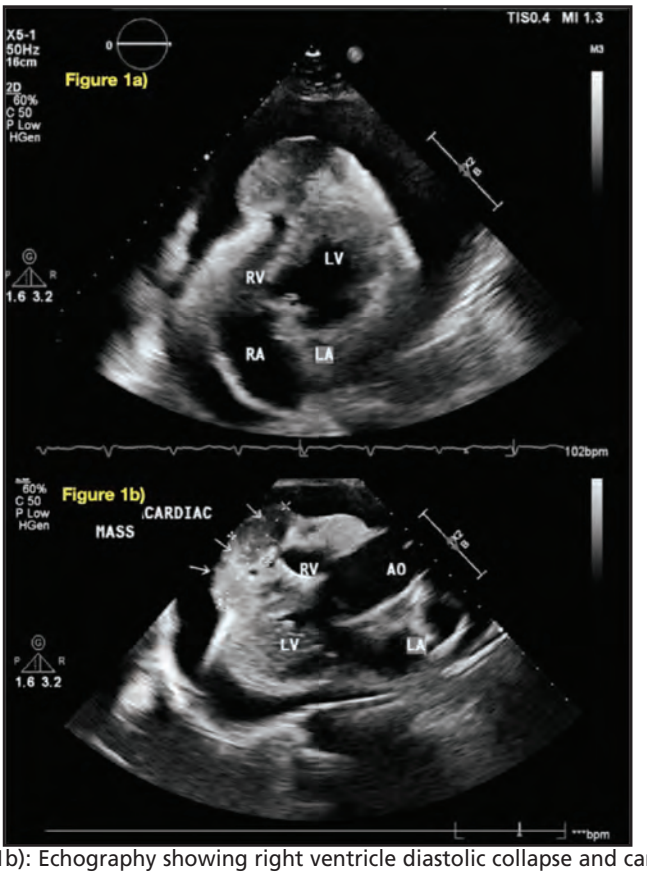

Fig. 1: 1a) and 1b): Echography showing right ventricle diastolic collapse and cardiac mass

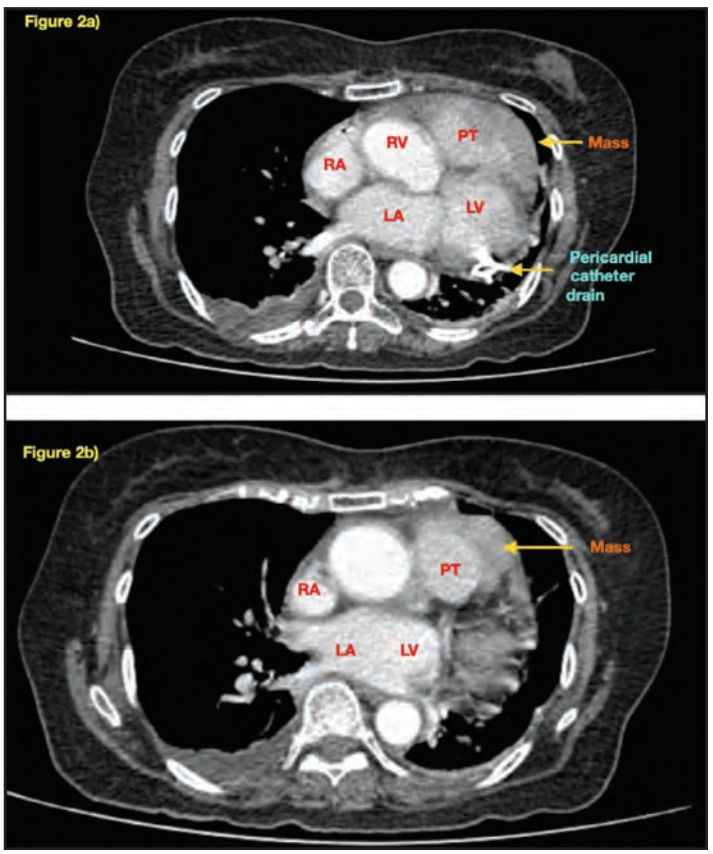

Fig. 2: 2a) and 2b) CECT scan showing cardiac mass (hyperdense crescentic lesion), pericardial catheter drain in situ surrounding pericardial effusion (RA = right atrium, LA= left atrium, RV= right ventricle, LV= left ventricle, PT= pulmonary trunk)

