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Case Report

Replacement myocardial fibrosis at the site of late gadolinium enhancement on magnetic resonance imaging in a patient with diffuse cutaneous systemic sclerosis: An autopsy report

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ABSTRACT

Late gadolinium enhancement (LGE) on cardiac magnetic resonance (CMR) is a well-known finding indicative of cardiac involvement in systemic sclerosis (SSc heart). However, few studies have reported the precise histopathology at the site of LGE. We present an autopsy report of a 51-year-old man diagnosed with diffuse cutaneous SSc according to a systematic diagnostic workup, including skin biopsy. CMR indicated left ventricular (LV) dilatation and broadly distributed subendocardial LGE in the LV walls. The patient was treated with methylprednisolone pulse therapy because of multiple episodes of ventricular tachycardia, whereas he subsequently died of left heart failure. An autopsy study revealed broad subendocardial replacement fibrosis, concomitant with the distribution of LGE on CMR, without inflammatory or edematous changes. Notably, myocardial fibrosis was evident around the intramural coronary arteries, although the arteries themselves were intact. These findings demonstrated that broad subendocardial LGE on CMR reflected replacement myocardial fibrosis in a patient with diffuse cutaneous SSc. These clinicopathological observations suggested that spasms in the intramyocardial arteries or the cardiac Raynaud's phenomenon may have provoked broad subendocardial fibrosis of the LV walls.

<**Learning objective:** The present autopsy report pathologically validated the presence of broad myocardial fibrosis in the area of subendocardial late gadolinium enhancement (LGE) on cardiac magnetic resonance in a patient with systemic sclerosis (SSc). Lack of inflammatory changes along with intact coronary arteries suggested the involvement of intramural coronary spasm in the development of cardiac involvement in SSc. Such an LGE pattern may suggest end-stage cardiac involvement and portend a poor prognosis.>

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Introduction

Systemic sclerosis (SSc) is a progressive disease that affects various organs, including the skin, lungs, kidneys, and heart. Cardiac involvement in SSc (SSc heart) is a well-recognized condition, characterized by fibrotic and inflammatory changes in the myocardium [1,2]. Clinically, SSc heart negatively affects the prognosis of patients with SSc [3]; thus, its early and accurate recognition is critical in the management of SSc.

Among various imaging modalities for the heart, cardiac magnetic resonance (CMR) is being increasingly used to evaluate SSc heart. Thus far, many studies have reported late gadolinium enhancement (LGE) as a promising indicator of SSc heart [4–7]. However, few of these studies have directly compared LGE images with gross or microscopic images of the heart; thus, the precise histopathology at the site of LGE remains unclear. The treatment of SSc heart varies depending on the pathological condition. Hence, clinicopathological studies comparing CMR images with histopathological observations of the heart are important in the clinical setting.

In the present autopsy report, we demonstrated that broad subendocardial LGE on CMR reflected replacement myocardial fibrosis in a patient with diffuse cutaneous SSc. Moreover,

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clinicopathological observations suggested that spasms in the intramyocardial arteries or cardiac Raynaud's phenomenon may have provoked broad subendocardial fibrosis of the left ventricular (LV) walls.

Case report

A 51-year-old man with Raynaud's phenomenon, sclerodactyly, and exertional dyspnea was referred to the Hokkaido University Hospital. Sclerotic change in the skin was distributed on the upper and lower extremities (forearms, upper arms, and lower legs), face, and trunk. He had an elevated antinuclear antibody titer of ×2560, and he was positive for anti-centromere antibody (index 179) and anti-ribonucleoprotein antibody (index 18.1) and negative for Scl-70 (14.4 index). A skin biopsy showed subdermal fibrotic change concomitant with SSc, and he was diagnosed with diffuse cutaneous SSc. The modified Rodnan total skin thickness score was 32. He had not exhibited chest symptoms indicative of angina pectoris or myocardial infarction.

His chest X-ray showed mild cardiomegaly, and the electrocardiogram revealed complete right bundle branch block and grade 1 atrioventricular block. An echocardiogram revealed a thin inferior LV wall and reduced LV wall motion. CMR indicated LV dilatation (LV end-diastolic volume, 221 mL), diffusely reduced LV wall motion (LV ejection fraction, 22.4%), and global subendocardial LGE in the LV wall (Fig. 1A–D, white arrowheads). LGE was also observed at the ventricular insertion point (Fig. 1B and C, white arrows). There were no relevant findings in T2-weighted images. High-resolution chest computed tomography revealed slight ground-glass opacities in both lower lobes, whereas a spirogram showed normal results. Coronary angiography exhibited normal coronary arteries. He reported multiple episodes of ventricular tachycardia with circulatory collapse and was treated with methylprednisolone pulse therapy (1 g/day). He did not develop renal crisis; however, his response to the steroid treatment was not favorable, and he subsequently died of left heart failure.

