Predominant primitive right ventricular involvement in systemic sclerosis

Sirs,

Systemic sclerosis (SSc) is a connective tissue disease that primarily affects the vascular system (1). Cardiac involvement, a common occurrence in SSc, may affect any part of the heart (1, 2). Right ventricular (RV) alterations are usually assumed to be consecutive to respiratory insufficiency and/or pulmonary arterial hypertension (3, 4). Primary RV involvement is seldom reported (5), but may be underestimated (6).

We report the case of symptomatic RV cardiomyopathy in an SSc patient with only limited primary left ventricular (LV) involvement and normal pulmonary artery pressure.

A 50-year-old man presented in March 2006 with severe dyspnea. Diffuse cutaneous subset of SSc with positive anti-topoisomerase antibodies was diagnosed in June 2003.

Until hospital admission, he had been treated with nifedipine 20mg BID, aspirin 100mg/day, omeprazole 20 mg/day, methotrexate 15 mg/week with folic acid substitution in addition to prednisone 10 mg/day: therapeutic compliance was however non-optimal. In November 2005, he complained of mild dyspnea (class II of the New York Heart Association). An echocardiography was performed and showed normal LV (end-diastolic diameter (EDD) 44mm, ejection fraction (EF) 62%), but mild RV enlargement (33 mm - long axis parasternal view. N < 26mm) with functional moderate tricuspid insufficiency. Cardiac catheterization showed no pulmonary arterial hypertension; CT-scan showed only limited pulmonary fibrosis and routine spirometry was normal. Captopril 50 mg/day were added to nifedipine and he was encouraged to be extremely adherent to this treatment. However, he developed lower limbs oedema and was hospitalised for dyspnea in March 2006. Clinical examination revealed unchanged Rodnan's skin score at 28/51, heart rate 60 beats/min, blood pressure 120/70mmHg, leg oedema up to midthighs associated with spontaneous jugular swelling and a tricuspid regurgitation murmur. Blood count, creatinemia and electrolytes were within normal range; erythrocyte sedimentation rate was 14mm at the 1st hour. EKG was normal. Echocardiography demonstrated enlarged hypokinetic RV (36 mm - long axis parasternal view) and massive tricuspid regurgitation, with normal LV (EDD 48mm, EF 60%); no other abnormality was demonstrated. Cardiac catheterization showed a typical adiastole pattern, without pulmonary hypertension.

Radionuclide ventriculography confirmed reduced RVEF (40%). Routine spirometry and CT-scan results were unchanged.

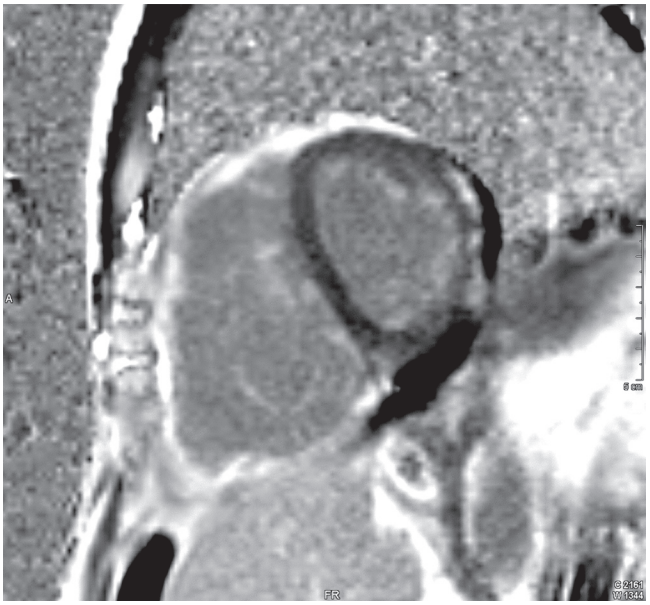

Fig. 1. Delayed contrast-enhanced cardiac MRI showing predominant RV fibrosis.

Lastly, a contrast-enhanced cardiac MRI showed diffuse transmural delayed enhancement of right myocardium and focal subepicardial delayed enhancement of the left myocardium suggestive of fibrosis, unrelated to coronay artery distribution (Fig. 1). Diuretics (furosemide 80 mg/day) were added to his treatment and he was discharged while still having symptoms. A 6-months follow-up period is uneventful, although the limitation persists.

The pathogenic mechanisms of cardiac lesions in SSc is complex; a growing body of evidence suggests that vascular system may be the primary target of the disease (1, 2). RV may have several origin, including primary myocardial involvement (from repeated ischemia-reperfusion injury, with or without accompanying structural vascular disease), and primary and/or secondary pulmonary hypertension associated with severe lung fibrosis (2-4). Our case-report may be singular because our patient had symptomatic RV cardiomyopathy in the absence of pulmonary hypertension or lung involvement that was associated to minimal LV involvement. This was confirmed by the MRI scan (7). It should be noted that these lesions have occurred in a SSc patient with the diffuse cutaneous subset and in the early period of the disease known to be at the higher risk for disease progression and severe complications.

In conclusion, symptomatic right cardiomyopathy may be a clinical feature of SSc. Primary cardiac involvement, a cardinal feature of SSc, may target both RV and LV separately or together. Our case report may reinforce the need to deeply investigate all patients for global cardiac involvement, since the earliest phase of the disease.

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