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CASE REPORT
Cardiovascular imaging

Masquerade of an emergency: cardiac tamponade as a deceptive presentation of primary cardiac diffuse large b-cell lymphoma—a case report

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Background Primary cardiac diffuse large B-cell lymphoma (CDLBCL) is an exceptionally rare entity, estimated to represent less than 1% of all primary cardiac tumours. In this case report, we emphasize the diagnostic importance of multimodality imaging and the need for additional procedures, such as tissue biopsy, in a case with a primary cardiac lymphoma presenting with cardiac tamponade.

Case summary An 80-year-old male was admitted to the emergency department with a life-threatening tamponade demanding immediate sternotomy. Pre-operative echocardiography unveiled pericardial effusion and a thickened apex. While computed tomography ruled out an aortic dissection, surgery revealed an unexpected vascular-rich mass at the right ventricle and apex, too perilous for biopsy. Post-operative imaging misinterpreted this mass as a benign haematoma. Subsequently, the patient was admitted to the intensive care unit, but after a conservative treatment strategy, the patient died. An autopsy revealed a primary CDLBCL.

Discussion This case demonstrates the deceptive nature of primary CDLBCL, often complicated by cardiac tamponade. It underscores the pivotal role of pathologic assessment, even amidst the perils of sternotomy, to determine the origin of abnormal cardiac masses. A heightened awareness among physicians is imperative, for such elusive diagnoses may slip by, with potentially fatal outcomes.

Keywords Diffuse large-cell B-cell lymphoma • Cardiac tamponade • Case report • Echocardiography • Cardiac magnetic resonance • Primary cardiac lymphoma • Biopsy

ESC curriculum 2.1 Imaging modalities • 2.2 Echocardiography • 2.3 Cardiac magnetic resonance • 6.8 Cardiac tumours • 7.5 Cardiac surgery

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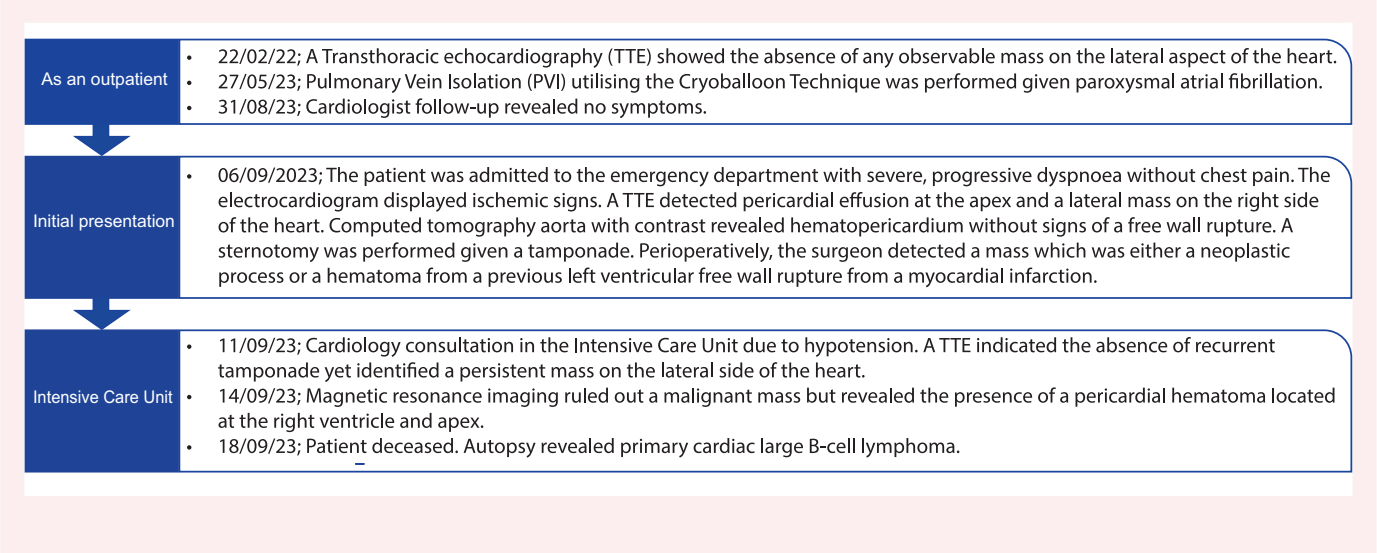
Learning points

- The case highlights the importance of histopathological investigation together with multimodality imaging in the diagnosis of primary cardiac tumours.
- The case is a reminder of the deceptive nature of primary cardiac diffuse large B-cell lymphoma, aiming to raise awareness among physicians regarding primary cardiac tumours when encountering tamponade with the presence of a right ventricular free wall mass.

