ESC European Society of Cardiology

European Heart Journal - Case Reports (2025) **9**, ytaf047
https://doi.org/10.1093/ehjcr/ytaf047

CASE REPORT
Inherited cardiac conditions/cardiomyopathies

Inflammation clashing onto myocardial susceptibility: a tale of two rare diseases—a case report

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Received 18 August 2024; revised 2 December 2024; accepted 23 January 2025; online publish-ahead-of-print 29 January 2025

Background Sarcoidosis is a rare inflammatory disease characterized by the presence of myocardial non-caseating granulomas. Heart failure, conduction abnormalities, and/or life-threatening arrhythmias are the main manifestations of cardiac sarcoidosis (CS). Cardiac magnetic resonance plays a major role in the diagnostic suspicion of cardiac involvement in sarcoidosis. However, late gadolinium enhancement (LGE) patterns are non-specific, and one should consider alternative or additional aetiologies for myocardial disease.

Case summary We report the case of a 40-year-old male with a past medical history remarkable for pulmonary and cutaneous sarcoidosis, presenting with asymptomatic premature ventricular contractions and severe left ventricular (LV) dilation and moderately reduced systolic function. Computed tomography angiography excluded coronary artery disease. Cardiac magnetic resonance revealed myocardial oedema in the anterior, anterolateral, and inferolateral walls and the presence of septal intra-mural and anterior, inferior, and lateral sub-epicardial 'ring-like' LGE. He had elevated inflammatory plasma biomarkers. N-terminal pro-brain natriuretic peptide was 110 pg/mL, and high-sensitivity troponin T was 20 ng/dL. Positron emission tomography computed tomography scan showed increased myocardial uptake consistent with inflammatory disease. Endomyocardial biopsy was normal. Thus, a presumptive diagnosis of isolated CS was made, and immunosuppression therapy was initiated, with full LV function recovery. Given the 'ring-like' LGE pattern, we recommended genetic testing, which identified a deletion in the dystrophin gene, classified as likely pathogenic.

Discussion This case highlights the contemporary diagnostic pathway for primary cardiomyopathies, emphasizing the increased likelihood of genetically influenced myocardial vulnerability to continuous harm when coupled with an acquired precipitant of myocardial damage. We describe a case of CS likely superimposed on a genetic myocardial substrate.

Keywords Cardiac sarcoidosis • Becker muscular dystrophy • Late gadolinium enhancement • Case report

ESC curriculum 2.3 Cardiac magnetic resonance • 2.5 Nuclear techniques • 6.5 Cardiomyopathy

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Handling Editor: Valentina Rossi
Peer-reviewers: Boldizsar Kovacs; Maria Lucia Narducci
Compliance Editor: Elton Luo

Learning points

- Ring-like late gadolinium enhancement pattern should alert the physician to other causes of primary cardiomyopathies, mainly genetic ones.
- Comprehensive diagnostic testing for dilated cardiomyopathy, including genetic analysis, helps identify pathogenic variants, guiding patient care and family screening.
- We report the case of a patient exhibiting dystrophinopathy, characterized by heightened susceptibility of the myocardium to a secondary affliction, such as sarcoidosis.
- This case illustrates the necessity of a multidisciplinary approach, integrating genetic counselling and tailored therapeutic interventions to optimize patient outcomes.

