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CASE REPORT

# A case of cardiac sarcoidosis mimicking cardiac amyloidosis on cardiovascular magnetic resonance

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## Abstract

A 52-year-old male visited our hospital with abnormal electrocardiogram and exertional fatigue. The electrocardiogram showed first-degree atrioventricular block, complete right bundle branch block, and inverted T waves in Leads II, III, aVF, V3, and V4. Echocardiography showed biventricular wall thickening involving granular sparkling of the interventricular septum. Late gadolinium enhancement on cardiovascular magnetic resonance (CMR) was found at the circumferential right ventricular wall and patchy regions of the left ventricle. Although these findings strongly suggested cardiac amyloidosis, he was finally diagnosed with systemic sarcoidosis due to the following. First, endomyocardial biopsy revealed non-caseating epithelioid granuloma with giant cells. Second, $^{18}$F-fluorodeoxyglucose positron emission tomography showed uptake in bilateral hilar lymph nodes, para-aortic lymph nodes, and the biventricular wall of the heart. Although echocardiography and CMR are very useful tools for diagnosis of cardiomyopathies, their specificity and accuracy need to be considered.

**Keywords** Cardiac sarcoidosis; Cardiac amyloidosis; Late gadolinium enhancement; $^{18}$F-Fluorodeoxyglucose positron emission tomography; Endomyocardial biopsy

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## Introduction

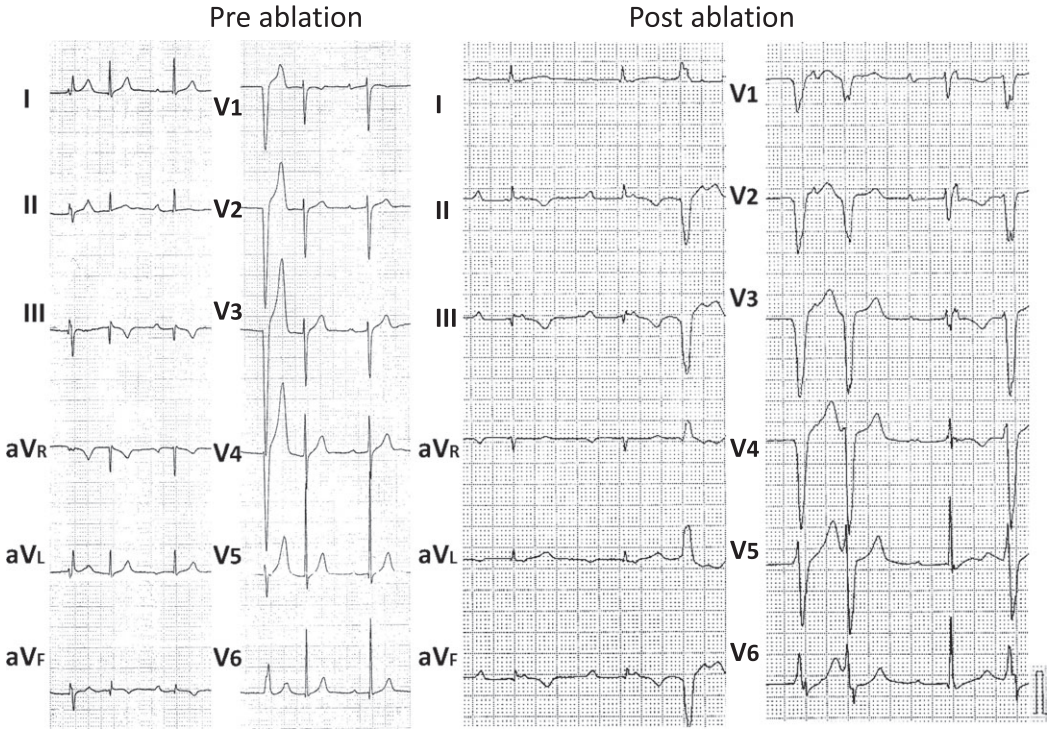
Cardiovascular magnetic resonance (CMR) is a useful diagnostic tool for cardiomyopathies.[1] It is well known that cardiac amyloidosis presents a characteristic pattern as global subendocardial late gadolinium enhancement (LGE) on CMR,[2] as well as on echocardiographic findings.[3] In cardiac sarcoidosis (CS), the LGE distribution on CMR shows a wide variety of types such as nodular, circumferential, subepicardial, and subendocardial types.[4,5] The differential diagnosis between cardiac amyloidosis and sarcoidosis is critical because it leads to different treatments and prognoses.

