

# Primary cardiac diffuse large B-cell lymphoma presenting with cardiac tamponade: a case report

Laurens Berton <sup>1\*</sup>, Valerie Van Ballaer <sup>2</sup>, Olivier Ghekiere<sup>2,3</sup>,  
and Eva De Caluwé <sup>4</sup>

<sup>1</sup>Department of Cardiology, Sint-Trudo Hospital, Diestersteenweg 100, 3800 Sint-Truiden, Belgium; <sup>2</sup>Department of Radiology, Jessa Hospital, 3500 Hasselt, Belgium; <sup>3</sup>Faculty of Medicine and Life Sciences, Cardiology and Organ systems, Hasselt University, 3500 Hasselt, Belgium; and <sup>4</sup>Department of Cardiology, Sint-Trudo Hospital, 3800 Sint-Truiden, Belgium

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## Background

Primary cardiac tumours are extremely rare with an autopsy incidence of 0.05%. They can present with a variety of symptoms, including life-threatening arrhythmia and cardiac tamponade. In this case report, we focus on the diagnostic process and management of a primary cardiac lymphoma (PCL) presenting with cardiac tamponade.

## Case summary

We report on a 71-year-old male presenting with a large pericardial effusion, tamponade, and a mass in the right atrioventricular groove. Multimodality imaging was performed, including transthoracic echocardiography, computed tomography, magnetic resonance imaging, positron emission tomography, and computed tomography-guided transthoracic biopsy. The final diagnosis of a double-hit diffuse large-cell B-cell lymphoma was made, for which treatment consisting of a combination of chemotherapy and immunotherapy was initiated. Low-dose colchicine was also added to the treatment.

## Discussion

Primary cardiac lymphoma remains a very rare diagnosis and this case highlights the need for multimodality imaging and imaging-guided biopsy to differentiate cardiac masses. First-line treatment for PCL remains a combination of chemotherapy with immunotherapy, with the addition of low-dose colchicine to prevent recurrence of malignant pericardial effusion.

## Keywords

Malignant pericardial effusion • Cardiac tamponade • Primary cardiac lymphoma • Diffuse large-cell B-cell non-Hodgkin lymphoma • Case report

## ESC Curriculum

2.1 Imaging modalities • 2.2 Echocardiography • 2.3 Cardiac magnetic resonance • 2.4 Cardiac computed tomography • 6.6 Pericardial disease

## Learning points

- Primary cardiac tumours are rare but can present with life-threatening complications such as tamponade.
- Detailed review of the transthoracic echocardiogram is essential to identify any cause of effusion in patients presenting with cardiac tamponade.
- Diagnosis of primary cardiac tumours and concurrent pericardial effusion heavily relies on multimodality imaging and (excisional) biopsy.

\* Corresponding author. Tel: +32 477 927 148, Email: [laurens.berton@gmail.com](mailto:laurens.berton@gmail.com)

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## Introduction

Primary cardiac tumours are extremely rare (0.05% in a case series of 12 000 autopsies) and cardiac lymphomas only represent 1% of these. They are even more rare in immunocompetent patients and present with tamponade in only 20% of cases.<sup>1,2</sup> In this case report, we focus on the diagnostic process and investigations in a rare case of primary cardiac lymphoma (PCL) with pericardial effusion.

## Timeline

20-03-2021	Emergency department presentation. Computed tomography pulmonary angiography shows pericardial effusion. Transthoracic echocardiography confirms pericardial effusion with tamponade and a mass in the right atrioventricular groove. Urgent pericardiocentesis and start of colchicine.
26-03-2021	Magnetic resonance imaging shows an infiltrating mass surrounding the right coronary artery.
01-04-2021	<sup>18</sup> F-fluorodeoxyglucose positron emission tomography shows pathological tracer uptake of the mass with two enlarged lymph nodes surrounding the aortic arch. No other foci.
07-04-2021	Computed tomography-guided biopsy: double-hit large-cell B-cell non-Hodgkin lymphoma.
10-05-2021	Start treatment with combination of chemotherapy and immunotherapy.
30-07-2021	Completion of three cycles of therapy with complete metabolic remission.