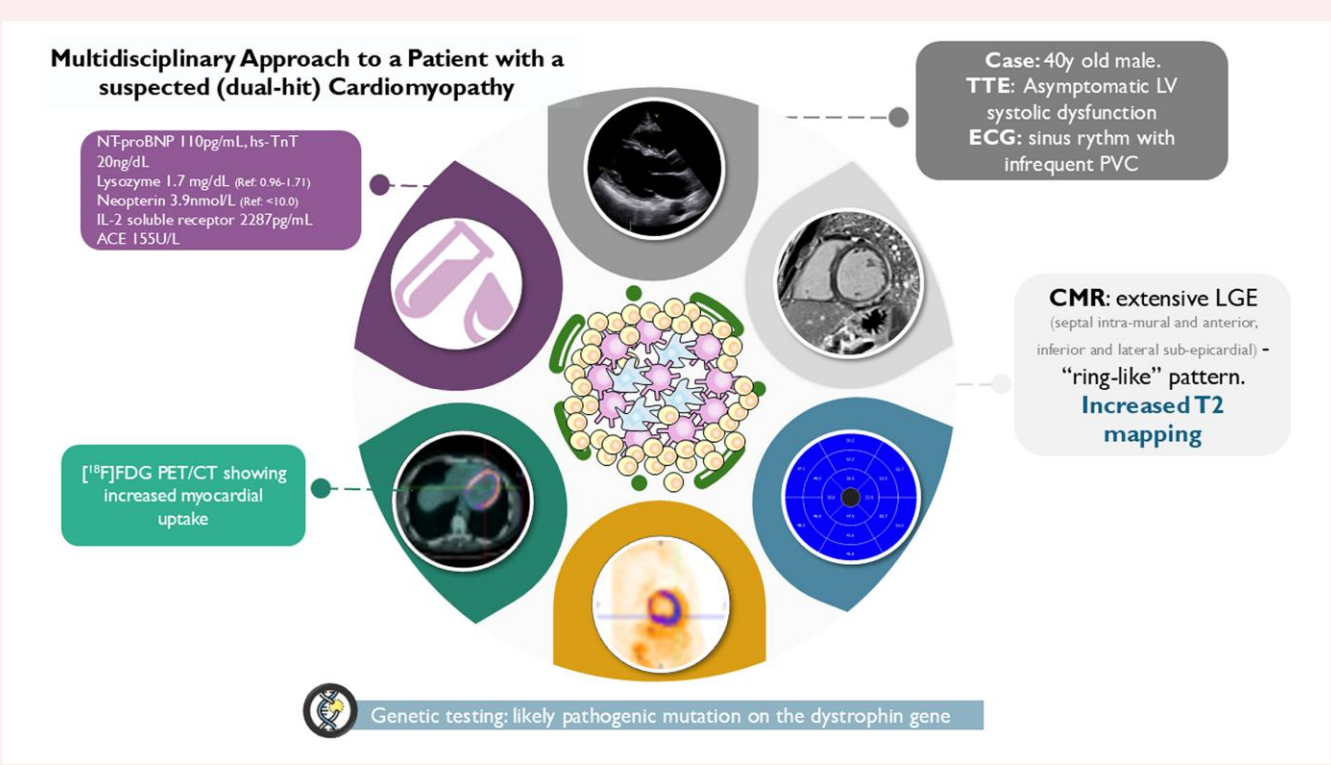
Introduction

Sarcoidosis is a multisystem, granulomatous disease of unknown aetiology, with ~25% of the patients having asymptomatic cardiac involvement, while 5% present with overt cardiac disease.[1] Clinical manifestations of cardiac sarcoidosis (CS) include new rhythm abnormalities and left ventricular (LV) systolic dysfunction, which, in a patient with a history of sarcoidosis, should alert the physician for possible CS.[2,3] Cardiac magnetic resonance (CMR) and 2-[^{18}F]fluoro-2-deoxy-D-glucose positron emission tomography computed tomography ([^{18}F]FDG PET/CT) scan are essential for diagnosing suspected CS, as they provide detailed structural and metabolic insights, improving disease detection and assessment[3] and monitoring response to immunosuppressive therapy.[4]

Increasing evidence highlights the crucial role of genetic variants in developing inflammatory cardiomyopathy/CS, potentially influencing disease pathophysiology, management, and risk stratification.[5] Genetic testing in CS patients has revealed that pathogenic or likely pathogenic variants in cardiomyopathy-susceptible genes—such as lamin A/C, desmoplakin, and titin—are present in 21–36% of patients.[6,7] This suggests that a pre-existing myocardial vulnerability may often present in CS.

We describe a case of a patient with asymptomatic CS in whom a genetically determined myocardial substrate for acquired inflammatory cardiac disease was identified.

Summary figure



[^{18}F]FDG PET/CT, 2-[18F]fluoro-2-deoxy-D-glucose positron emission tomography computed tomography; ACE, angiotensin-converting enzyme; CMR, cardiac magnetic resonance; hs-TnT, high-sensitivity troponin T; LGE, late gadolinium enhancement; NT-proBNP, N-terminal pro-brain natriuretic peptide; TTE, transthoracic echocardiogram; Ref, reference values. For the remaining reference values, please refer to text.