We experienced an unusual case of CS mimicking cardiac amyloidosis on echocardiography and magnetic resonance imaging findings, which was finally diagnosed by endomyocardial biopsy and $^{18}$F-fluorodeoxyglucose positron emission tomography (FDG-PET).

## Case report

A 52-year-old male came to our hospital with exertional fatigue. One year ago, he underwent radiofrequency catheter ablation at another hospital for frequent monofocal premature ventricular contractions originating from the right ventricular (RV) moderator band near the apex. Pre-operational echocardiogram showed first-degree atrioventricular block and inverted T waves in Leads III and aVF. At the first visit to our hospital, complete right bundle branch block and inverted T waves in Leads II, III, aVF, V3, and V4 were also recorded (*Figure 1*). On physical examinations, he had no leg oedemas, lymphadenopathies, or exanthemas. Chest X-ray did not show any pulmonary congestion, cardiomegaly, or bilateral hilar lymphadenopathy. With regard to laboratory data, serum creatinine level, BNP level, and Troponin I level were elevated (1.27 mg/dL, 154.2 pg/mL, and 0.131 ng/mL, respectively). However, serum angiotensin-converting enzyme level and

**Figure 1** Echocardiogram changes. Pre-ablation: A first-degree atrioventricular block (PQ intervals: 360 ms), inverted T waves in Leads III and aVF, and ventricular premature contraction are shown. Post-ablation: Complete right bundle branch block; inverted T waves in Leads II, III, aVF, V3, and V4; and ventricular premature contraction are shown.




soluble interleukin 2 receptor were not elevated (8.6 U/L and 116 U/mL, respectively). Echocardiography showed biventricular wall thickening with granular sparkling echogenicity of the interventricular septum (*Figure 2*). On echocardiographic assessment, enlarged RV (RV diastolic area 34 cm/m$^2$ and RV systolic area 28 cm/m$^2$), decreased RV fractional area change (18%), and diffusely reduced left ventricular (LV) ejection fraction (48%) with normal LV dimension (in diastole 49 mm and in systole 38 mm) were also recorded. His Doppler cardiac index was significantly decreased (1.42 L/min/m$^2$). CMR also demonstrated diffuse biventricular wall thickenings, and LGE was shown in both ventricles with high signal under T2-weighted image (*Figure 3*). Coronary artery disease was excluded from coronary computed tomography angiography. Although these findings strongly suggested cardiac amyloidosis, he was finally diagnosed with CS by FDG-PET (*Figure 4*), showing invasive uptake in the free wall of the RV and the anteroseptal wall of the LV, and endomyocardial biopsy

**Figure 2** Two-dimensional echocardiography. (A) On short axial views, the septal wall thickening (15.2 mm) with granular sparkling echo is shown, and diastolic dysfunction (E/e′ = 16.9) is prominent. (B) On enlarged image, granular sparkling echo is a characteristic.

