ESC European Society of Cardiology

European Heart Journal - Case Reports (2021) **5**(6), 1–2
doi:10.1093/ehjcr/ytab247

Brobdingnagian monstrosity at the right heart

Eslam Abbas [1]* and Ehab Al-Hefny [2]

[1]Department of OncoCardiology, Dar El Salam Cancer Center, 987 Nile Corniche, Al Kafor, Cairo 11559, Egypt; and [2]Department of Cardiology, Al Azhar University, Cairo 11651, Egypt

Received 26 January 2021; first decision 9 February 2021; accepted 25 May 2021

For the podcast associated with this article, please visit https://academic.oup.com/ehjcr/pages/podcast

A 52-year-old female, not known to be a smoker, diabetic, or hypertensive patient, presented with atypical chest pain and dyspnoea on exertion. Clinical examination was unremarkable, and the electrocardiogram showed atrial flutter. Transthoracic echocardiogram revealed an ill-defined large mass occupying the majority of the right atrium and infiltrating the free wall of the right ventricle for tissue characterizations via cardiac magnetic resonance (CMR) imaging. As shown in the pictures, CMR additionally revealed a voluminous infiltrating soft tissue mass (8.5 cm × 17 cm × 9.5 cm) arising from the lateral wall of the right atrium and centred upon the atrioventricular groove with patent right coronary artery and no extension outside the pericardium. The mass tissue elicited an isointense SI in the cine and T1 black-blood sequences, high signal intensity (SI) in the turbo inversion recovery magnitude (TIRM) sequences, and a heterogeneous enhancement in the first-pass perfusion as well as late gadolinium series. Tumour site, infiltration, and tissue characterization were suggestive of malignant, infiltrative cardiac tumour, mainly primary cardiac lymphoma. Additional differential diagnoses comprised undifferentiated sarcoma, rhabdomyosarcoma, fibrosarcoma, or metastasis[1,2] for histopathological correlation. Despite the size of the infiltrating mass; the right ventricle showed a fair systolic function

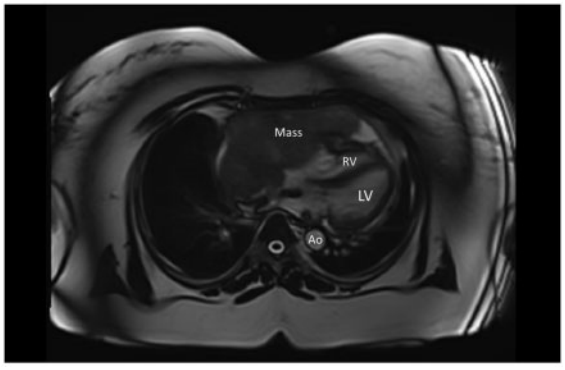


Figure 1 A cardiac magnetic resonance image (cine four-chamber) showing a large voluminous infiltrative soft tissue mass centred upon the right atrioventricular groove. The mass arises from the lateral wall of right atrium and extending to the right atrium cavity inseparable from the basal segment of the free wall of the right ventricle as well as the related overlying pericardium with no extracardiac extension.

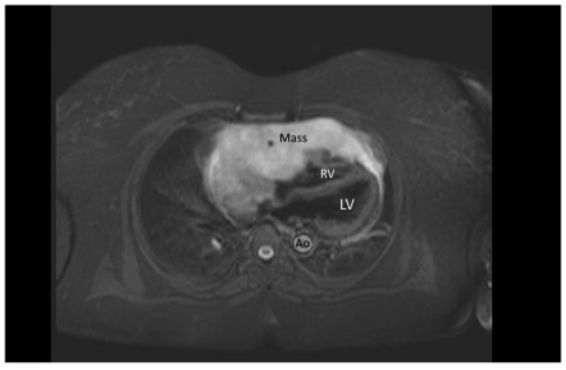
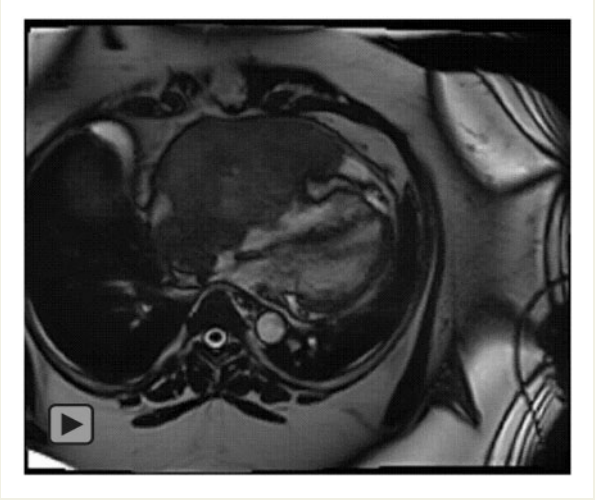