Case presentation

A 40-year-old man was referred to the outpatient cardiomyopathy clinic due to newly diagnosed asymptomatic LV systolic dysfunction on cardiac imaging performed after the incidental finding of premature ventricular contractions (PVCs) on pre-sports electrocardiogram. His medical history included previous cutaneous and pulmonary sarcoidosis, diagnosed at the age of 25 years and in remission since 2017 after corticosteroid treatment. He reported no known cardiovascular disease or relevant family history. He denied taking any regular medication. His physical examination was unremarkable. The blood work demonstrated NT-proBNP = 110 pg/mL, high-sensitivity troponin T = 20 ng/dL, and elevated inflammatory markers [soluble interleukin-2 receptor (sIL-2R) = 2287 pg/mL (normal: 458–1997); serum level of angiotensin-converting enzyme = 155 U/L (normal: 8–52)]. The 24-h Holter showed sinus rhythm and infrequent (9/h) polymorphous PVCs.

Transthoracic echocardiography revealed a severely dilated LV (indexed end-diastolic volume = 112 mL/m^2), 'scooping' (basal thinning) of the anterior septum, and moderate systolic dysfunction [LV ejection fraction (LVEF) = 41%, global longitudinal strain = −15.4%] (*Figure 1*; Supplementary material online, *Video S1*).

Cardiac magnetic resonance demonstrated diffuse myocardial oedema [elevated T2 mapping = 51 ± 6 ms (institutional cut-off = 50 ms)] mostly in the anterior, anterolateral, and inferolateral walls and an extensive (26% of LV mass) late gadolinium enhancement (LGE) with two patterns: intra-mural in the LV septum and sub-epicardial in the anterior, inferior, and lateral walls (*Figure 2*; Supplementary material online, *Video S2*). Coronary CT angiography excluded coronary artery disease.

Considering the initial investigation and his past medical history, the diagnosis of CS was suspected, and a whole-body [^{18}F]FDG PET/CT scan was performed, revealing increased myocardial uptake consistent with inflammatory myocardial disease (*Figure 3*), without uptake elsewhere. The right ventricle endomyocardial biopsy (EMB) was negative for sarcoid (see Supplementary material online, *Figure S1*). Hence, considering the lack of a histological diagnosis (either cardiac or extra-cardiac), a presumptive diagnosis was made based on multimodal imaging.

The patient was started on immunosuppression treatment with prednisolone 30 mg, and methotrexate regimen up-titrated to 20 mg plus ramipril, bisoprolol, and dapagliflozin. Three months later, follow-up imaging evidenced noticeable improvement with CMR depicting LVEF = 52% and normalization of the T2 values (49 ms). The LGE pattern and its extensiveness persisted (*Figure 4A*; Supplementary material online, *Figure S2*). There were no PVCs on follow-up 24-h Holter. Six-month [^{18}F]FDG PET/CT showed normalized myocardial uptake (*Figure 4B*).

Genetic testing was recommended and performed after counselling. A large hemizygous deletion was identified on the dystrophin gene (c.(93+1_94-1)_(960+1_961-1)del—classified as likely pathogenic), responsible for Becker or Duchenne muscular dystrophy (BMD/DMD). Given the possibility of CS as a 'second hit' for myocardial injury and despite LV function recovery with treatment, the 'heart team' recommended complete cessation of moderate-to-intense physical activity, including his usual competitive sports (hockey), and the implantation of a implantable cardio defibrillator (ICD) for primary prevention of sudden cardiac death.