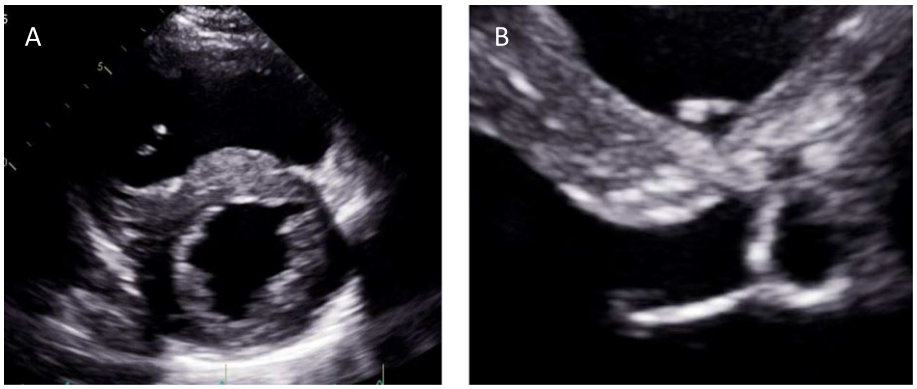


**Figure 3** Cardiovascular magnetic resonance images. On cine images, diffuse biventricular wall thickenings are shown. Late gadolinium enhancement is present in multiple locations, including both ventricles. On the T2-weighted image (Pre), a clear bright signal in the right ventricle (RV) and the anteroseptal wall of the left ventricle (LV) is noted, but after steroid treatment (Post), these lesions are not well delineated.

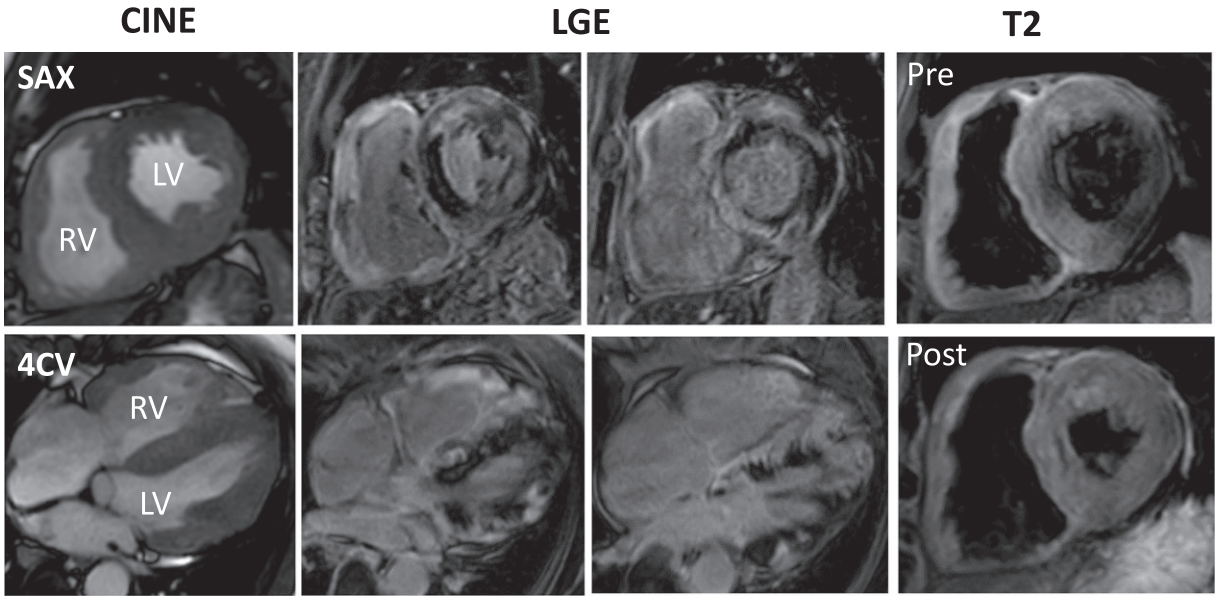
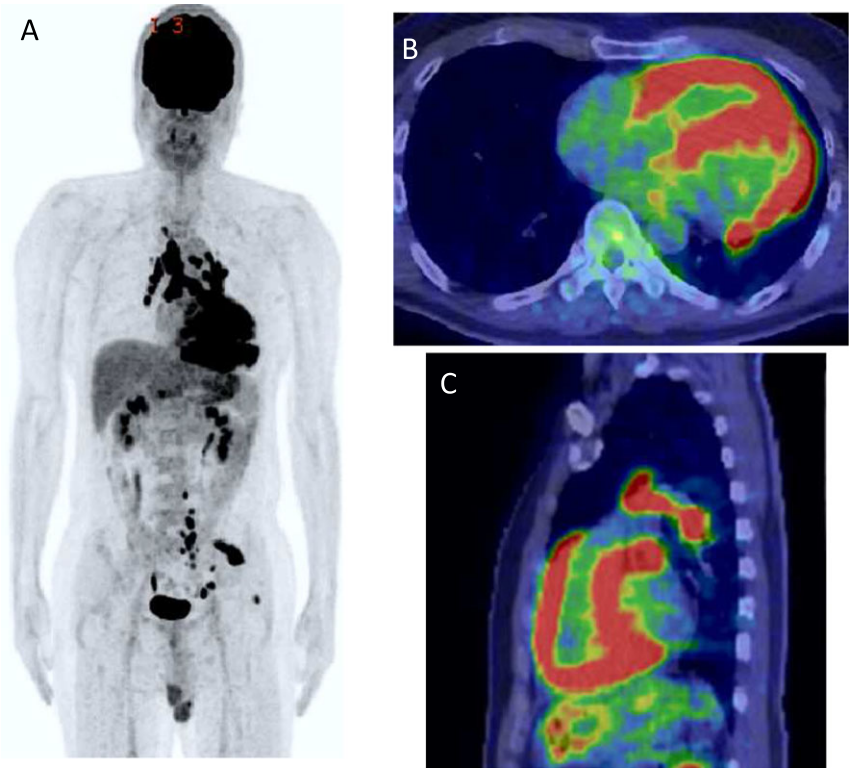



