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CARDIOLOGY
Case Report

Cardiac Magnetic Resonance in Dermatomyositis Induced Myocarditis: Should we treat the Patient or the Images?

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We report a case of 42 years old woman, smoker, with history of biopsy proven dermatomyositis (DM), 8 years ago and normal heart function, who was admitted due to fever, rapidly progressive dyspnoea and orthopnoea. Her ECG presented sinus tachycardia (120 bs/min), LAH, mitral P waves and poor progression of R in the precordial leads. She had gallop rhythm and the biochemical profile showed SGOT = 118, SGPT = 100, γGT = 105, cTnI = 0.16. The 2D evaluation revealed pericardial effusion, pulmonary hypertension, left atrial and biventricular dilatation, diffuse LV hypokinesia and severely impaired LV-RV function. Coronary angiography and right heart catheterization were normal.

Carvedilol 6.25 mg ½ bid, furosemide 40 mg 1 bid, spironolacton 50 mg ½ once per day, and ramipril 4 mg opd was applied to treat the cardiac emergency. Additionally, intravenous, methtylprednisone (250 mg) and immune globulin (50 gr) were given. Few days after treatment, marked improvement of cardiac symptoms was occurred and she was discharged with intravenous immune globulin (100 gr) and methylprednisone (125 mg), once per month for six months and subcutaneous methotrexate 10 mg once per week. After six doses of immune globulin and methylprednisone, she started a monthly therapy with Rituximab 1000 mg and she did not report any cardiac symptom again.

Cardiac magnetic resonance (CMR) was performed 10 days after the development of cardiac symptoms and also on an annual basis every year for the next two years. STIR T2 - W and T1 - W multislice spin-echo images were obtained with identical parameters before and after 0.1m mol/kg Gd-DTPA. Early gadolinium enhancement measurement (EGE) was calculated within 1 minute of Gd-DTPA injection in the same area of myocardium and in latissimus dorsi, as in T2-W. Immediately after the second set of T1-W, 0.1m mol/kg Gd-DTPA was given again and late gadolinium-enhanced images (LGE) were taken 15 min later. T2 > 2, EGE > 4 and in case of unmeasurable EGE, due to muscular inflammation, early postcontrast myocardial signal (PostCMSI) > 50% higher than the precontrast, were considered abnormal [2-4]. The study was considered as positive, if at least 2/3 criteria [3] were abnormal. For left ventricular (LV) function, a steady-state free-precession (SSFP) sequence was used. Ejection fraction (EF) was calculated as: EF = [(volume at end-diastole – volume at end-systole)/volume at end-diastole].

In T2-W and EGE the signal ratio was measured in LV myocardium as well as within latissimus dorsi in the same slice. To assess LGE, all short-axis LGEs were summed and yielded the total volume, expressed as LV proportion (% LGE).

The 1 rst CMR study revealed T2 = 3.5, EGE = 32 and extensive intra-myocardial LGE in intraventricular septum, inferior and lateral wall, due to acute myocarditis. Additionally, diffuse hypokinesia, with left ventricular end diastolic (LVEDV) and end systolic (LVESV)

volume 117 and 74 ml respectively and LVEF = 37%, was also assessed. The 2nd and 3rd CMR performed after one and two years respectively, assessed LV function amelioration, but without reaching normal values. Additionally, T2, EGE and LGE remained abnormal (Figure 1A-B).

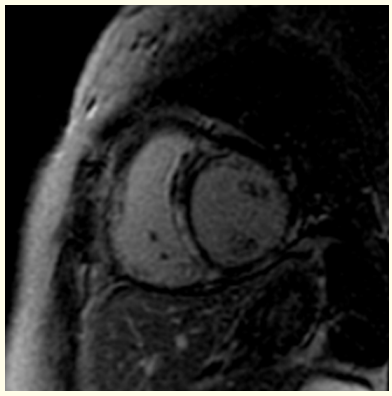

Figure 1A: *Short axis of the heart showing increased LGE (first evaluation).*

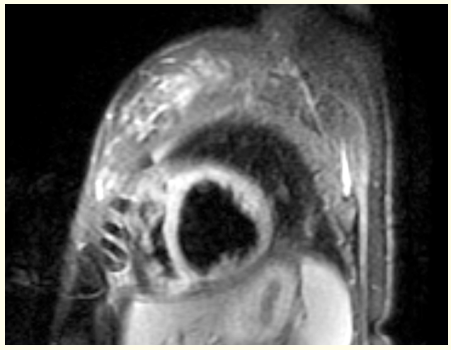

Figure 1B: *Short axis of the heart showing increased LGE (first evaluation).*

Polymyositis (PM)/dermatomyositis (DM) are inflammatory myopathies (IM), occasionally complicated by myocarditis. However, the correlation of cardiac manifestations with overall disease severity is controversial. To diagnose myocarditis in IM remains a challenge, due to atypical clinical presentation, high risk of death and lack of standardized diagnostic criteria. Until now, the algorithm used for IM myocarditis documentation is based on ECG, laboratory and echocardiographic findings, but these criteria are of limited value, due to lack of sensitivity and specificity [1].

CMR has been proven of value for the diagnosis and follow-up of various types of myocarditis, ranging from viral to autoimmune diseases [2-4], because of the advantage to diagnose oedema, cell infiltration and fibrotic lesions in one examination [2,3], even in cases with silent clinical presentation. Reported experience about CMR for heart evaluation in PM/DM is quite limited [5-7]. There are only two CMR studies in IM, before and after treatment. In one of them, Allanore., *et al*. [6] reported both clinical and CMR amelioration in 4 IM evaluated after six months of immunosuppression. In the other study, published by our group [7], the majority of clinically improved IM, had abnormal CMR three months after treatment, although the disease presented complete clinical remission.

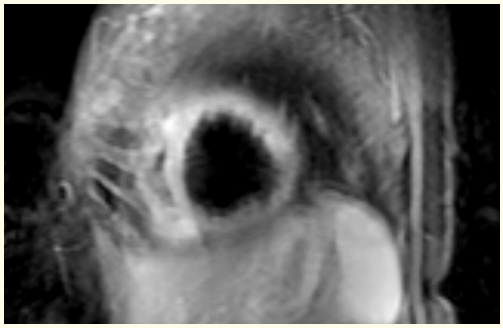

Figure 1C: *Short axis of the heart showing increased LGE (first evaluation).*

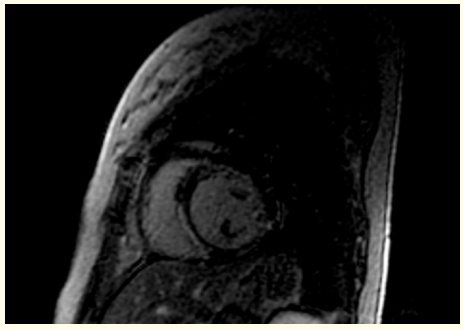

Figure 2: *Short axis of the heart showing LGE persistence (evaluation after 2 years).*

To our knowledge, this is the only case of IM induced myocarditis in which CMR was still positive for acute myocarditis two years after intensive cardiac and immunologic treatment. There are no data comparing clinical findings, CMR and endomyocardial biopsy in IM. However, it was documented in other immunologic disorders, like lupus nephritis, that there is not always agreement between clinical improvement and biopsy findings [8]. Under these circumstances, in IM induced myocarditis, which is usually characterized by lack of overt clinical presentation, the persistence of abnormal CMR findings should not be ignored. However, it remains to be proved the best type and duration of immunologic treatment and the clinical implications of the persistently abnormal CMR in IM survival.

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