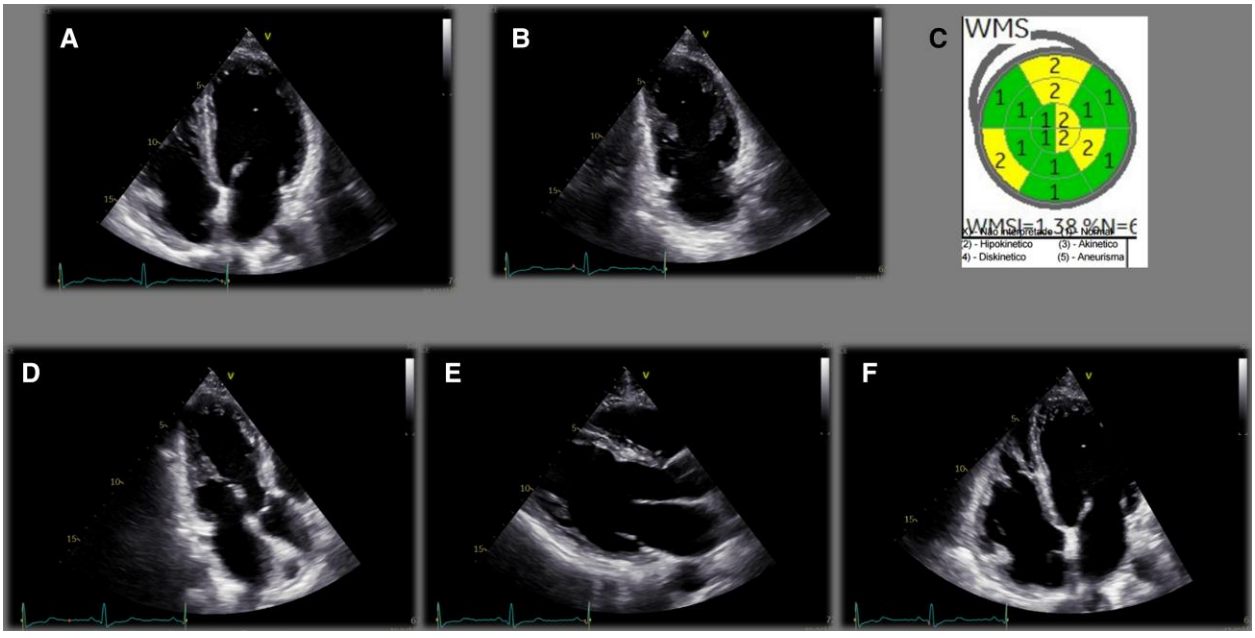


Figure 1 Initial transthoracic echocardiogram. Severe left ventricular dilation (left ventricular indexed end-diastolic volume = 112 mL/m^2) with moderate systolic dysfunction (Simpson biplane ejection fraction 41%, ejection fraction 3D 39%, global longitudinal strain −15.4%) due to non-segmental hypokinesia. 'Scooping' in the anterior basal septum (white arrow in *E*). Preserved cardiac index (2.7 L/min/m^2). Normal diastolic function. Mild dilatation of the left atrium (left atrium index volume 35 mL/m^2). Right atrium non-dilated. Slightly dilated right ventricle (basal diameter 51 mm, right ventricular indexed end-diastolic volume 67 mL/m^2) with normal systolic function (tricuspid annular plane systolic excursion 24 mm, S' 18 cm/s; free wall strain: −23.5%; radial: fractional area change 58%, right ventricle ejection fraction 3D 59%). No valvular abnormalities. No pericardial effusion. White arrow: basal thinning of the ventricular septum. (*A*) Apical four-chamber view. (*B*) Apical two-chamber view. (*C*) Wall motion bull´s eye. (*D*) Apical three-chamber view. (*E*) Parasternal long-axis view. (*F*) Right ventricle-focused apical four-chamber view.