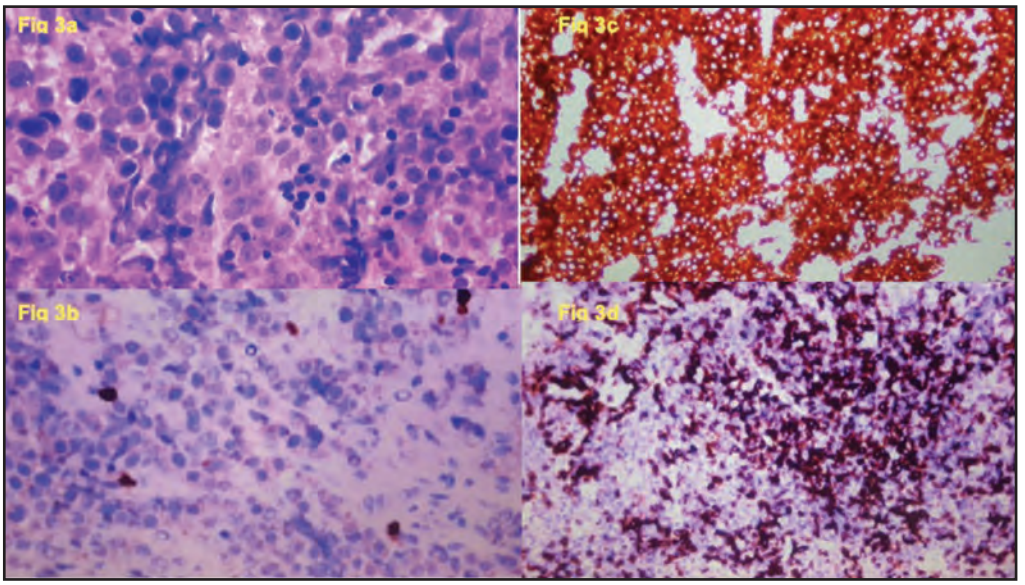

Fig. 3: Immunohistopathology alides from paricardial window biopsy
a) Hematoxylin & Eosin stain: ill defined tumour cells with nucleolar prominence. Moderate to high mitotic activity seen
b) Positive immunohistochemical staining for CD3
c) Positive immunohistochemical staining for CD9a
d) Positive immunohistochemical staining for CD20 Subsequently, the patient was referred to the Haematology team. Chemotheraphy was initiated with Rituximab, Cyclophosphamide, Etoposide, Vincristine, and Prednisolone (CEOP) after staging CT scan. She responded well with resolved pericardial effusion as per repeated echocardiogram post first cycle of R-CEOP

Management and Outcome

Pericardiocentesis drained a total of 1.5L of haemoserous fluid which relieved the patient's failure symptoms drastically and improved hemodynamically. Her blood pressure increased to 140/71 with a pulse rate of 100 post procedure. Pericardial drainage was discontinued after 2 days as there was no further fluid output. CECT showed a hyperdense crescentic lesion within anterior and left pericardium measuring 2.3x6.3x5.0 cm with minimal mass effect onto the adjacent right lateral ventricle; besides mediastinal lymphadenopathies over right lower paratracheal, subcarinal and paraaortic - largest measuring 1.5cm in short axis.

