BIOPSY-PROVEN ACTIVE MYOCARDITIS IN SYSTEMIC SCLEROSIS PATIENTS WITH RECENT-ONSET CARDIAC INVOLVEMENT

ACC Poster Contributions
Georgia World Congress Center, Hall B5
Tuesday, March 16, 2010, 9:30 a.m.-10:30 a.m.

Session Title: Myocarditis and dilated cardiomyopathy
Abstract Category: Cardiomyopathies/Myocarditis/Pericardial Disease
Presentation Number: 1231-43

Authors: Costantino Smaldone, Maurizio Pieroni, Maria De Santis, Gaetano Zizzo, Mara Campioni, Antonella Ventrone, Antonia Camporeale, Andrea Macchione, Anna Severino, Fulvio Bellocci, Gianfranco Ferraccioli, Filippo Crea, Catholic University of the Sacred Heart, Rome, Italy

Purpose: Cardiac involvement is a frequent finding and a main prognostic determinant in systemic sclerosis (SSc) patients, but the mechanisms and the histological substrate of myocardial damage are still largely unknown. We evaluated endomyocardial biopsy (EMB) findings in SSc patients with evidence of cardiac involvement.

Methods: Scleroderma patients presenting recent-onset symptoms or signs (arrhythmias, decrease in ejection fraction, wall motion abnormalities) of cardiac involvement associated with an increase of cardiac enzymes, were submitted to cardiac magnetic resonance (CMR), coronary angiography and right ventricular EMB. Myocardial specimens were processed for histology, immunohistochemistry and polymerase chain reaction for cardiotropic viruses.

Results: Among 189 consecutive SSc patients, 8 (4.2 %; 6 F, 2 M) showed evidence of recent-onset cardiac involvement with increased cardiac enzymes. Clinical presentation included dyspnea in 6 cases, wall motion abnormalities and/or reduced ejection fraction in 5, ventricular arrhythmias and chest pain in 3 and 2 patients respectively. CMR identified areas of delayed enhancement (DE) compatible with the diagnosis of myocarditis in 5 cases. Coronary arteries were normal in all patients. Histology showed the presence of inflammatory infiltrates with necrosis of adjacent myocytes diagnostic for active myocarditis in 7 patients. Immunohistochemistry showed a prevalence of activated T lymphocytes in all cases. In the remaining patient a borderline myocarditis was observed. No evidence of vasculitis nor thickening of arteriolar walls was found. Polymerase chain reaction identified parvovirus B19 genome in 3 patients with active myocarditis. Three patients with no evidence of virale genome received high dose steroids therapy and showed an improvement of symptoms with regression of DE areas at cardiac MRI.

Conclusions: Active myocarditis represents the pathological substrate of recent-onset cardiac involvement in SSc patients. Diagnosis through endomyocardial biopsy may have important therapeutic implications. The role of parvovirus B19 in determining both skin and cardiac damage in SSc patients remains to be clarified.