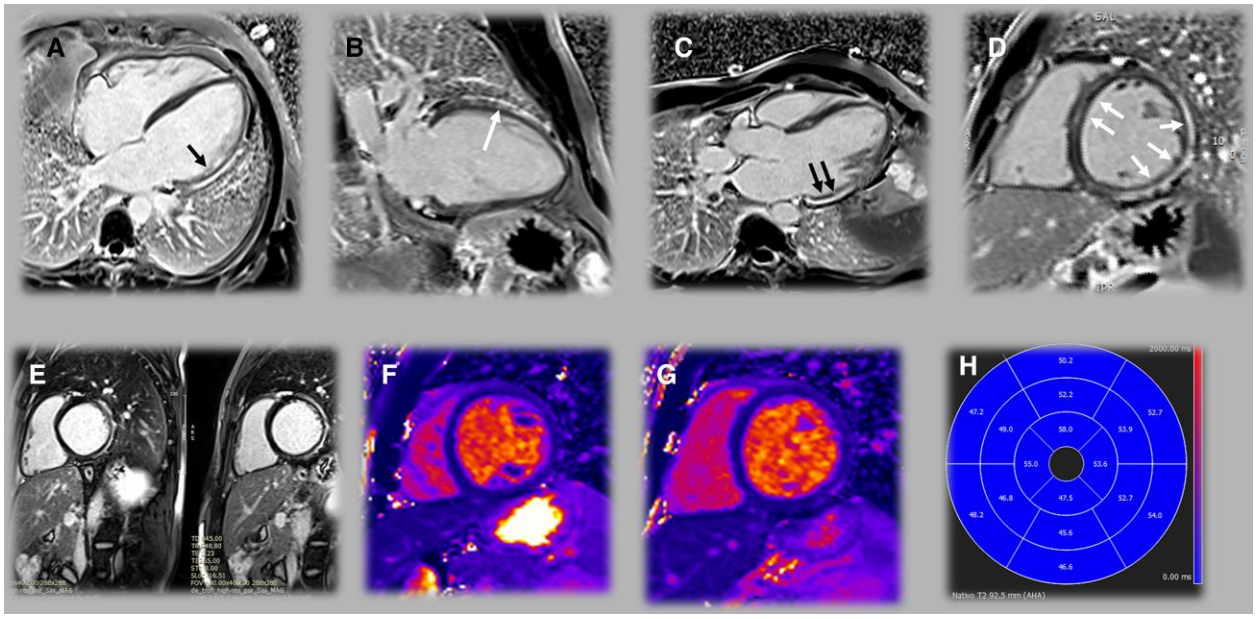


Figure 2 Cardiac magnetic resonance. Severe left ventricular dilation (left ventricular indexed end-diastolic volume = 140 mL/m^2), with moderate systolic dysfunction (left ventricular ejection fraction = 41%). Right ventricle with dimensions at the upper limit of normality (indexed end-diastolic volume = 122 mL/m^2) and preserved systolic function (ejection fraction = 50%). Extensive late gadolinium enhancement (white and black arrows): mid-wall septal pattern, extending into the sub-epicardial region in the anterior, inferior, and lateral segments and anterior, inferior, and lateral sub-epicardial ('ring-like' pattern). Native T1 globally within normal range (1019 ms; institutional cut-off 1079 ms), although slightly increased in the basal and mid-inferior and inferolateral segments (~1100 ms). T2 mapping sequence shows signs of active myocardial oedema (global T2 51 ± 6 ms; institutional cut-off 50 ms), predominantly in the anterior, anterolateral, and inferolateral walls. (*A*–*E*) Late gadolinium enhancement images depicting 'ring-like' pattern. (*F*, *G*) T2 mapping with P2-prep steady-state free precession sequence showing diffusely increased T2 values. (*H*) T2 mapping bull´s eye.

A final diagnosis of Becker muscular dystrophy with a cardiac phenotype exacerbated by the presence of a concomitant acquired inflammatory disease was made. The patient and his family were referred for genetic counselling. His mother is currently awaiting genetic results: if she carries the genetic mutation (indicating that it is not a *de novo* mutation in the index patient), we will recommend that the patient's brother undergo genetic testing (see Supplementary material online, *Figure S3*). As for the index case, follow-up includes regular clinic and laboratory evaluations every 3 months, cardiac imaging every 6 months, and a planned [^{18}F]FDG PET/CT scan at 2 years.

Discussion

This case represents a challenging scenario that underscores the critical importance of considering a primary myocardial disease exacerbated by inflammatory myocarditis. Given the patient's pulmonary sarcoidosis history and new LV dysfunction, CS was firstly considered.

Histological confirmation of CS is the gold standard for diagnosis, with the identification of typical epithelioid granulomas, but such an approach is hampered by the invasive nature and low sensitivity resulting from the patchy nature of the disease,[8] being positive in only 20–50% of cases.[3,9,10] While a definite diagnosis of CS requires an EMB and histopathology, a high diagnostic likelihood can be achieved by combining extra-cardiac histological confirmation of sarcoidosis with clinical manifestations and findings on multimodality cardiac imaging.[8]