Figure 2 The corresponding TIRM sequence shows that the basal segment of the free wall of the right ventricle is infiltrated by the mass while the mid-ventricular and the apical free wall are compressed by the mass with no evidence of infiltration. TIRM sequence improves imaging contrast because of the increased T1-weighting and the inherent fat suppression. The high signal intensity of the examined mass in the TIRM sequence enables accurate evaluation of tumour extension and the presence of vascular infiltration. The mass encases the right coronary artery, yet it appears patent with no evidence of occlusion.

* Corresponding author. Tel: +20 102 305 4574, Email: islam.omr@med.au.edu.eg
Handling Editor: Edoardo Conte
Peer-reviewers: Raja Ezman Faridz Raja Shariff and Dimitrios Terentes-Printzios



Video 1 Imaging in the cine four-chamber plane depicts myocardial contractility and cardiac volumes in the presence of the soft tissue mass.

[ejection fraction (EF = 50%)] while the systolic function of the left ventricle was effectively preserved (EF = 65%); therefore, urgent surgical debulking was not warranted. Histopathological examination of the cardiac mass ultrasound-guided biopsy revealed large malignant lymphocytes consistent with diffuse large B-cell lymphoma. After a negative metastatic workup, a diagnosis of primary cardiac lymphoma was established. The treatment plan included (i) a prophylactic direct oral anticoagulant (Edoxaban) therapy against pulmonary embolism and (ii) a subcutaneous implantable cardioverter-defibrillator as primary prophylaxis against sudden cardiac death, followed by (iii) the chemotherapeutic R-CHOP protocol (rituximab 375 mg/m^2, cyclophosphamide 750 mg/m^2, doxorubicin 50 mg/m^2, vincristine 1.4 mg/m^2, and prednisone 100 mg)[3] of which she already received the first cycle. Doxorubicin is an extremely potent chemotherapeutic against B-cell lymphoma but may pose a cardiotoxic side-effect. To curb the incidence of toxic cardiomyopathy, special consideration has been given to maintaining the cumulative lifetime dose of doxorubicin under 450 mg/m^2, besides pre-treatment with angiotensin-converting enzyme inhibitors and beta-blockers as cardioprotective agents.[4,5]

The incidence of primary cardiac lymphoma is 0.01% of all cardiac tumours with no standard care. However, early detection and rapid diagnosis of the disease provide the best chance for achieving an optimum response in the form of complete remission. This case depicts an unusual presentation of a rare cardiac tumour, highlighting the role of CMR in the diagnosis, evaluation of mass extension, and accurate assessment of biventricular function in the presence of a bulky intracardiac mass.

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

Conflict of interest: none declared.

Funding: None declared.

References

1. Motwani M, Kidambi A, Herzog BA, Uddin A, Greenwood JP, Plein S. MR imaging of cardiac tumors and masses: a review of methods and clinical applications. *Radiology* 2013;**268**:26–43.
2. Aggeli C, Dimitroglou Y, Raftopoulos L, Sarri G, Mavrogeni S, Wong J et al. Cardiac masses: the role of cardiovascular imaging in the differential diagnosis. *Diagnostics* 2020;**10**:1088.
3. Coiffier B, Lepage E, Brière J, Herbrecht R, Tilly H, Bouabdallah R et al. CHOP chemotherapy plus rituximab compared with CHOP alone in elderly patients with diffuse large-B-cell lymphoma. *N Engl J Med* 2002;**346**:235–242.
4. Ewer MS, Ewer SM. Cardiotoxicity of anticancer treatments. *Nat Rev Cardiol* 2015;**12**:547–558.
5. Armenian SH, Lacchetti C, Barac A, Carver J, Constine LS, Denduluri N et al. Prevention and monitoring of cardiac dysfunction in survivors of adult cancers: American Society of Clinical Oncology Clinical Practice Guideline. *J Clin Oncol* 2017;**35**:893–911.