Left Ventricular Wall Motion Abnormalities in 80 Patients with Systemic Sclerosis

I. HEGEDŰS, L. CZIRJÁK*

Summary Segmental analysis of left ventricular wall motion was performed in 80 patients with systemic sclerosis by echocardiography. Half of the cases showed normal wall motion. Eighteen of the 57 patients with limited cutaneous systemic sclerosis (31.6%) exhibited hypokinetic wall motion abnormalities, while five of the 23 cases with diffuse cutaneous systemic sclerosis (21.7%) had similar findings. The three cases showing akinesis also belonged to the limited scleroderma subgroup. Our findings show that hypokinetic wall motion abnormalities can be detected in a remarkably high proportion of cases with limited cutaneous systemic sclerosis.

Key words Scleroderma, Systemic Sclerosis, Clinical Study, Myocardium, Heart.

INTRODUCTION

Systemic sclerosis (SSc) is characterized by fibrosis, capillary endothelial injury, and obliterative vasculopathy affecting the skin and certain internal organs including the heart. Various forms of cardiac involvement can be detected in SSc including pericardial disease (1-3), myocardial fibrosis (2,4), disturbances of the conduction system, impaired left ventricular systolic and/or diastolic function (3,5), asymmetrical left ventricular thickening or thinning (3,6) and the consequences of primary or secondary pulmonary hypertension (7). Cardiac symptoms are important in SSc because of their significant influence on survival (1,8,9).

Left ventricular wall motion abnormalities are well documented in patients with coronary diseases (10). No data have been available on the left ventricular segmental wall motion in SSc. In the present study, the wall motion abnormalities were investigated in SSc.

PATIENTS AND METHODS

Eighty patients with SSc were investigated. The female/male ratio of patients was 72/8. The mean age of the patients was 50.1±12.5 years (from 20 to 79). At the time of the investigation, all patients fulfilled the diagnostic criteria for SSc (11). Twenty-three cases belonged to the subset of diffuse cutaneous systemic sclerosis and the number of cases with limited cutaneous systemic sclerosis was 57 (12). The mean disease duration was 9±8 years. The clinical and laboratory data of patients were evaluated according to a standard protocol as previously described (13,14). Lung involvement was found in 59 (74%) cases, while oesophageal dysmotility was demonstrated in 36 (45%) patients. Twenty cases (25%) had sicca syndrome. Subcutaneous calcinosis was found in 6 (7%) patients. A positive antinuclear antibody test on HEp-2 cell was detected in 69 (86%) cases. The mean blood pressures of the patients and the controls were 130/82 and 126/84 Hgmm, respectively. Only three patients showed a mild hypertension during the period of this study. No patients with diabetes mellitus, hyperthyroidism and azotaemia were included in this study. All drug administration was stopped 16 hrs. before the examination. The investigation was not possible in three additional cases with SSc because of extensive skin involvement of the chest.

Forty-five healthy control individuals were also investigated. The mean age of the controls was 47.2±8 years; the female/male ratio was 39/6. No significant difference was detected by Student's t test between the mean age of patients and controls.

A Hitachi EUB 151 equipment (Japan) with 3.5 MHz and Pedoff transducers was used for this investigation. M mode, 2D and doppler echocardiography were performed as previously described (3). The same investigator (I.H.) evaluated all the echocardiography. Segmental wall motion was recorded on videotape and blindly...
evaluated. The left ventricle was divided into 12 segments (10). Each segment was separately evaluated. The following scores were used. Dyskinetic wall movement: 0, akinesis: 1, hypokinesis: 2. A normal segmental motion received a score of 3, while hyperkinetic wall motion was graded as a score of 4 (10). If any of the walls received a score of 0 or 1, the patient was categorized as having dyskinesis or akinesis, respectively. Five cases exhibiting hyperkinetic wall motion of one segment as a secondary consequence of the hypokinesis of the adjacent segment(s), were grouped with hypokinesis patients. Hypokinesis was recognised with the total score of the 12 wall segments ≤ 34, while a total score ≥ 37 was categorized as hyperkinesis.

