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LETTER TO THE EDITOR

Contrast-Enhanced CMR Imaging Reveals Myocardial Involvement in Idiopathic Inflammatory Myopathy Without Cardiac Manifestations

Polymyositis (PM) and dermatomyositis (DM) are inflammatory diseases affecting skeletal muscles and other internal organs (1). Cardiac involvement occurs in up to 70% of patients and may be lethal (1). The diagnosis, based on electrocardiogram and laboratory and imaging investigations, remains a challenge because of nonspecific clinical presentation and the lack of standardized criteria (1). Cardiac magnetic resonance (CMR) is currently the best technique for diagnosing cardiac fibrosis (2).

Our aim was to evaluate patients with PM/DM without cardiac manifestations using CMR. Seven patients with PM (3 men/4 women) and 9 with DM (4 men/5 women) were studied. They were consecutively selected if they did not present any evidence of myocardial involvement, as shown by clinical and laboratory evaluation, and if they had no comorbidities, such as known coronary artery or valvular disease, obesity, diabetes, hypertension, hypercholesterolemia, alcoholism, and cardiac infections.

Their median age was 44 years (range 32 to 79 years) and the mean disease duration was 24 months (range 3 to 235 months), with 14 attaining remission of myopathy (6 PM, 8 DM, 8 to 12 months before the study), as shown by normal inflammatory indexes (erythrocyte sedimentation rate, C-reactive protein level), muscle enzyme assays, and improved muscle strength tests. Disease-associated involvement other than inflammatory myopathy was observed in all patients, including polyarthritis (5 PM, 1 DM), Raynaud phenomenon (3 PM, 2 DM), pulmonary fibrosis (2 DM), and sicca symptoms (1 PM, 1 DM); skin rashes appeared only in patients with DM.

Left ventricular (LV) function and late gadolinium-enhanced (LGE) areas were evaluated by 2 readers, and the kappa value was 0.86. Data, presented as medians and ranges, were compared by 2-sided Student *t* test and Fisher exact test. Values of $p < 0.05$ were considered to be statistically significant. In all patients, values were within normal range for LV end-systolic volume (median 38 ml, range 23 to 64 ml), LV end-diastolic volume (median 123 ml, range 86 to 184 ml), and LV ejection fraction (median 70%, range 52% to 78%).

LGE was detected in 9 of 16 patients with PM/DM (56.3%, including 6 of 7 patients with PM and 3 of 9 with DM). There was a trend for more frequent LGE among patients with PM, compared with those with DM ($p = 0.060$). LGE was epicardial or intramyocardial, sparing the subendocardium (typical of past inflammation) and involved approximately 5% of myocardial mass (median 4.4%, range 3% to 7%). LGE involved the intraventricular septum in 8 patients (5 with PM, 3 with DM), inferior wall in 3 (2 PM, 1 DM), and lateral wall in 5 (3 PM, 2 DM). The anterior wall and apex were affected in 1 and 2 patients with PM, respectively. Lesions involving more than one area were identified in 4 patients with PM and 3 with DM. LGE images are presented in Figures 1 and 2.

The frequency of heart involvement in PM/DM varies between 6% and 75% (1,3). Although clinically overt disease is rare, cardiac events are common causes of death (1). Cardiac troponin I is the most reliable index of myocardial damage (1,3). Reported experience about CMR in PM/DM is quite limited (3).

In this study, we retrospectively assessed the possibility of silent myocardial involvement in PM/DM using CMR. Our results revealed LGE in patients with PM/DM without

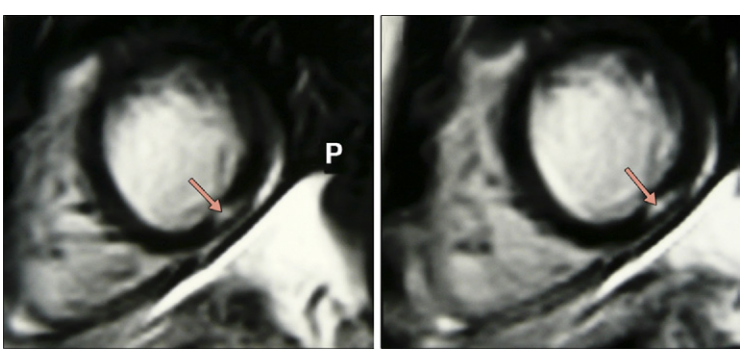

Figure 1. LGE Area in the Inferior Wall of the LV of a Patient With PM

Late gadolinium-enhanced (LGE) area in the inferior wall of the left ventricle (LV) in a patient with polymyositis (PM) **(arrows)**, without clinical evidence of heart involvement. P = posterior.

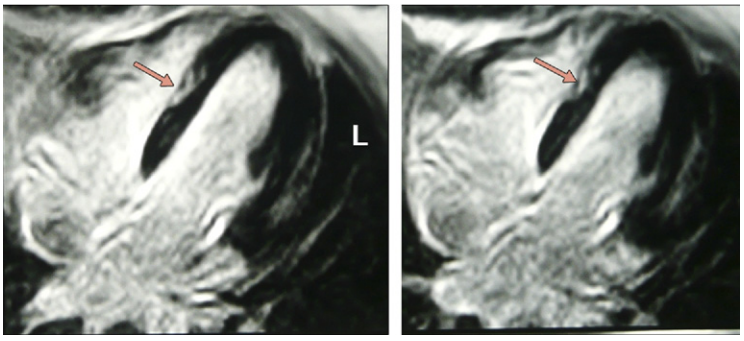

Figure 2. LGE Area in the IVS of the LV of a Patient With DM

LGE area in the intraventricular septum (IVS) of the LV in a patient with dermatomyositis (DM) **(arrows)**, without clinical evidence of heart involvement. L = left; other abbreviations as in Figure 1.

previous cardiac manifestation. The trend toward more frequent occurrence of LGE in PM, compared with DM, is worth mentioning; however, a study with a larger number of patients is necessary for definitive conclusions. As a preliminary report, our study has several limitations. First, the imaging data were not supported by biopsy (arduous in PM/DM because of patchy lesions and the retrospective study design). Second, evaluation of T2 and early T1 was missing. However, LGE bears prognostic value, beyond other common predictors (4).

In conclusion, CMR unveiled silent myocardial involvement in PM/DM. To establish the utility of CMR as a routine diagnostic approach in PM/DM, further studies are mandatory, including CMR in early and late PM/DM and correlation with clinical/laboratory data.

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REFERENCES

1. Lundberg IE. The heart in dermatomyositis and polymyositis. Rheumatology 2006;45:iv18–21.
2. Mavrogeni S, Spargias K, Markussis V, et al. Myocardial inflammation in autoimmune diseases: investigation by cardiovascular magnetic resonance and endomyocardial biopsy. Inflamm Allergy Drug Targets 2009;8:390–7.
3. Allanore Y, Vignaux O, Arnaud L, et al. Effects of corticosteroids and immunosuppressors on idiopathic inflammatory myopathy related myocarditis evaluated by magnetic resonance imaging. Ann Rheum Dis 2006;65:249–52.
4. Kwong RY, Chan AK, Brown KA, et al. Impact of unrecognized myocardial scar detected by cardiac magnetic resonance imaging on event-free survival in patients presenting with signs or symptoms of coronary artery disease. Circulation 2006;113:2733–43.