She was referred to the Cardiothoracic (CTC) team for biopsy and discharged well post biopsy. A biopsy via pericardial window over ventricle mass and pericardium presented tumour tissue formed by diffused patternless sheets of malignant medium to large lymphoid cells. The tumour cells possess ill-defined, scanty to moderate amounts of cytoplasm and exhibit hyperchromatic non-cleaved nuclei with nucleolar prominence. Moderate to high mitotic activity and focal tumour necrosis are observed. These cells also expressed bcl 2, CD20, CD79a, LAC and PAX5. 80% of the tumour cells stain positive for c-myc. Ki-67 proliferative index is 70-80%. Stains for ALK, CD3, CD10, CD19, CD21, CD23, CD30, CD56, CK AE1/AE2, Tdt and TTF1 show negative reactivity in the malignant cells.

Combination of her clinical and histopathological features concluded as high grade DLBCL, non-GCB type of intracardiac origin, Ann Arbour stage IVe, International Prognostic Index (IPI) score 3 or National Comprehensive Cancer Network (NCCN) -IPI score of 5. This put her into high-intermediate risk with 51% 5-year progression-free survival and 64% of 5-year overall-survival.

DISCUSSION

This case highlights the importance of recognising uncommon presentations of PC-DLBCL, especially life threatening complications as seen in this patient presenting with cardiac tamponade requiring urgent treatment. The differential diagnosis of a pericardial mass presenting with cardiac tamponade includes malignant metastases, primary cardiac angiosarcoma, and pericardial mesothelioma. Clinical presentation for PCL can be nonspecific, however both DLBCL and PCL are aggressive; with PCL particularly associated with poor outcomes.[5]

Definite diagnosis can be achieved through multimodality diagnostic imaging and/or pathological study (fluid cytology looking for presence of atypical lymphoid cells, immunohistochemistry, flow cytometry, or biopsy). In our case, early echocardiography was a non-invasive simple yet crucial diagnostic tool aiding in diagnosis and relieving the patient's cardiac tamponade in order to stabilise her condition. After that CECT scan confirmed intracardiac mass location - enabling the CTC team to localise the lesion percutaneously for tissue biopsy. Early recognition, accurate multimodal diagnostic imaging, and timely biopsy can significantly influence management decisions and improve patient outcomes. Complication of PC-DLBCL includes heart failure, angina, pericardial effusion, tumour embolisation, direct infiltration/compression of coronary arteries and electrical conduction system.[5,6] The latter manifests as

electrographic (ECG) abnormalities such as atrial arrhythmias and atrioventricular blocks, risking sudden cardiac death.[5]

First-line treatment for PCL is by R-CHOP chemotherapy regime.[4] Myocardial cell necrosis and infiltration by lymphoma cells which lack intercellular cohesion can predispose patients to angina, myocardial dysfunction, arrhythmias, thrombosis or myocardial rupture.[6] Hence, it requires multidisciplinary team monitoring from Cardiology and CTC throughout chemotherapy.

Overall response rate of PC-DLBCL to chemotherapy is 79%. It is reported complete remission of 59% in literature. Radiotherapy can be utilized for cardiac masses refractory to chemotherapy. For patients with coronary stenosis and/or haemodynamic compromise, surgical excision may be used as first line treatment. Despite advances in medicine, patients with PCL have grave prognosis with median survival of 7 months.[7] Although it has a high mortality rate, it can be cured with intense chemotherapy and post chemotherapy surveillance.

CONCLUSION

Isolated cardiac mass presenting as cardiac tamponade is rare in PC-DLBCL. Prompt diagnosis and coordinated multidisciplinary management involving cardiologists, haematologists, and cardiothoracic surgeons are crucial. Chemotherapy tailored by imaging and biopsy findings can achieve favorable outcomes even without surgical intervention, as demonstrated in this case.

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ETHICAL CLEARANCE

Ethical clearance has been obtained from NMRR Secretariat (NMRR ID-24-00408-M3) . Informed consent was obtained from patient/ patient's family members in line with COPE standards for his/her images and other clinical information to be reported in this journal. Due efforts are made to conceal their identity.

DECLARATIONS

The authors have no conflict of interest with respect to the case report.

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