In this case, extra-cardiac histologic confirmation of sarcoidosis was not pursued due to the isolated cardiac involvement, the invasiveness of extra-cardiac biopsy procedures, and the clinical parameters supporting the diagnosis. Notably, the patient had a history of pulmonary sarcoidosis, elevated angiotensin-converting enzyme and sIL-2R levels, and demonstrated a favourable response to immunosuppression, with normalization of LVEF and resolution of cardiac inflammation as evidenced by [^{18}F]FDG PET/CT imaging. According to the Heart Rhythm Society criteria,[2] CS is established based on a biopsy-proven diagnosis, while in the Japanese Circulation Society criteria,[3] histology is no longer mandatory, in the presence of typical imaging findings. Indeed, the diagnostic yield of EMB is low and a false negative may create the risk of the diagnosis being missed.[8] Electroanatomic mapping of the ventricular endocardium can help address this limitation by guiding EMB to scarred areas (i.e. the arrhythmogenic substrate), greatly increasing the diagnostic accuracy,[11] prompting an earlier diagnosis and targeted treatment. Nonetheless, in cases of highly probable CS with a history of extra-cardiac sarcoidosis, multimodality imaging may support the diagnosis of CS.[8]

Echocardiographic abnormalities are variable and non-specific, although thinning of the basal septum is a typical feature of CS.[2,3] Cardiac magnetic resonance offers a precise assessment of cardiac structure and function, the ability to detect myocardial oedema (i.e. active inflammation) although T2 imaging, and is highly sensitive to detect myocardial fibrosis, although LGE assessment.[8] There is no pathognomonic LGE pattern for CS[2,3]; however, it is usually 'patchy' and multifocal, mostly seen in the basal segments of the LV septum and lateral walls, with an intramural and/or sub-epicardial pattern,[1] although the 'ring-like' pattern has been infrequently described in CS. Whole-body [^{18}F]FDG PET/CT further corroborates our initial suspicion of CS, demonstrating increased myocardial inflammation typically

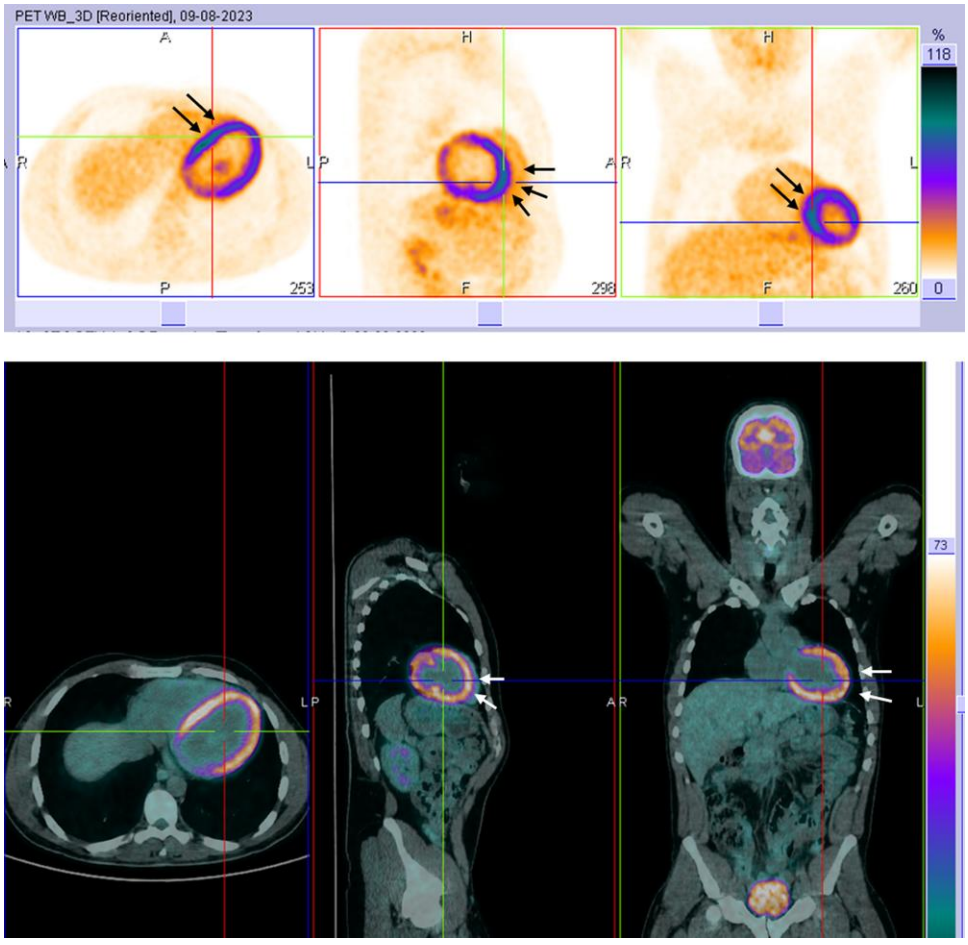


Figure 3 Whole-body 2-[^{18}F]fluoro-2-deoxy-D-glucose positron emission tomography computed tomography. Diffuse increased uptake in the myocardium (black and white arrows).