Introduction

Malignant primary cardiac tumours are exceptionally rare, as indicated by a meta-analysis of 22 studies that reported an autopsy incidence of approximately 0.005%.[1,2] Cardiac lymphomas, which represent <1% of all malignant primary cardiac tumours, are even more uncommon.[3] We report a case of a patient with primary cardiac lymphoma who presented with a life-threatening tamponade which demanded immediate sternotomy. This case report highlights the importance of conducting additional diagnostic procedures, alongside multimodality imaging to procure tissue samples for pathological analysis, in the situation where a vascular-rich mass located in the heart's left ventricle and apex resulted in cardiac tamponade necessitating sternotomy.

Summary figure



Case presentation

An 80-year-old man was admitted to the emergency department with severe, progressive dyspnoea without chest pain. On initial evaluation, blood pressure was 75/54 mmHg, heart rate was 57 beats per minute, and the oxygen saturation was 95% with the use of a non-rebreathing mask, and oxygen flow was 15 L/min. Physical examination revealed muffled heart sounds and cold and pale extremities which displayed early marbling. The jugular veins were not distended. The patient's history included chronic obstructive pulmonary disease and recurrent paroxysmal atrial fibrillation after failed pulmonary vein isolation. The history of medical therapy included the use of an anticoagulant (Xa inhibitor), a beta-blocker, an angiotensin receptor blocker, and a beta2-sympathomimetic, among others. The electrocardiogram (ECG) indicated, sinus bradycardia with ST-elevation in V2–V3 and negative T-segments in II, III, and aVF. The troponin T level was elevated (0.044 µg/L), and lactate levels were 3.0 mmol/L. The initial transthoracic echocardiogram (TTE) which was performed by the emergency physician showed a pericardial mass. The mass raised suspicion of either a covered perforation or a left ventricular free wall rupture (LVFWR) caused by myocardial infarction or aneurysm and warranted further investigation. Subsequent TTE performed by the cardiologist confirmed a tamponade caused by a small rim of pericardial fluid and a solid mass of 4.6 cm. The mass pressured the left and right ventricle and caused concomitant poor left and right ventricular function (*Figure 1*; Supplementary material online, *Video S1*). Subsequent contrast-enhanced aortic computed tomography (CT) ruled out an aortic dissection and an LVFWR, although signs suggested a haematopericardium.

Because pericardiocentesis was deemed unfeasible given the suspected presence of a haematoma, the proposed treatment approach was sternotomy. In the operating room, in total 400 mL of haemorrhagic pericardial fluid was removed. Active bleeding and the origin of the fluid were not detected perioperatively. However, the surgeon detected a mass which was either a haematoma or a neoplasm (*Figure 2*). The haemorrhagic pericardial fluid originated from either another cardiac mass or a post-aneurysmatic LVFWR from a myocardial infarction which was covered by a haematoma. A biopsy was not performed given the high risk of bleeding complications.

One day after admission to the intensive care unit, the patient was extubated. An magnetic resonance imaging (MRI) was performed to assess lesion viability, characterize the cardiac mass, and determine the origin of the pericardial effusion (*Figure 3A* and *B*; Supplementary material online, *Video S2*). The quality of the cardiac MRI was suboptimal because the patient was on mechanical ventilation and was experiencing cardiac arrhythmias. The pericardial mass at the apex did not show enhancement during perfusion or late enhancement. Therefore, the image results were not suggestive of a neoplastic mass but rather with a haematoma. A multidisciplinary team which included

