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CASE REPORT
Cardiac imaging

A case report of a primary cardiac lymphoma causing superior vena cava obstruction: the value of multimodality imaging in the clinical workup

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Background This is a case report of a primary cardiac lymphoma with an unusual clinical presentation. We hereby illustrate the characteristic features of cardiac lymphomas by multimodality imaging and particularly cardiac magnetic resonance (CMR) that can help reach a timely diagnosis non-invasively and guide treatment decisions.

Case summary A 58-year-old woman, without significant past medical history, presented with a 3-week history of shortness of breath associated with facial and neck swelling. Transthoracic echocardiogram confirmed the presence of a cardiac mass in the right atrium. Cardiac magnetic resonance helped to characterize the mass, assess its haemodynamic significance and relation to cardiac structures, and reach a non-invasive diagnosis that was crucial for guiding treatment decisions and interventions.

Discussion Cardiac masses have distinct imaging features that can help differentiate malignant from benign cardiac tumours. More specifically, primary cardiac lymphomas can be relatively easy diagnosed by CMR in most cases thanks to their characteristic imaging appearance.

Keywords Case report • Cardiac lymphoma • Cardiac magnetic resonance • Multimodality imaging

Learning points

- Primary cardiac lymphoma are rare, typical sizable tumours that usually involve the right heart chambers.
- Cardiac magnetic resonance has an important role in delineating mass characterization, relation to cardiac structure, and haemodynamic significance.
- Multimodality imaging can be used to reach a non-invasive diagnosis in most cases of cardiac lymphomas and guide treatment decisions and interventions.

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Introduction

Primary cardiac lymphomas are rare cardiac sizable masses that typically involve the right ventricular chambers. If left untreated cardiac lymphomas may have a poor prognosis and cause haemodynamic complications and conduction abnormalities. We hereby present a rare case of primary cardiac lymphoma with an unusual presentation. We highlight the role of multimodality imaging and mainly cardiac magnetic resonance (CMR) in reaching an early non-invasive diagnosis and guiding timely therapeutic management.

Timeline

Day 1	Emergency department admission with shortness of breath, increased face and neck swelling for 3 weeks. Computed tomography demonstrates an irregular mass in the right atrium with possible superior vena cava obstruction.
Day 7	Cardiac magnetic resonance and positron emission tomography (PET) show a large metabolically active irregular cardiac mass with features suggestive of cardiac lymphoma.
Day 15	Biopsy of myocardial mass. Histology diagnostic for diffuse large B-cell lymphoma of probable non-germinal centre phenotype (MUM1+).
Day 21	Complete heart block. Permanent pacemaker implanted. Chemotherapy initiation.
6 months	Completed 1× RCVP (rituximab, cyclophosphamide, vincristine sulfate, and prednisone) and 6× R-CHOP (rituximab, cyclophosphamide, doxorubicin hydrochloride, vincristine and prednisolone) chemotherapy.
7 months	Computed tomography PET shows a complete metabolic response.

Case presentation

A 58-year-old woman, without significant past medical history, presented with a 3-week history of shortness of breath associated with facial and neck swelling. On examination, she was found to be haemodynamically stable with distended neck veins, flushed face and blue tinge to lips. Laboratory studies revealed elevated C-reactive protein (69 mg/L; normal range 0–10) and cardiac troponin (150 ng/L; normal range <11.6) levels. Chest X-ray showed cardiomegaly with an otherwise normal heart and mediastinal contour (*Figure 1A, Videos 1* and *2*).

Computed tomography (CT) pulmonary angiogram ruled out pulmonary embolism but demonstrated an irregular mass in the right atrium with a moderate pericardial effusion (*Figure 1B*). Whole-body CT did not detect a primary lesion elsewhere.

Transthoracic echocardiogram showed a large echogenic mass in the right atrium with evidence of vascularity on contrast echo, protruding into the right ventricular cavity, causing mild tricuspid inflow obstruction, and superior vena cava (SVC) obstruction (*Figure 1C*). A large global pericardial effusion with signs of mild haemodynamic compromise and a large left pleural effusion were also seen.

Cardiac magnetic resonance imaging was performed for further assessment and for tissue characterization. A sinister-looking ill-defined soft-tissue mass was seen infiltrating the right atrial free wall and extending into the atrioventricular groove, infiltrating the right ventricular anterior and inferior walls. The left ventricular inferior wall was also infiltrated by the mass. Cardiac magnetic resonance velocity mapping demonstrated caudal flow in the azygos vein, suggestive of SVC obstruction (*Figure 1D* and *E*). The mass was encasing the aortic root, the right coronary artery[1] and the main pulmonary artery without invasion or compression (*Figure 1F* and *G*). There was a lobulated extension of the mass into the right atrial cavity with limited extension to the suprahepatic inferior vena cava. The pulmonary veins appeared patent. There was possible involvement of the base of the anterior tricuspid valve leaflet, but no inflow obstruction. The mass was isointense on T1 weighted images and mildly hyperintense on T2 weighted images (*Figure 1H*). There was mild enhancement of the mass during the first pass perfusion and heterogeneous enhancement on the early and late phase post gadolinium images (*Figure 1I*). Anatomical location and tissue characteristics by magnetic resonance imaging were highly suggestive for malignancy. The CMR features were suggestive of lymphoma rather than any other malignant tissue, but tissue biopsy was recommended for confirmation.