found in the active phase of CS. False-positive [^{18}F]FDG PET/CT results can occur if the pre-exam requirements of prolonged fasting (12–18 h) and a high-fat, lacking carbohydrates diet for 24–48 h are not followed, to minimize physiological uptake of the FDG tracer.[8] However, in our patient, these recommendations were strictly adhered to before each exam. The complete resolution of myocardial inflammation was observed in the follow-up exam, following immunosuppression. Lastly, both exams were conducted at the same centre, minimizing the likelihood of inter-observer variability. Nonetheless, we cannot entirely exclude the hypothesis of a false-positive result, particularly given the lack of a histopathological diagnosis of CS. Thus, based on the current guidelines,[3] a clinical diagnosis of CS based on imaging was made and immunosuppressive therapy was initiated.

Myocardial inflammation and fibrosis due to CS may cause ventricular arrhythmias and are associated with sudden cardiac death.[3] European Society of Cardiology guidelines recommend ICD implantation if LGE affects ≥22% of LV mass,[12] which has been associated with arrhythmic endpoints.[13,14] Moreover, our patient presented with a 'ring-like' LGE pattern, that, although rarely described in CS, suggested another possible cause for dilated cardiomyopathy.[15] It is often associated with arrhythmogenic cardiomyopathy, mainly desmoplakin and filamin C-truncating variants,[16] which can also present with 'myocarditis-like flares' and elevated T2 mapping values, complicating the differential diagnosis with CS. Hence, in the presence of these patterns, genetic testing is recommended for further aetiologic clarification, since it may lower the threshold for ICD implantation.[5]

A deletion on the dystrophin gene was found in the case presented. Dystrophin links actin cytoskeleton to the extracellular matrix, stabilizing sarcolemma and maintaining muscle cell integrity.[17] The documented deletion spans Exons 3 to 9, which encodes actin-binding sites (ABS) 2 and 3. In-frame deletions and missense mutations in Exon 2 (ABS1) are associated with a severe form of BMD/DMD, especially in cardiac phenotype, due to the low actin affinity and degradation of dystrophin.[17,18] Patients with DMD Exon 3–9 deletion, in which ABS1 is spared, are usually asymptomatic or may have a very mild BMD phenotype.[18] This supports why the disease may have emerged later in life in our patient, triggered by a 'second hit' (CS), and also clarifies the patient's favourable response to immunosuppressive therapy and neurohormonal blockade. Despite BMD being capable of showing myocardial inflammation on [^{18}F]FDG PET/CT,[19] several findings support our second-hit hypothesis of superimposed CS, namely prior pulmonary and cutaneous sarcoidosis history, elevated inflammatory markers indicative of activated macrophages, septal 'scooping' on TTE, and positive response to immunosuppressive therapy.

Lastly, the genetic mutation further strengthened the recommendation for ICD implantation.[12] Considering the likelihood of future atrioventricular block and the patient's history of mild BMD and severe CS flare, we opted for a transvenous ICD.

