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Case Report

Isolated cardiac sarcoidosis diagnosed by electroanatomic voltage mapping-guided endomyocardial biopsy combined with magnetic resonance imaging and positron emission tomography

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ABSTRACT

Cardiac involvement in sarcoidosis is related to lethal arrhythmias and is considered a serious condition. Because steroid therapy is an effective treatment, early diagnosis of cardiac sarcoidosis (CS) is of paramount importance in respect to improving prognosis. However, the diagnostic yield of histologic examination by endomyocardial biopsy (EMB) in CS is usually low. We report the case of isolated CS histopathologically proven by electroanatomical voltage mapping (EVM)-guided EMB combined with cardiac magnetic resonance imaging (CMR) and ^{18}F-fluorodeoxyglucose positron emission tomography (FDG-PET). A 53-year-old man presented with general fatigue. Electrocardiography showed intermittent complete atrioventricular block and echocardiography showed reduced cardiac function. CMR showed late gadolinium enhancement (LGE) in the areas of myocardium with suspected sarcoidosis. Next, we performed an EVM-guided EMB and found a non-caseating epithelioid granuloma in the right ventricular septum, which showed low voltage on EVM and LGE on CMR. FDG-PET showed accumulation in the same cardiac region. This case shows that EVM-guided EMB combined with diagnostic imaging can be a valuable approach in cases of suspected isolated CS.

<**Learning objective:** To reduce inflammation and prevent ventricular remodeling by early corticosteroid treatment, detecting cardiac sarcoidosis (CS)-affected regions is important. Cardiac magnetic resonance imaging and positron emission tomography help detect such regions. However, histopathological diagnosis of CS by endomyocardial biopsy (EMB) is usually difficult. The present case suggests that electroanatomical voltage mapping-guided EMB combined with diagnostic imaging improves detection of CS-affected regions.>

Introduction

Sarcoidosis is a multisystem non-caseating granulomatous disease of unknown etiology. The rates of cardiac involvement in sarcoidosis have been reported to be higher in patients from Japan than in those from Europe and the USA [1]. Cardiac involvement in patients with sarcoidosis affects cardiac function and causes ventricular failure, disrupts the cardiac conduction system, and leads to advanced atrioventricular (AV) block, malignant ventricular tachycardia (VT), and sudden cardiac death. Therefore, early diagnosis and treatment of this progressive and potentially severe condition are critically important.

The detection rates of non-caseating epithelioid granulomas by endomyocardial biopsy (EMB) in patients with cardiac sarcoidosis (CS) have been reported to be only 20% [2]. Therefore, the

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histopathological diagnosis is not easy especially in patients with isolated CS.

Diagnostic imaging methods such as cardiac magnetic resonance imaging (CMR) and positron emission tomography (PET) have been advancing rapidly. In patients with CS, a scar or intense inflammation can expand the extracellular space and causes a slower washout of gadolinium, resulting in areas of late gadolinium enhancement (LGE). PET with ^{18}F-fluorodeoxyglucose (FDG) is also useful in detecting infection and inflammation foci in patients with cardiomyopathy. Both CMR and FDG-PET are effective in assessing the extent and degree of disease in sarcoidosis [3]. The electroanatomical voltage mapping (EVM)-guided EMB is reported to be a useful method in detecting affected regions in isolated CS [4]. There have been no reports of histopathological diagnosis by EVM-guided EMB combined with diagnostic imaging in patients with suspected isolated CS.

In the current report, we present a case of CS diagnosed by EVM-guided EMB of an affected cardiac area with abnormal electrical activity, selectively combined with CMR and PET imaging.

Case report

A 53-year-old man was referred to our department because of aggravating dyspnea for several months, enlargement of the cardiac silhouette on chest radiograph, reduced cardiac function on echocardiography [left ventricular ejection fraction (LVEF) of 38%], and intermittent complete atrioventricular block (cAVB) on electrocardiography. He reported dyspnea and fatigue during work and AV block was detected on a medical examination earlier in the year.

