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Identification of left ventricular chamber-like aneurysm related to cardiac sarcoidosis

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DESCRIPTION

A 58-year-old woman was referred to us with an abnormal ECG, showing complete right bundle branch block and left anterior fascicular block. An echocardiogram showed left ventricular (LV) septal thinning and hinted at the presence of an LV aneurysm. An enhanced CT indeed revealed an LV aneurysm-like structure. After appropriate testing to exclude coronary artery disease, this LV anomaly led us to suspect the possibility of cardiac sarcoidosis (CS). Cine cardiac magnetic resonance (CMR) imaging showed an apparent chamber-like aneurysm, and late gadolinium enhancement (LGE) CMR showed transmural positive enhancement in the mid-portion of the LV at the region of the aneurysm (figure 1A,B). As our institution lacked an FDG-PET scanner, gallium scintigraphy was instead performed, which showed uptake in the heart and bilateral hilar lymph nodes (figure 2A,B); hilar lymphadenopathy was undetected on enhanced CT, and its presence strongly indicated sarcoidosis. We finally made a clinical diagnosis of CS with LV aneurysm, as per the criteria set in the relative Japanese guidelines.

Steroid and anticoagulant therapy was initiated and the patient was followed up at our outpatient clinic. At 3-year follow-up, her CMR did not show any significant change in the LGE pattern or aneurysm size, and neither did gallium scintigraphy.

However, she had heart failure (HF) aggravated by ventricular tachycardia (VT), a QRS of 166 ms and a low LV ejection fraction, and we concluded that the patient would benefit from cardiac resynchronisation therapy-defibrillator (CRT-D) implantation to resynchronise the contractions and protect her from future fatal arrhythmias.

Sarcoidosis is a systemic disease characterised by epithelioid granulomas of unknown cause. CS is detected in 2%–7% of patients, but more than 20% of cases are clinically silent.¹ It is complicated with arrhythmias, resulting in sudden cardiac death or aggravating HF.² About 66% of patients with CS have the disease isolated to the heart.¹ Complete heart block, bundle branch block, VT, HF and sudden cardiac death are common presentations; however, CS-related ventricular aneurysms are only found in 10% of cases.² Implantation of a defibrillator is indicated for secondary prevention in patients surviving ventricular arrhythmias, including sustained VT and ventricular fibrillation.³ Interestingly, in our patient the arrhythmogenic area was mapped around the area of the LV aneurysm during the electrophysiological study, although the beneficial effect of CRT-D is at best only indirectly linked to the aneurysm. We emphasise the need for awareness of CS when ventricular aneurysms are discovered.

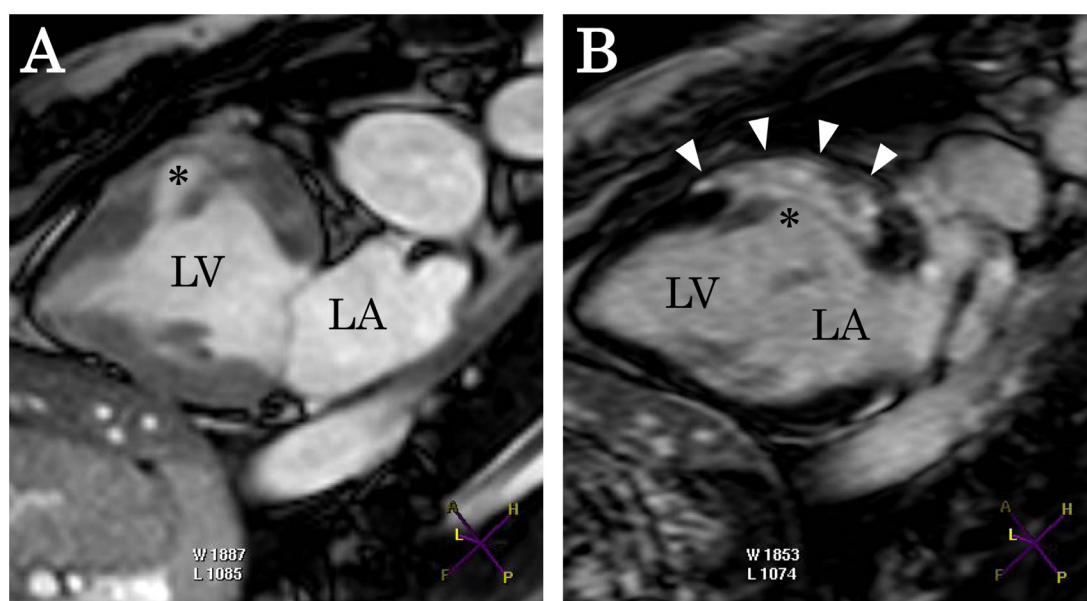


Figure 1 Cine CMR imaging showing the region of the LV chamber-like aneurysm (*) and late gadolinium enhancement CMR showing the transmural positive enhancement (arrowheads) at the region of the aneurysm. (A) Cine CMR and (B) late gadolinium enhancement CMR. CMR, cardiac magnetic resonance; LA, left atrium; LV, left ventricle.



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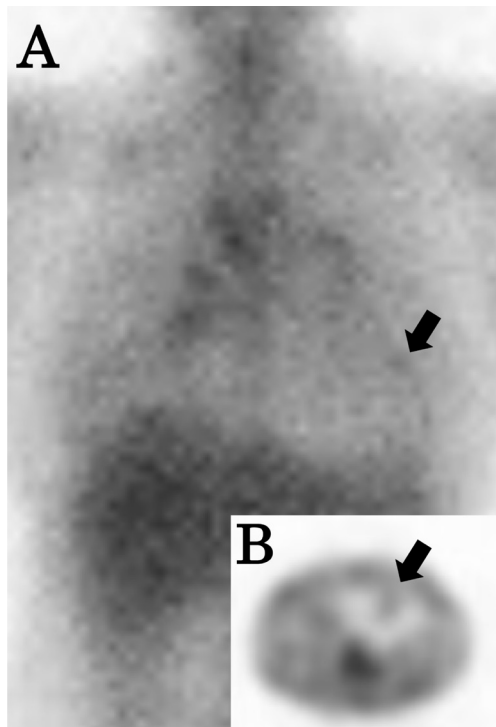


Figure 2 Gallium scintigraphy showing uptake in the heart (arrows) and bilateral hilar lymph nodes. (A) Coronal section view and (B) transverse section view.

Contributors All authors were involved in the clinical management of the patient. MS drafted and revised the manuscript. DK, SK and AH revised the manuscript.

Learning points

- Identification of an aneurysm in the left ventricle prompts consideration of cardiac sarcoidosis in the differential diagnosis.
- Cardiac MRI, especially with late gadolinium enhancement, is useful in the diagnosis of cardiac sarcoidosis.
- Cardiac sarcoidosis can be complicated with ventricular tachycardia and advanced heart failure.

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