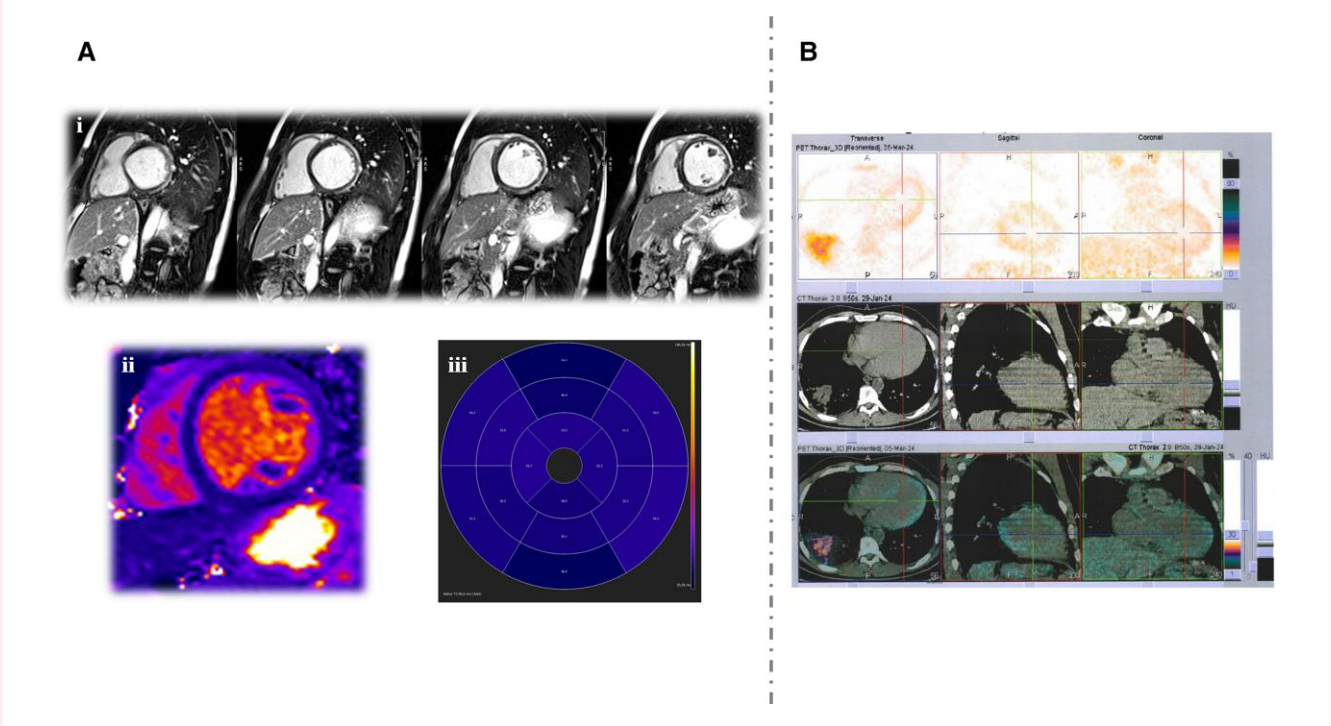


Figure 4 Follow-up imaging. (*A*) Cardiac magnetic resonance imaging: recovery of left ventricular ejection fraction (left ventricular ejection fraction 52%) and left ventricular volumes reduction (indexed end-diastolic volume 85 mL/m^2), with slight reduction of native T1 and T2 mapping (global T1 1019>>977 ms, global T2 52>>49 ms). Persistence of late gadolinium enhancement pattern previously described (*Figure 2*). (i) Late gadolinium enhancement images showing 'ring-like' pattern. (ii) T2 mapping with P2-prep steady-state free precession sequence. (iii) T2 mapping bull's eye. (*B*) 2-[^{18}F]fluoro-2-deoxy-D-glucose positron emission tomography computed tomography showing no abnormal myocardial uptake.

Conclusion

This case highlights the complex interplay between genetic predisposition and acquired inflammatory myocardial disease in a patient with a clinical diagnosis of isolated CS. The detection of a pathogenic variant emphasizes the importance of genetic testing in patients with atypical imaging patterns, which may help guide treatment, preventive strategies, family screening, and counselling.

Lead author biography

Maria Rita Lima received her master's degree in Medicine from the Faculty of Medicine, University of Lisbon in 2019. She is currently a cardiology intern at Santa Cruz Hospital, Lisbon, Portugal. She has previously published a review article regarding septic cardiomyopathy. Her current focus is on the pathophysiology of heart failure and primary cardiomyopathies and its management. She also has a special interest in cardiac advanced imaging.

Supplementary material

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

Acknowledgements

The authors would like to express their gratitude to all colleagues who cooperate in providing data for the current analysis.

Consent: The authors have obtained consent from the patient to publish this case report, including images, in accordance with the COPE guidelines.

Conflict of interest: None declared.

Funding: The present study was a local centre investigation, without funding by external entities.

Data availability

All data exposed in this case report were acquired from our institution, after obtaining informed consent from the patient.

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