## Case presentation

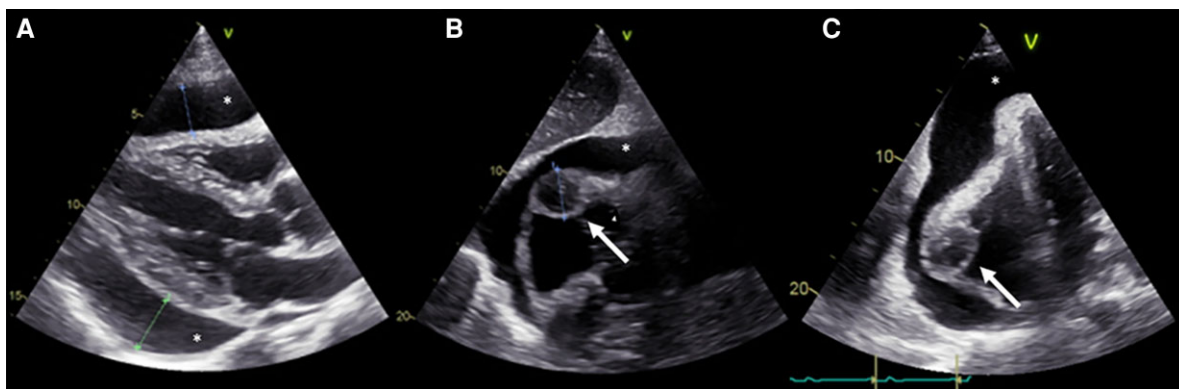
A 71-year-old male patient was referred to the emergency department by his general practitioner because of dyspnoea, thoracic

pain, and elevated D-dimers. On initial evaluation, blood pressure was 135/92 mmHg, heart rate was 102 beats per minute, and oxygen saturation was 97%. Physical examination revealed elevated jugular venous pressure, indicative of raised central venous pressure, and muffled heart sounds. His medical history included recurrent prostatitis and recent Guillain-Barré syndrome without any previously reported cardiac antecedents. He was not taking any medication.

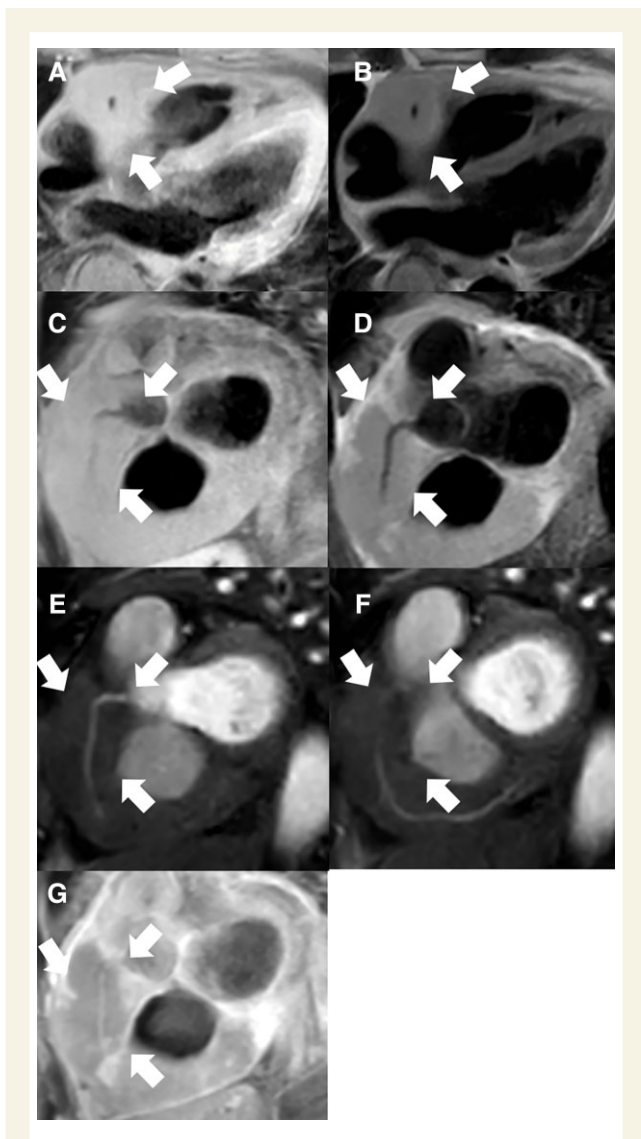
An electrocardiogram revealed a sinus tachycardia with low voltages. Due to the elevated D-dimers in combination with distended jugular veins, a computed tomography pulmonary angiography was performed. Computed tomography pulmonary angiography excluded pulmonary embolism but did show a large pericardial effusion. Urgent transthoracic echocardiography (TTE) confirmed a pericardial effusion of 3 cm (*Figure 1A*) with partial collapse of the right atrium and right ventricle, compatible with tamponade. Transthoracic echocardiography also showed a right-sided cardiac mass of 36 × 23 mm located in the atrioventricular groove (*Figure 1B and C*), which needed further investigation. An urgent pericardiocentesis was performed with the removal of 1200 mL of haemorrhagic pericardial fluid. Histopathologic examination of the pericardial fluid showed atypical lymphocytes, not otherwise specified.