Cardiac positron emission tomography (PET) imaging with ^{18}F-fluorodeoxyglucose (FDG) after proper dietary preparation (prolonged fast and a no carbohydrate diet) was performed to determine if the mass was indeed malignant. The PET imaging features were in keeping with a large intensely metabolically active irregular mass in the right side of the heart with further extension into the aortic wall, main pulmonary trunk and left ventricular pericardium (*Figure 1J*). There were also metabolically active lesions in the spleen, possibly kidneys, thoracic and abdominal lymph nodes, likely to be due to distant metastases.

Endomyocardial biopsy of the right ventricle was performed. Fragments of myocardium with sheet-like proliferation of small to moderately large malignant cells with large nuclei and minimal cytoplasm were detected. Numerous mitoses and focal necrosis were also noted. Histology was diagnostic for diffuse large B-cell lymphoma (DLBCL) of probable non-germinal centre phenotype (MUM1+) corresponding to a stage IIIE DLBCL. The clinical course was complicated by complete heart block requiring urgent permanent pacemaker implantation. Six cycles of R-CHOP (rituximab, cyclophosphamide, doxorubicin hydrochloride, vincristine and prednisolone) chemotherapy in addition to previous R-CVP (rituximab, cyclophosphamide, vincristine sulfate, and prednisone) treatment were completed without complications. A complete metabolic response with no residual disease was documented by a PET/CT at 6 months after the initial diagnosis (*Figure 1K*).

Discussion

Primary cardiac lymphomas are rare, sizable cardiac tumours,[2–4] which can lead to mechanical complications and usually have poor

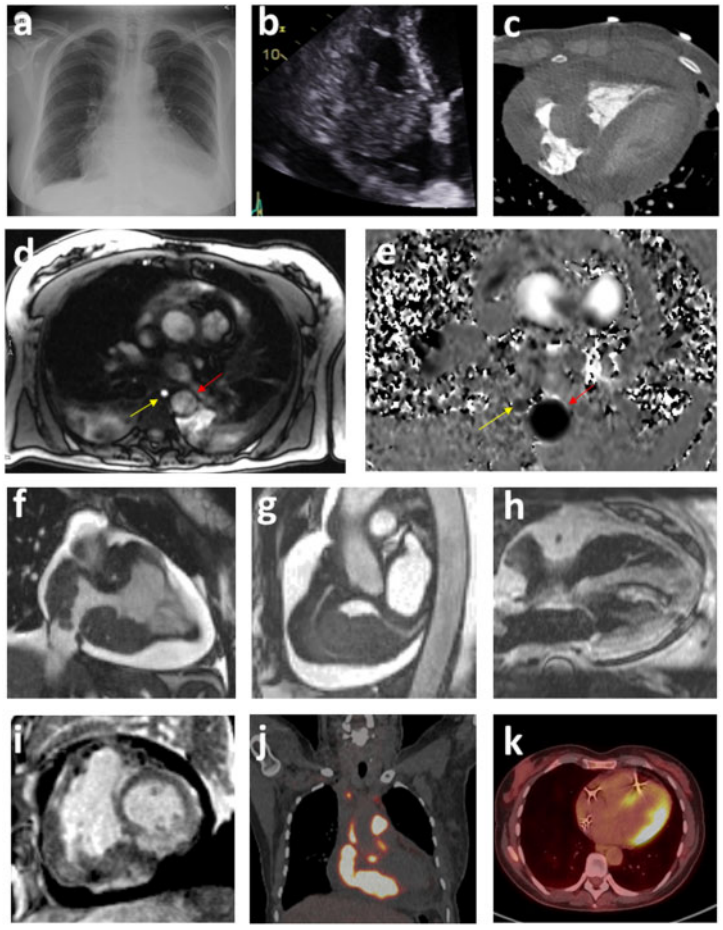
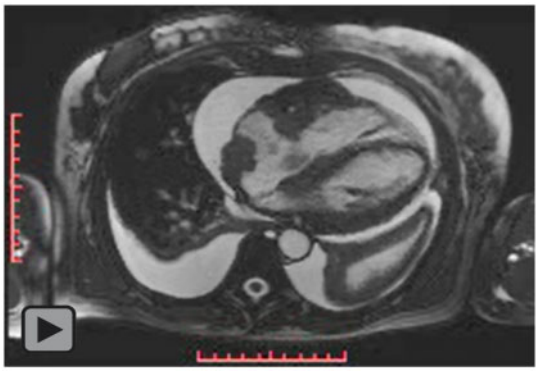
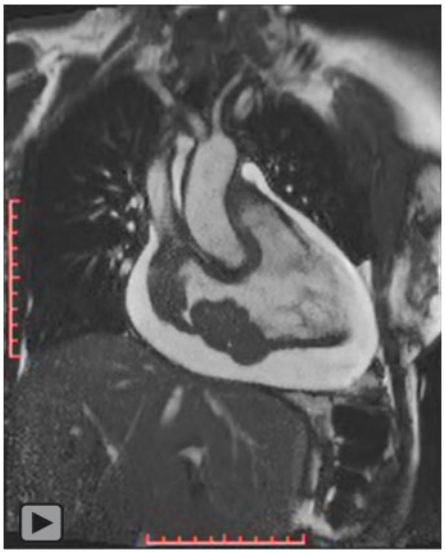