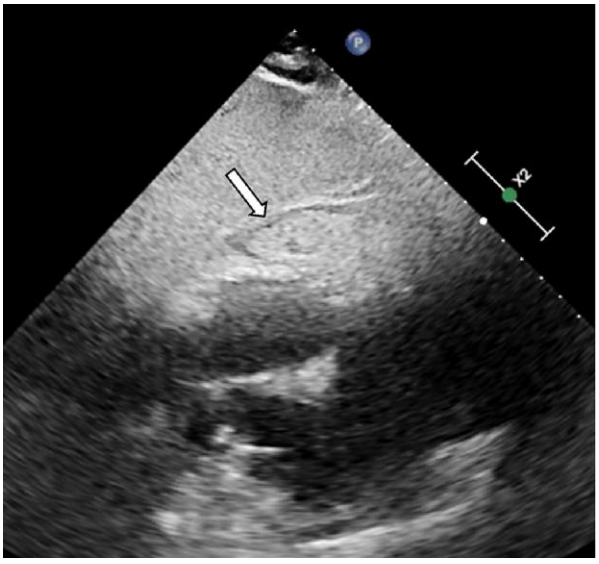


Figure 1 Transthoracic echocardiography image upon admission illustrates a combination of a small rim pericardial effusion which initially measured 7 mm and ultimately contained 400 mL of pericardial fluid and a pericardial mass (arrow) which measured 46 mm × 15 mm in parasternal axis located laterally on the right side of the heart.

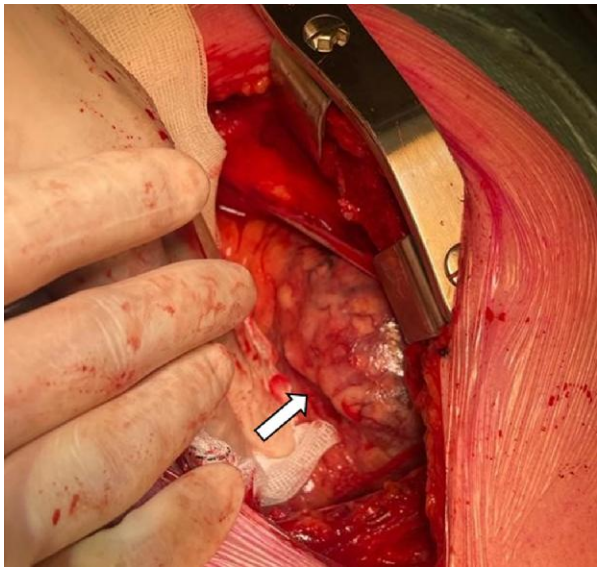


Figure 2 Intraoperative depiction of the pericardial mass (arrow) located on the apex and left side of the heart. The precise location of the mass on the right side of the heart was difficult to ascertain. Given the vascularity of the mass, obtaining a biopsy to prevent a recurrent tamponade would have been challenging.

intensivists, cardiothoracic surgeons, and radiologists determined that complete safe excision of the cardiac anomaly was unfeasible given its integration with myocardial tissue. Further diagnostic evaluation was not discussed. Considering the patient's gradual decline in the intensive care unit, and the lack of available treatment strategies, the patient and family agreed to discontinue further treatment. The patient and family agreed to a post-mortem investigation of the lesion.

Macroscopic investigation of the heart at autopsy revealed enlargement with diffuse epicardial and pericardial adhesions which were caused by a tumour process. The tumour was mainly localized in the epicardium and pericardium, but tumour cells were also found in the myocardium which exhibited nodular configuration and grey-white colouration (*Figure 4A*). Histologic analysis of the tumour lesion showed diffuse sheets of large cells (*Figure 4B*); several large cells were multinucleated, with scant cytoplasm and polymorphic enlarged nuclei. The nuclei exhibited a crude speculated chromatin pattern and one or multiple nucleoli. Additional immunohistochemical analysis indicated that the lesion was typed as a diffuse large B-cell lymphoma (DLBCL). The lesional cells were positive for the B-cell markers CD20 (*Figure 4C*) and Pax5 and demonstrated a nongerminal centre phenotype (BCL2 was diffusely positive, BCL6 and MUM1 were partially positive, and CD10 was negative). The KI67 index was nearly 100%. Epstein-Barr encoding region (EBER) *in situ* hybridization was negative. An additional fluorescence *in situ* hybridization (FISH) analysis to detect a cMYC translocation was performed; however, no translocation was present. No other nodal or extra-nodal localization of a DLBCL was observed. The lymphoma was classified as a primary cardiac DLBCL.

Discussion

Primary CDLBCL, which represents less than 1% of all primary cardiac tumours, is exceptionally rare. Our patient was presented with a life-threatening complication—cardiac tamponade—which necessitated sternotomy for the removal of haemorrhagic pericardial fluid. In this case, the CDLBCL was wrongly diagnosed as a concealed perforation (haematoma) which was potentially linked to post-aneurysmatic LVFWR from a myocardial infarction. Imaging modalities failed to provide a precise diagnosis, and histopathological material was not obtained. Therefore, raising awareness among physicians of the need for a broad differential diagnosis and appropriate additional diagnostic modalities in the management of rare cardiac masses is crucial.