**Figure 4** $^{18}$F-Fluorodeoxyglucose positron emission tomography imaging. (A) On planar imaging, distinct and focal uptake of fluorodeoxyglucose (FDG) is shown in the bilateral hilar lymph nodes of the lung and the heart. (B) On a transverse image, intense uptake of FDG is shown in the right ventricle and anteroseptal wall of the left ventricle. (C) On a sagittal image, intense uptake of FDG is shown in the right ventricle and anteroseptal wall of the left ventricle and hilar lymph nodes. The regions of high FDG uptake show signal enhancement by T2-weighted cardiovascular magnetic resonance.





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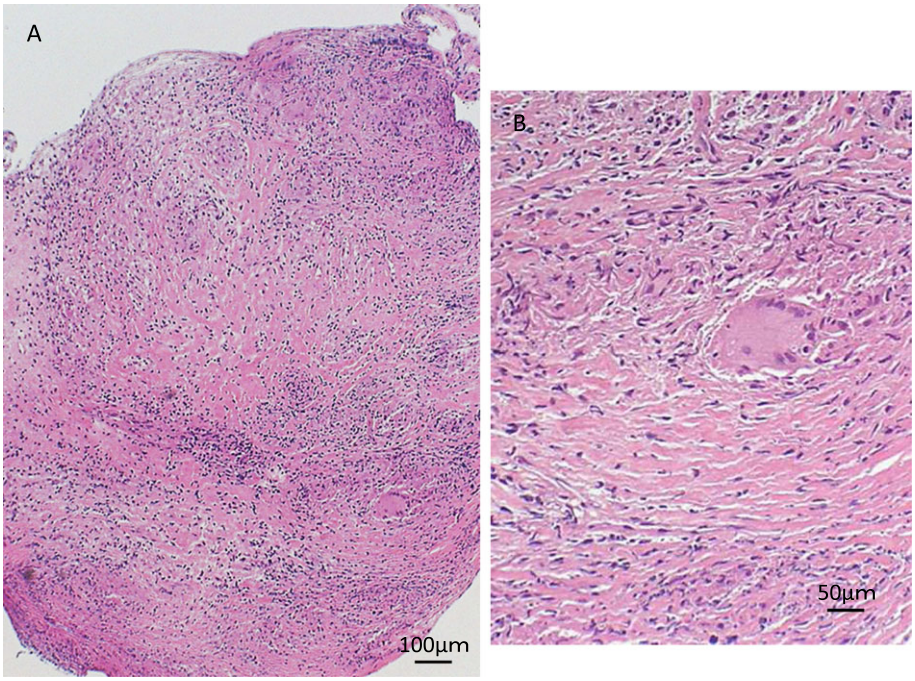
(*Figure 5*), showing non-caseating epithelioid granuloma with giant cells. The patient was treated with methyl prednisolone pulse therapy (1000 mg/day)[6] for 3 days, followed by oral prednisolone (30 mg/day). The dosage of prednisolone was gradually reduced. During hospitalization, sustained ventricular tachycardia (VT) originating from the RV lateral and inferior walls was frequently observed. VT still persisted in spite of radiofrequency ablation and administration of beta-adrenergic receptor blocker (bisoprolol fumarate 1.25 mg/day) and Class III anti-arrhythmic agent (sotalol hydrochloride 80 mg/day). However, after 3 months of steroid therapy, his cardiac function has significantly improved (LV ejection fraction 70%, RV fractional area change 40%, and Doppler cardiac index 2.34 L/min/$m^2$). Although his cardiac function improved, an implantable cardioverter defibrillator was implanted considering patient's age and preference. The patient has been followed at an outpatient clinic for a year. He experienced no worsening of cardiac function, heart failure, and VT thereafter.