To further differentiate the cause of this pericardial effusion and to characterize the cardiac mass, magnetic resonance imaging (MRI) of the heart was performed. This confirmed a large sharply outlined infiltrating mass surrounding the right coronary artery (RCA), with the extension of the mass from the proximal to the distal segment of the RCA (arrows). The mass was iso- to hyperintense on T<sub>1</sub>-weighted spectral presaturation with inversion recovery (*Figure 2A and C*) and T<sub>2</sub>-weighted turbo spin-echo (*Figure 2B*) sequences. Short-TI inversion recovery sequence (*Figure 2D*) showed a slightly hyperintense mass when compared with the normal myocardium.

Short axis rest perfusion shows no early contrast enhancement of the mass surrounding the RCA (*Figure 2E and F*), while inhomogeneous contrast enhancement can be observed on T<sub>1</sub>-weighted images with fat suppression after intravenous administration of gadolinium (*Figure 2G*, acquisitions obtained 10 min after administration of



**Figure 1** Transthoracic echocardiography on admission illustrating diffuse pericardial effusion (\*) with a right-sided cardiac mass in the atrioventricular groove (arrow) [parasternal (A), subcostal (B), and apical (C) approach].



**Figure 2** Cardiac magnetic resonance imaging work-up of a primary cardiac diffuse large B-cell lymphoma surrounding the right coronary artery. Four chamber imaging shows a mass located in the right atrioventricular groove and surrounding the right coronary artery, on iso- to hyperintense on T<sub>1</sub>-weighted spectral presaturation with inversion recovery (Figure 2A) and T<sub>2</sub>-weighted turbo spin-echo (Figure 2B) sequences. Short axis imaging shows the extension of the mass from the proximal to the distal segment of the right coronary artery (arrows), on the T<sub>1</sub>-weighted spectral presaturation with inversion recovery sequence (Figure 2C) and short-TI inversion recovery (Figure 2D) sequences. Short axis rest perfusion shows no early contrast enhancement of the mass surrounding the right coronary artery (Figure 2E and F), while inhomogeneous contrast enhancement can be observed on T<sub>1</sub>-weighted images with fat suppression 10 min after intravenous administration of gadolinium (Figure 2G).

gadolinium). Cardiac MRI also showed normal ventricular and valvular function (Figure 3).

Further staging by <sup>18</sup>F-fluorodeoxyglucose positron emission tomography-computed tomography (<sup>18</sup>F-FDG PET-CT) revealed



**Figure 3** CT coronary angiography (curved MPR reconstructions) shows complete patency of the right coronary artery without luminal stenosis or external compression by the mass.

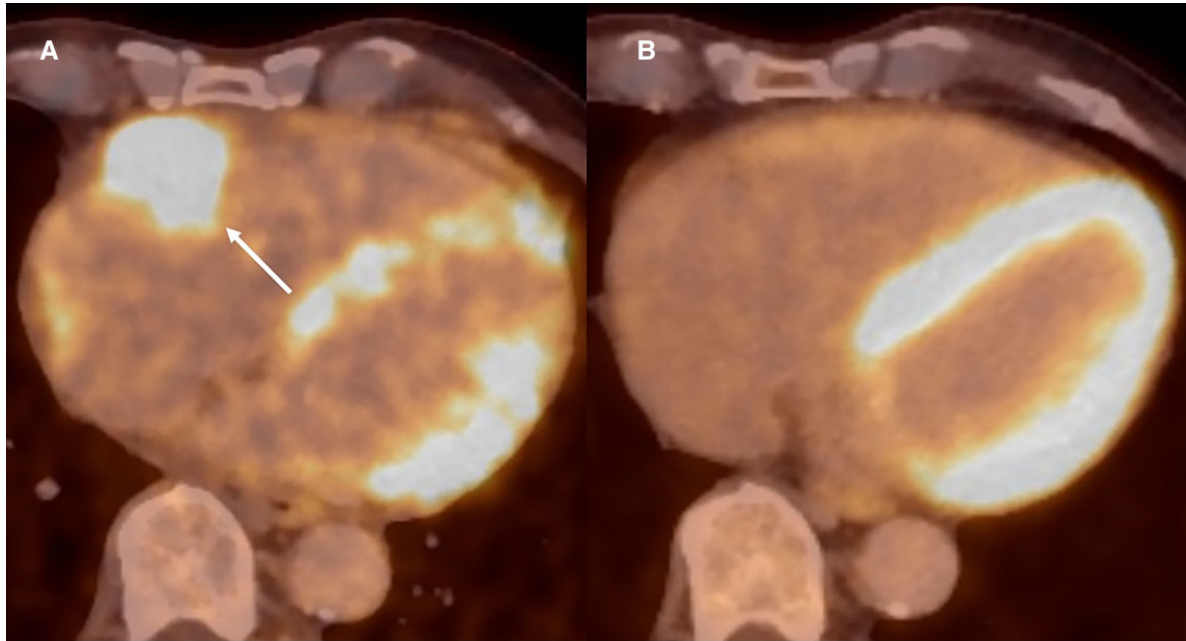
pathological tracer uptake of the mass (Figure 4A) and two enlarged lymph nodes surrounding the aortic arch. No extranodal involvement was seen.