The importance of utilizing multiple imaging modalities, such as TTE, multidetector row CT, MRI, and ^{18}F-FDG PET-CT is acknowledged; these modalities are crucial in the diagnosis of cardiac tumours.[4] Specifically, MRI exhibits high specificity and sensitivity in differentiating between benign and malignant cardiac masses.[5] Distinct morphological features such as right-sided location, invasive growth patterns, and the presence of pericardial effusion have emerged as single indicators of malignancy.[6] Nevertheless, the differentiation between primary cardiac lymphoma, secondary neoplasms, and other cardiac masses such as myxomas, angiosarcomas, or concealed perforations (haematoma) requires histopathology. In our case, the presence of ECG abnormalities together with pericardial fluid and a mass on the heart suggested a covered perforation after an LVFWR. The incidence of LVFWR after acute myocardial infarction ranges between 0.01% and 0.5%, and LVFWR is associated with mortality rates between 39% and 92%.[7,8] Given the variable clinical presentations and high mortality, LVFWR remains a diagnostic challenge for clinicians.[8] Although obtaining histopathological material is not desirable in the presence of a haematoma after post-aneurysmatic LVFWR from a myocardial infarction, it is crucial for an accurate diagnosis and the initiation of appropriate treatment in patients with DLBCL. Obtaining intraoperative excisional biopsies can be challenging given the risk of bleeding associated with their proximity to a coronary artery or to well-vascularized tumours which also may resemble a haematomas after LVFWR.

Upon the intraoperative acquisition of histopathology, an early diagnosis which allowed for the initiation of treatment in our patient could have been established. The primary management approach of DLBCL involves systematic therapy. Among the 70% of patients who present

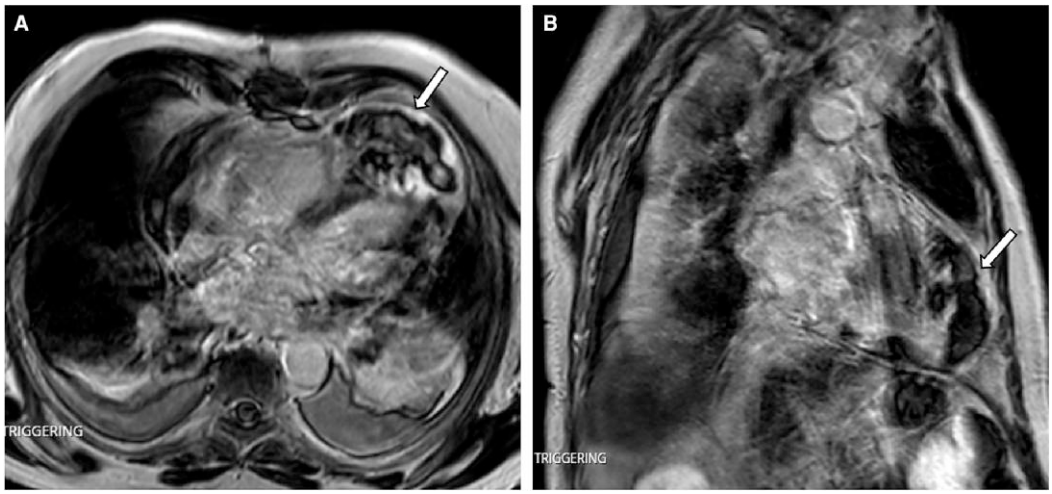


Figure 3 Cardiac magnetic imaging was performed one week postoperatively. The mass did not show enhancement in perfusion or late enhancement, indicating characteristics which are consistent with haematoma formation. (*A*) In the four-chamber view, the mass (arrow) was located on the lateral wall of the right ventricle and the apex. (*B*) In the sagittal view, the mass (arrow) extended from the lateral side of the right atrium to the apex; its dimensions measured 84 mm × 24 mm.