At the first physical examination, his blood pressure was 116/72 mmHg. There were no rales and murmurs on auscultation, and no edema was noted on the extremities. He was diagnosed as having New York Heart Association (NYHA) functional class II, and his B-type natriuretic peptide (BNP) level was 98.5 pg/mL. His angiotensin-converting enzyme level was 10.3 IU/L (8.3–21.4 IU/L), and lysozyme level was 5.0 μg/mL (5.0–10.2 μg/mL). Thoracic computed tomography (CT) scan showed no hilar lymphadenopathy. Echocardiography showed low LVEF (35%), and thinning of the basal septum. His CMR showed a significantly increased signal intensity of LGE in the anteroseptal area and entire circumference of the area of basal and lateral wall (Fig. 1). Despite negative Ga-67 imaging, CS was suspected based on the significantly hyperintense epicardial LGE signal on CMR. Cardiac catheterization showed a left ventricular aneurysm and no significant coronary stenosis. Myocardial tissue samples were taken from the right ventricular (RV) apex during the first catheterization, but the biopsy failed to capture a non-caseating epithelioid granuloma.

The patient met the major criteria for a diagnosis of CS, according to the revised Japanese Ministry of Health, Labour and Welfare guidelines for the diagnosis of CS [5]: cAVB, thinning of the septum, and reduced LVEF. Apart from the heart, no other areas of suspected sarcoidosis were detected in the other organs. Histopathological analysis of the affected cardiac region was necessary for a diagnosis of isolated CS.

Therefore, we performed an EVM-guided EMB (Fig. 2A), performed concomitantly with induction and ablation of VT. In this study, sustained monomorphic VT was induced by RV apex pacing, but that was unmappable. We made a RV voltage map and recognized that the low voltage area from basal to mid lateral and basal to mid septal RV wall (Fig. 2B) was the same region as the increased LGE signal intensity area on CMR, and performed a biopsy of the mid septal RV wall. Selective EMB of the low-voltage areas was performed using a bioptome (Boston Scientific Corp., San Jose, CA, USA) with a 6-Fr long sheath placed into the right femoral vein.

The abnormal FDG accumulation only in the heart suggested isolated CS (Fig. 3A and B). A fused PET and CT image was reconstructed and significant myocardial FDG uptake was found in

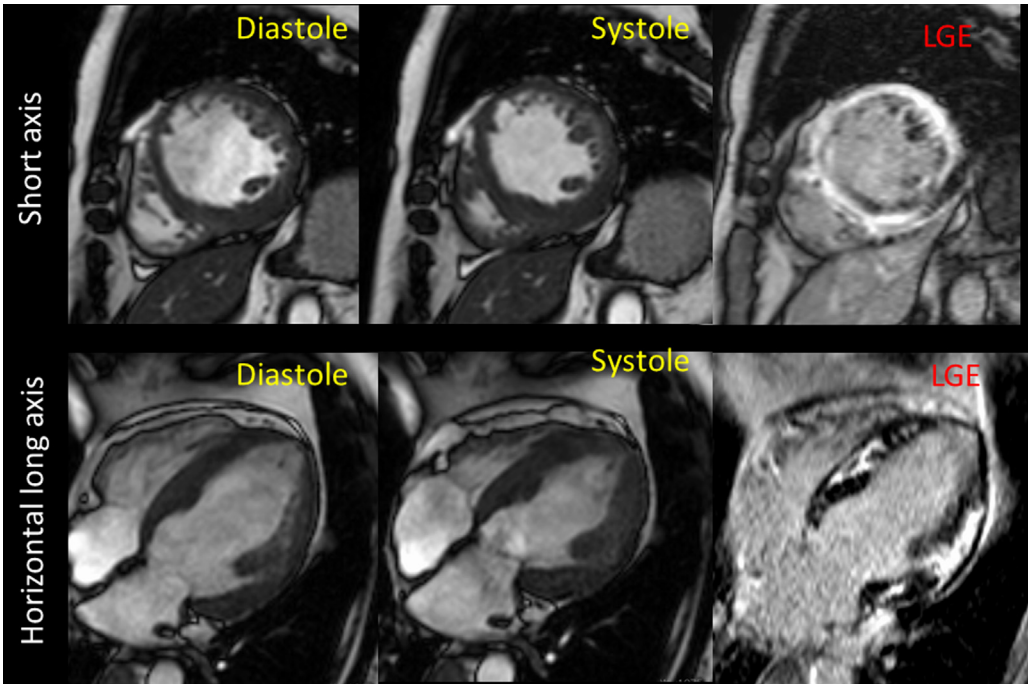

Fig. 1. Cardiac magnetic resonance imaging (CMR). CMR showed the thinning of the basal septum. Late gadolinium enhancement (LGE) showed a significantly increased signal intensity in anteroseptal area, and entire circumference of the area of basal and lateral wall.

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