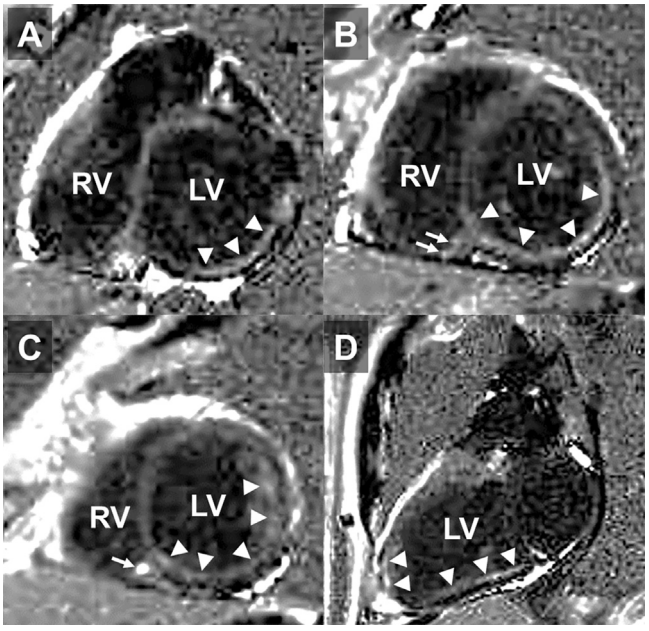

Fig. 1. Myocardial late gadolinium enhancement on contrast-enhanced magnetic resonance imaging. Late gadolinium enhancement (LGE) is globally noted in the subendocardial area of the LV walls (white arrowheads, panels A–D) and at the ventricular insertion point (white arrows, panels B and C). The LV is dilated with an increased LV end-diastolic volume (221 mL), and the ejection fraction is decreased to 22.4%. LV wall hypertrophy, as seen with cardiac amyloidosis, was not observed.
We used phase sensitive inversion recovery (PSIR) sequence for LGE more than 15 min after the administration of gadolinium. PSIR imaging can show the full negative-to-positive value range of magnetization and therefore, delivers better contrast between the normal myocardium and myocardium with fibrotic change. Because we waited for a sufficiently long time delay, the signal of the LV cavity showed a dark signal (the blood signal may be almost null at TI 325 ms). The scan parameters were TR700, TE1.1, TI325, and FA40.
RV, right ventricle; LV, left ventricle.

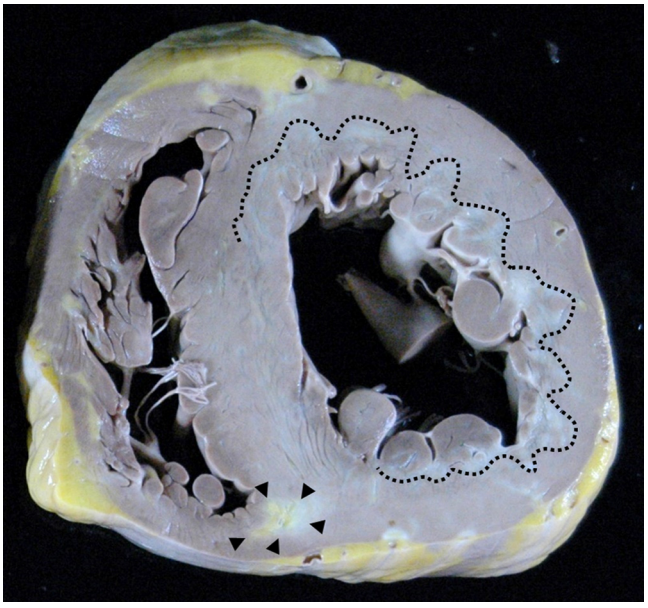

Fig. 2. Macroscopic findings of the heart. A white-to-gray colored area is noted in the subendocardium of the left ventricular walls (inside the dotted line) and at the ventricular insertion point (black arrowheads). The area mostly corresponded to the site of late gadolinium enhancement in the cardiac magnetic resonance image.

Macroscopic evaluation of his heart showed broad fibrotic change in the subendocardial area (Fig. 2, inside the dotted line) and at the ventricular insertion point (Fig. 2, black arrowheads), concomitant with the distribution of LGE on CMR. On microscopic evaluation (Fig. 3), there was replacement fibrosis predominantly in the subendocardial area (white arrows) and notably around the intramural coronary arteries, although the arteries themselves were intact (asterisks). There were no inflammatory or edematous changes and no amyloid deposition. Gross and microscopic evaluations noted no significant luminal narrowing with atherosclerotic changes in the epicardial coronary arteries.

Discussion

Previous studies have shown that the distribution of LGE in the heart is highly variable in patients with SSc. LGE has been reported to be linear [4,6,7] or patchy [2,6,7]; located in the midwall [2,4], subepicardial [2], or transmural area [6]; or present at ventricular insertion points [6,7]. However, few of these reports have documented pathological observations of the sites where LGE has been noted. Pieroni et al. and Krumm et al. conducted pathological analyses in patients with SSc with LGE on CMR [2,7]; however, they used myocardial biopsy, which allows investigations on only the right side of the interventricular septum. Some studies have reported subendocardial distribution of LGE on CMR in patients with SSc. However, histopathological evaluations at the LGE area were not performed [5]. Thus, the present report notably demonstrated that linear subendocardial LGE is one of the CMR findings suggestive of SSc heart.

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