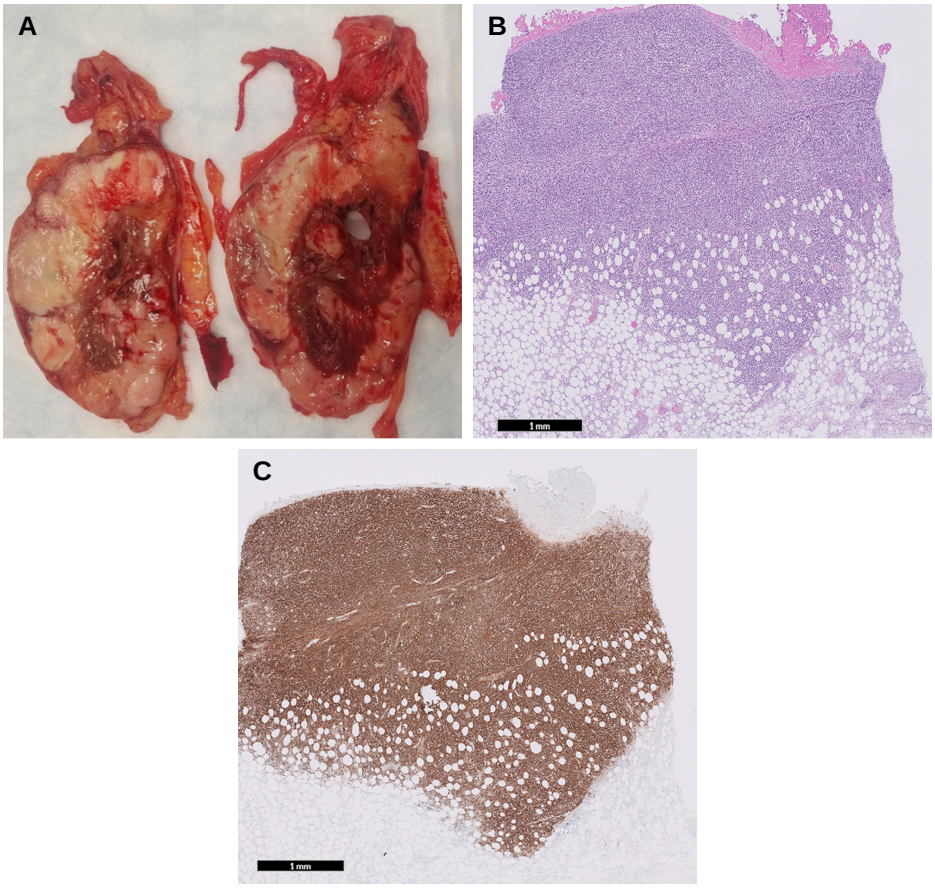


Figure 4 Macroscopic and microscopic pathologic features. (*A*) Slices of the heart at obduction showed white nodular lesions in the epicardium and myocardium. (*B*) A haematoxylin and eosin stain of a section of the white nodular lesion in the epicardium demonstrated a cell-rich lesion. (*C*) An immunohistochemical stain for CD20 of the same area as in B demonstrated a B-lymphocyte lineage.

with advanced-stage disease, over 60% have shown partial or complete remission with R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone).[9] Our patient exhibited the B-cell markers CD20 and Pax5 B-cell markers, a BCL2-positive phenotype, and the absence of a cMYC translocation and would likely have had a relatively better prognosis with R-CHOP.[10,11] The median age at treatment initiation for patients with DLBCL is in the mid-60 s, and 30% are older than 75 years of age.[9] Nevertheless, the prognosis with chemotherapy remained poor for our 80-year-old patient; the median survival for this patient was 7 months.[9]

This case highlights the importance of obtaining a tissue biopsy and using multimodality imaging in the diagnosis of primary cardiac tumours. The case further underscores the need for physicians to consider the presence of primary cardiac tumours. The diagnostic process can be challenging. To prevent the fatal consequences of CDLBCL, obtaining histopathological material is crucial for the diagnosis. In this case, the absence of pathological assessment impeded the initiation of appropriate treatment in our patient.

We conclude this case report and discussion with a few key learning points:

- The case highlights the importance of histopathological investigation together with multimodality imaging in the diagnosis of primary cardiac tumours.
- Awareness of primary cardiac tumours must be raised among physicians when tamponade in the presence of a right ventricular free wall mass is encountered.
- The case is a reminder of the deceptive nature of primary cardiac diffuse large B-cell lymphoma.
- Imaging modalities (especially MRI) must be better correlated to histopathological findings to improve their diagnostic yield.

Lead author biography

Tijn Heeringa is currently a third-year student in the medical MSc program, a second-year student in the epidemiology MSc program and is in the first year of a PhD program at the University Medical Center Utrecht. He has a special interest in Cardiothoracic Surgery and Cardiology.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

Consent: The authors confirm that written consent for submission and publication of this case report including the images and associated text have been obtained from the patient's family in line with COPE guidance.

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Data availability

The data underlying this article will be shared on reasonable request by the corresponding author.

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