Objectives: To investigate the prevalence of myocarditis in patients affected by IIM in and to determine whether the presence and extent of myocardial involvement identify a distinct disease phenotype.

Methods: 42 longitudinally followed IIM patients were routinely screened for myocardial involvement during a median [IQR] follow-up time of 4.2 [2.8-5.5] years. Patients with secondary causes of myocardial dysfunction were not included. Patients were considered to have myocarditis in case of: i) abnormal elevation of both circulating troponin T and troponin I, ii) signs of myocardial inflammation or necrosis/ fibrosis at cardiac MRI, or iii) positive myocardial tissue histology. Demographic, clinical and serologic features of patients with myocarditis were compared to those with no sign of myocardial involvement. Moreover, we determined whether the extent of myocardial involvement based on troponin levels predicts skeletal muscle disease severity.

Results: 57.1% (24 of 42) of patients had myocarditis. The frequency of myocardial dysfunction was similar among patients with DM, PM, IBM or IMNM and was not related to autoantibody positivity. Myocarditis was not associated with sex or ethnicity. Patients with or without myocarditis were similar in terms of age at disease onset and extra- muscular manifestations including dysphonia, dysphagia, arthralgias or arthritis. Raynaud phenomenon or intermittent lung disease. Independent of the IIM subtype, the presence of perimysial macrophages at skeletal muscle biopsy seems to protect from myocarditis development (p=0.04). Patients with myocarditis had higher median [IQR] levels of aldolase (10.9 [7.8-15.8] vs. 5.6 [4.9-8.6], p=0.014) and creatine kinase (1785 [966-5852] vs. 685 [168-2255], p=0.04) compared to patients with no myocardial dysfunction. Among patients with myocarditis, levels of troponin I negatively correlated with manual muscle testing 8 (MMT8) score (r=-1, p<0.01), strength in biceps (r=-0.95, p<0.014) and wrist extensors (r=-0.95, p=0.014) at last visit. Troponin T and troponin I titers were similar among patients with different IIM subtypes. C-reactive protein (p>0.04) but not erythrocyte sedimentation rate was found to predict myocardial involvement.

Conclusion: Our findings suggest that myocarditis is a frequent occurrence among patients with IIM and should be routinely ruled out. A more severe skeletal muscle disease is associated with an increased likelihood of myocarditis development, presumably due to higher systemic disease activity or inefficient disease control. The extent of myocardial damage faithfully reflects the severity of skeletal muscle dysfunction.

References:

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