## Discussion

This patient provided two important clinical suggestions. First of all, CS can present with biventricular wall thickening involving a granular sparkling appearance of the interventricular septum. Although it is well known that this ventricular wall thickening with granular sparkling pattern of the septum on echocardiography is characteristic of infiltrative cardiomyopathy such as cardiac amyloidosis,[3] septal thickening in CS is quite rare.[7,8] In CS, echocardiographic characteristics vary according to the disease activity and may reveal wall thickening due to inflammation of granulomatous legions and thinning due to fibrous scar formation. Furthermore, this case also showed a global high signal intensity in the RV and the septum and anterior region of the LV on T2-weighted images,[9] which correspond to the myocardial uptake regions on FDG-PET indicating granulomatous infiltration. Compared with that in pre-steroid condition, improved T2-weighted high signal intensity was observed after steroid therapy (*Figure 3*). This T2-weighted high signal intensity should be considered as a sign of cardiac inflammation and oedema in ongoing CS.

Second, CS can show global LGE in both ventricles on CMR. In our patient, LGE was observed over a wide range of the RV and in patchy and transmural regions of the LV, mimicking cardiac amyloidosis. In cardiac amyloidosis, accumulated abnormal proteins expand more widely in the interstitial space,[10] resulting in several different patterns of LGE on CMR such as entire subendocardial circumference, focal intramural, and subepicardial patterns in both ventricles.[2,11] On the other hand, it is well documented that LGE in CS is patchy and typically involves the basal septum and lateral LV walls, but rarely in the free wall of the RV.[4,5] Yared *et al*.[12] reported a case of CS imitating arrhythmogenic RV

**Figure 5** Histopathological pictures of a cardiac biopsied specimen from the right ventricle. Non-caseating epithelioid granuloma with giant cell and inflammatory cell infiltration is observed. Lower magnification (A) and higher magnification (B).





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dysplasia suspected by echocardiography, in which LGE was shown in the portion of the RV free wall and the subepicardial portion of the LV. On FDG-PET, intense uptake was observed in the RV and patchy in the LV, suggesting that these findings were similar to our case.

Recently, hybrid imaging of FDG-PET and CMR has been applied for non-invasive imaging of cardiac diseases.[13] In particular, cardiac positron emission tomography–magnetic resonance imaging is reported to be clinically feasible for the detection of cardiac inflammation such as active CS.[14,15] A combination of positron emission tomography and CMR may be more desirable in non-invasive imaging for CS to differentiate it from other types of cardiac hypertrophy.

In conclusion, CS can present as biventricular wall thickening involving granular sparkling of the interventricular septum and also shows a wide range of transmural and patchy LGE on CMR mimicking cardiac amyloidosis. Although CMR is a useful diagnostic tool, its interpretation needs to be considered.

## Conflict of interest

None declared.

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