The blind reevaluation of the segmental wall motion abnormalities in all cases by the same investigator (I.H.) showed a 96% accordance with the previous investigation. Forty patients were blindly evaluated by another investigator with a correlation of 94%.

RESULTS

Normal wall motion (with a score of 3 in all 12 segments) was detected in 40 patients; 39 of the 45 controls also showed normal wall motion. No difference was demonstrated in the means of the scores between the patients with limited and diffuse systemic sclerosis (Table I).

Eighteen of the 57 patients with ISSc (31.6%) exhibited hypokinetic wall motion abnormalities, while five of the 23 (21.7%) cases with dSSc had a similar finding.

All three cases with akinesis belonged to the ISSc subgroup. The akinesis was observed in the posterior wall with a Q wave on II-III- aVF leads in the ECG. Hypokinesis was detected in 18 cases with ISSc (31.6%) and 5 patients with dSSc (21.7%) (Table I).

Hyperkinetic wall motion abnormality was found in 6 cases with dSSc (26.1%), though 6 patients with ISSc also had a similar finding (10.5%) (Table I). All the hypokinetic/hyperkinetic segments were exclusively found on the septal and posterior wall regions. Among the 12 cases with hyperkinesis, one showed an aortic regurgitation, four had mild anaemia which could be responsible for this abnormality. No apparent secondary cause of the hyperkinesis was revealed in the remaining seven patients. Four of these cases belonged to the dSSc subset (data not shown).

No statistically significant differences were found in any of these wall motion parameters between ISSc and dSSc groups by the Chi square test. Female and male patients developed similar abnormalities (data not shown). No correlation was found between myocardial wall motion parameters and the left ventricular thickness of the septum or the posterior wall (data not shown). No differences were found in the wall motion abnormalities of patients aged below 40 and above 40 years (data not shown). The mean age ± SD of cases with hypokinesis, normokinesis and hyperkinesis did not show any remarkable differences (49.8±9.1, 49.4±12.2, 52±9.6 years, respectively). The mean disease duration of patients with or without wall motion abnormalities was 10.2±9.9 and 8.4±7.1 years, respectively.

With regard to the findings causing secondary wall motion abnormality, pulmonary hypertension was found by a high velocity tricuspid regurgitation only in three patients. The velocity of yet flow was above 2.5 m/sec in these cases. One of these patients from the ISSc subset, exhibited a right ventricular hypertrophy on ECG and on echocardiography as well. A paradoxical septal and hyperkinetic left ventricular posterior wall motion was observed in this case. The two other patients showed a normokinetic wall motion. Both of them belonged to the dSSc subgroup and had an incomplete right bundle branch block on ECG. One patient with dSSc had a left bundle branch block with diffuse hypokinesia. Three patients with ISSc had a Q wave on II-III avL-leads, and an akinetic postero-inferior wall motion.

Pericardial involvement was detected in 23 cases; 14 patients (5 with ISSc and 9 with dSSc) showed the presence of pericardial fluid less than 100 ml in all of these cases. Only three of the patients with pericardial fluid had hypokinesia of the posterior wall. In nine patients, the pericardium was thickened behind the left ventricle and atrium. A posterior wall hypokinesia was observed in 2 cases, both of them had a compensated septal hypokinesia.

Forty-two patients showed an abnormality which may affect the wall motion including pericardial fluid or pericarditis, thickened pericardium, tricuspid valve regurgitation, conduction abnormalities (left or right bundle branch block), and/or increased right ventricular diameter (above 27 mm). Comparing the wall motion of these cases with the remaining 38 patients without these abnormalities, no differences were found in the wall motion scores (34.7±3.8 and 35.1±5.1, respectively). Among the 38 cases without any abnormality potentially causing secondary wall motion disturbance, 11 patients (28.6%) (8 with ISSc and 3 with dSSc) showed a hypokinesia; also 28.6% of the 42 cases had hypokinesia.

No difference was found in the wall motion scores between the patients showing asymmetrical thickening or thinning of wall segments (data not shown). Nineteen patients remain if we further exclude cases showing a left ventricular asymmetrical thickening or thinning (3). Among these 19 cases, five (26.3%) had hypokinesia (one with dSSc and four with ISSc).