Figure 1 Multimodality imaging of a primary cardiac lymphoma. (*A*) Chest X-ray. (*B*) Transthoracic echocardiography—right ventricle view. (*C*) Contrast-enhanced computed tomography scan of the mass; cardiac magnetic resonance flow mapping. (*D* and *E*) Magnitude and velocity encoded image showing great vessel anatomy and a caudal flow in the azygos vein (yellow arrow) similar to the descending aorta flow (red arrow) suggestive of superior vena cava obstruction. (*F*) Cardiac magnetic resonance steady-state free precession imaging of the mass. (*G*) Steady-state free precession view of the mass and coronary artery encasement. (*H*) High signal of the mass on T2-short tau inversion recovery by cardiac magnetic resonance. (*I*) Late gadolinium enhancement imaging of the mass. (*J*) Highly increased ^{18}F-fluorodeoxyglucose radiotracer uptake by the mass with metabolic tissue volume by positron emission tomography/computed tomography. (*K*) Follow-up positron emission tomography/computed tomography after six cycles of chemotherapy showing complete remission of the tumour (see also text for details).



Video 1 Cardiac magnetic resonance (CMR) transaxial view steady-state free precession (SSFP) cine showing the infiltrating mass and pericardial effusion.

prognosis if left untreated. In this case, the clinical course was complicated by complete heart block. Multimodality imaging of cardiac masses can help to identify features that are suggestive of malignancy and guide prompt patient management.[5,6] A summary of these features is provided in *Table 1*. Combined assessment by transthoracic echocardiography and CMR can be of help in most cases of cardiac masses. Further valuable information can be provided by CT or ^{18}F-FDG PET/CT imaging; high radiotracer uptake by the mass is highly suggestive of a malignant cardiac tumour.[5] A stepwise multimodality imaging approach and judicious use of available resources can help reach a non-invasive diagnosis in most cases, and differentiate malignant tumours from benign cardiac masses or intracardiac thrombi.



Video 2 Cardiac magnetic resonance (CMR) right ventricle in & out view steady-state free precession (SSFP) cine showing the infiltrating mass and pericardial effusion.

Lead author biography

Dr Alessia Azzu is a Cardiologist with a special interest in Cardiovascular Imaging. After graduating from the University of Sassari and finishing her specialty training in Cardiology in Italy, she completed post-CCT Fellowships in Periprocedural Echocardiography and Cardiac Magnetic Resonance Imaging at the Royal Brompton Hospital in London. She is now a Clinical Research Fellow at the National Heart and Lung Institute, Imperial College London, UK.

Supplementary material

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

Table 1 Imaging features suggestive of a malignant cardiac mass and cardiac lymphoma

	Malignant cardiac mass	Primary cardiac lymphoma
General imaging features	Direct invasion	Common involvement of RA, RV, and AV groove frequent involvement of epicardial surface
	Irregular borders	
	Large size, multiple lesions	Usually large sized mass
	Pericardial involvement	Pericardial effusion
		Encasement of large vessels and coronary arteries without obstruction
Echocardiography	Tissue perfusion (contrast)	Limited tissue perfusion
	Haemodynamic impact	
	Valvular abnormalities	
CMR	Tissue heterogeneity	Tissue heterogeneity
	T1w-TSE: variable intensity[a]	T1w-TSE: isointense/hypointense[a]
	T2w-TSE: isointense/hyperintense[a]	T2w-TSE: hyperintense[a]
	T2 STIR: isointense/hyperintense[a]	T2 STIR: hyperintense[a]
	Fat suppression: isointense/hypointense	First pass perfusion: mild enhancement
	First pass perfusion (vascularity)	LGE: heterogeneous with less enhancing central regions. Large areas of central necrosis or haemorrhage are less likely, as opposed to other cardiac tumours (e.g. angiosarcoma)
	LGE (+)	
CT	Calcification	Calcification not typical
	Solid component	Encasement of coronary arteries
	Isodense signal	Mediastinal lymphadenopathy may be present if it is not an isolated cardiac lymphoma
	Contrast enhancement	
^{18}F-FDG PET/CT	High SUVmax	Shows cardiac and extra-cardiac lesions
	High metabolic tissue volume	Reliable for staging
	High total lesion glycolysis	

^{18}F-FDG PET/CT, ^{18}F-flurodeoxyglucose positron emission tomography/CT; CMR, cardiac magnetic resonance; CT, computed tomography; LGE, late gadolinium enhancement; SUV, standardized uptake value; T1w-TSE, T1-weighted-Turbo Spin Echo; T2-STIR, T2 short tau inversion recovery; T2w-TSE, T2-weighted-Turbo Spin Echo.
[a]Signal intensity relative to myocardium.

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

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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