Based on multimodality imaging, the main differential diagnoses of the mass included IgG4-related disease and cardiac lymphoma, both of which can present with a (pseudo-)tumoural formation and both of which are histopathological diagnoses. Consequently, a computed tomography-guided transthoracic biopsy of the cardiac mass was performed. Histopathology was compatible with a diffuse large B-cell lymphoma (DLBCL). Further cytogenetics showed both C-MYC and BCL-6 translocation and a final diagnosis of a double-hit DLBCL with primary cardiac involvement was made.

The patient was treated with rituximab, cyclophosphamide, hydroxydaunorubicin, vincristine, prednisone (R-CHOP) every 3 weeks combined with high dose methotrexate to prevent spreading to the central nervous system. After initial pericardiocentesis, the symptoms of dyspnoea quickly resolved. Treatment with 0.5 mg colchicine twice a day (1 mg daily) was initiated to prevent the recurrence of pericardial fluid and tamponade. After three cycles, the patient was asymptomatic, and <sup>18</sup>F-FDG PET-CT showed a complete metabolic response (Figure 4B) with complete regression of the mass. Transthoracic echocardiography showed full resolution of the pericardial fluid and a stable left ventricular ejection fraction of 58%.

## Discussion

Primary cardiac tumours are extremely rare (0.05% in a case series of 12 000 autopsies), and cardiac lymphomas only represent a minority of these (1%). Primary cardiac lymphomas are defined as lymphomas only involving the heart and the pericardium.<sup>3</sup> This case illustrates that these patients can present with life-threatening complications, such as cardiac tamponade, for which urgent treatment is required.<sup>1,2</sup>



**Figure 4**  $^{18}\text{F}$ -fluorodeoxyglucose positron emission tomography–computed tomography in the axial plane shows pathological tracer uptake of the mass (arrow in *Figure 4A*), with complete metabolic response after three cycles of rituximab, cyclophosphamide, hydroxydaunorubicin, vincristine, and prednisone (*Figure 4B*).

It highlights the importance of multimodality imaging in the diagnostic process.

Primary cardiac lymphomas are most common in immunocompromised patients and are frequently aggressive, needing urgent treatment to prevent death. They must be differentiated from more common cardiac masses such as myxomas and angiosarcomas. Primary cardiac lymphomas can be anaplastic, plasmablastic, and T-cell but most often they are a B-cell. Cytogenetic characteristics of PCL are not well known.<sup>4</sup>

In a case series of PCLs, only 20% of patients had pericardial tamponade at first presentation. The most common presenting symptoms include dyspnoea and arrhythmia.<sup>4</sup> Guillain-Barré syndrome is very rarely associated with non-Hodgkin lymphoma.<sup>5</sup>

Multimodality imaging is crucial to characterize these lesions. Most often, TTE, CT, MRI, and PET-CT are used for diagnosis and staging. In a case series of five patients with DLBCL, the combination of a right-sided cardiac mass, a large pericardial effusion, and no apparent stenosis of the encased coronary artery were found to be very specific for DLBCL.<sup>6</sup> Histopathological diagnosis is ideally made on an excisional biopsy, but this is not always feasible because of localization close to the coronary artery with high risk of bleeding complication.<sup>7</sup>

More than 60% of people with a DLBCL can be cured by using R-CHOP. Double-hit DLBCLs, however, are associated with poorer outcomes.<sup>8</sup> Surgery can be used as a bail-out strategy or as a first-line treatment when haemodynamic compromise and/or coronary stenosis are present. Despite chemotherapy and/or surgery, the prognosis of patients with PCL remains poor with a median survival of 7 months.<sup>4</sup>

Colchicine was already an established part of treatment to prevent the recurrence of pericardial effusion in non-malignant causes of

pericardial fluid and pericarditis, but it is also useful in cases of malignant pericardial effusion with tamponade.<sup>9</sup>

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

- Primary cardiac tumours are rare but can present with life-threatening complications such as tamponade.
- Detailed review of the transthoracic echocardiogram is essential to identify any cause of effusion in patients presenting with cardiac tamponade.
- Diagnosis of primary cardiac tumours and concurrent pericardial effusion heavily relies on multimodality imaging and (excisional) biopsy.
- Low-dose colchicine remains standard of care in non-malignant causes of pericardial effusion and pericarditis, but it also appears to be safe and effective in malignant causes of pericardial effusion with tamponade.

## Lead author biography



Laurens Berton underwent his Medical Studies at the University Hospitals of Leuven. He completed his second year of general internal medicine at the Sint-Trudo hospital and is currently a third-year resident of general internal medicine at the University Hospitals of Leuven and has a special interest in Cardiology.

**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

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

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