Mod Rheumatol (2007) 17:518–520
DOI 10.1007/s10165-007-0622-7

CASE REPORT

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Left ventricular aneurysms developed in a patient with systemic sclerosis

Received: April 16, 2007 / Accepted: June 18, 2007

Abstract A 60-year-old woman with systemic sclerosis, lung fibrosis, and primary biliary cirrhosis was admitted to our hospital because of heart failure. Ventricular aneurysms were found in the apex and the posterior wall of the left ventricle by angiocardiography; however, there was no sign of coronary artery stenosis. A myocardial biopsy specimen revealed diffuse focal myocardial fibrosis. In this case, the patient with systemic sclerosis developed a rare complication of ventricular aneurysms without coronary disease.

Key words Myocardial fibrosis · Systemic sclerosis

Case report

A 46-year-old woman consulted a doctor complaining of Raynaud's phenomenon and sclerodactyly. A laboratory examination and liver biopsy revealed elevated liver enzyme levels, positive antitopoisomerase I (22 U/ml) and antimitochondrial antibodies, and lymphocytes infiltration around the bile duct. She was diagnosed as having systemic sclerosis and primary biliary cirrhosis and started taking prednisolone 5 mg/day. She visited our hospital when she was 48 years old and was described as having diffuse cutaneous sclerosis over her face, trunk, and extremities, lower lung fibrosis, and slightly elevated levels of alkaline phosphatase (393 IU/l) and creatine kinase (192 IU/l). Since then, she has been taking prednisolone 10 mg/day and ursodeoxycholic acid 600 mg/day.

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She had been well and had no record of an anginal attack. When she was 60 years old, she complained of exertional dyspnea and was admitted to our hospital. Her chest X-ray showed cardiomegaly (chest–thoracic ratio was 76%). Neither she nor her family had ischemic heart disease or diabetes mellitus. Her plasma glucose level as needed was 94 mg/dl. Her body temperature was 36.5°C, blood pressure 184/98 mmHg, pulse 104 beats/min, and body weight 54 kg. She had slightly anemic conjunctiva and edematous legs. Systolic murmur was heard mostly at the apex area, which had been newly recorded in her medical chart 6 months earlier, and fine crackle was heard in the bilateral lower lung fields. The liver and spleen were enlarged by 1.5 cm below the right and left costal margins, respectively.

The laboratory findings showed mild pancytopenia (leukocyte count 3500/μl, erythrocyte count 403×10^6/μl, hemoglobin 11.7 g/dl, hematocrit 37.7%, and platelet count 5.2×10^6/μl) as a result of liver cirrhosis, and the C-reactive protein level was slightly elevated to 0.6 mg/dl. The aspartate aminotransferase level was 50 IU/l, the alanine aminotransferase level 23 IU/l, the alkaline phosphatase level 322 IU/l, the lactate dehydrogenase level 619 IU/l, the total bilirubin level 1.89 mg/dl, the direct bilirubin level 1.03 mg/dl, the creatine kinase level 220 IU/l, the blood urea nitrogen level 8.4 mg/dl, and the serum creatinine level 0.5 mg/dl. The atrial antidiuretic hormone was 81.2 pg/ml (normal range, less than 43.0 pg/ml), and the thyroid function tests were normal.

Sporadic supraventricular and ventricular premature contractions by electrocardiogram at rest and partially bigeminal pulse by Holter electrocardiogram were revealed, whereas neither ST-T change nor abnormal Q was detected (Fig. 1). Cardiac ultrasonography showed a thinning of myocardium and hypokinesis in most of the left ventricle and in part of the interventricular septum. Ventricular aneurysms were found in the apex and the posterior wall of the left ventricle. The ejection fraction lowered to 40.3%. In addition, the tricuspid and the mitral insufficiency and moderate pericardial effusion were found. Myocardial scintigraphy (^{201}Tl) showed a low uptake of radioisotope in part of the left ventricular posterior and anterior walls.

We suspected that she had developed cardiac aneurysms as a result of old myocardial infarction, and she began to receive oxygen. Diuretics, antihypertensives, and antiplatelets were administered orally, and digoxin was also administered because of paroxysmal atrial fibrillation. Cardiac insufficiency was improved while she reduced her body weight by 5 kg, and her chest thoracic ratio decreased to 66% from 76%. After she regained her health, left ventricular and coronary angiographies were performed. Left ventricular aneurysms were found in the apex and the posterior wall (Fig. 2A); however, neither coronary occlusion nor insufficiency was detected (Fig. 2B). A punch biopsy of the myocardium was performed from the right ventricle, which was echo- and radiographically normal. The specimen of the myocardium showed a diffuse random arrangement of small focal fibrosis. There was no other abnormality, such as a granulomatous lesion (Fig. 3).

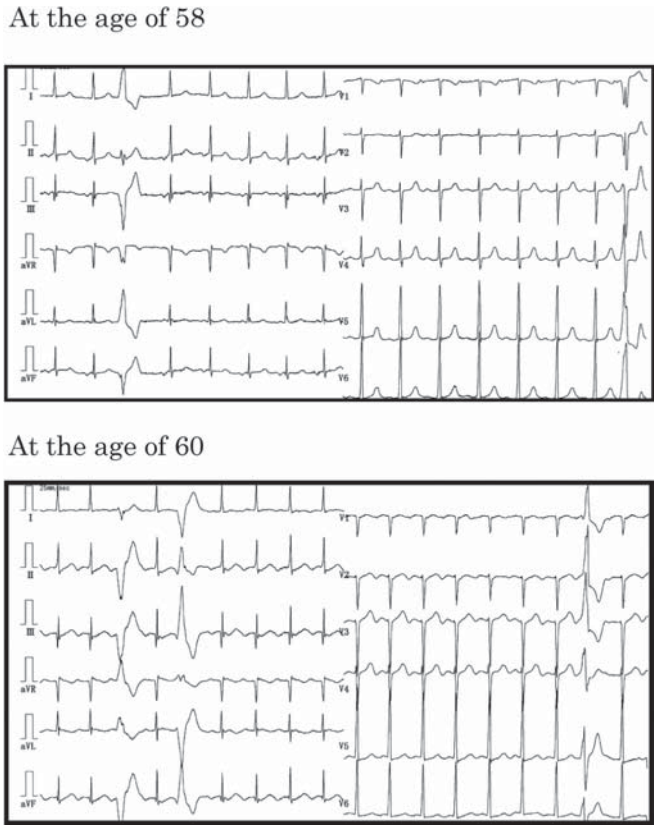

Fig. 1. Electrocardiograms at rest were recorded when the patient was 58 and 60 years old. Sporadic supraventricular and ventricular premature contractions were revealed, whereas neither ST-T change nor abnormal Q was detected

Discussion

We reported a woman with antitopoisomerase I antibody-positive systemic sclerosis associated with primary biliary cirrhosis and pulmonary fibrosis. She had developed cardiac insufficiency resulting from ventricular aneurysms.

In general, 20%–25% of patients with systemic sclerosis developed cardiac complications, including pericarditis, arrhythmia, and heart failure caused by akinesis or hypokinesis of the ventricular wall as a result of myocardial fibrosis.[1,2] Because cold stimulation in patients with systemic

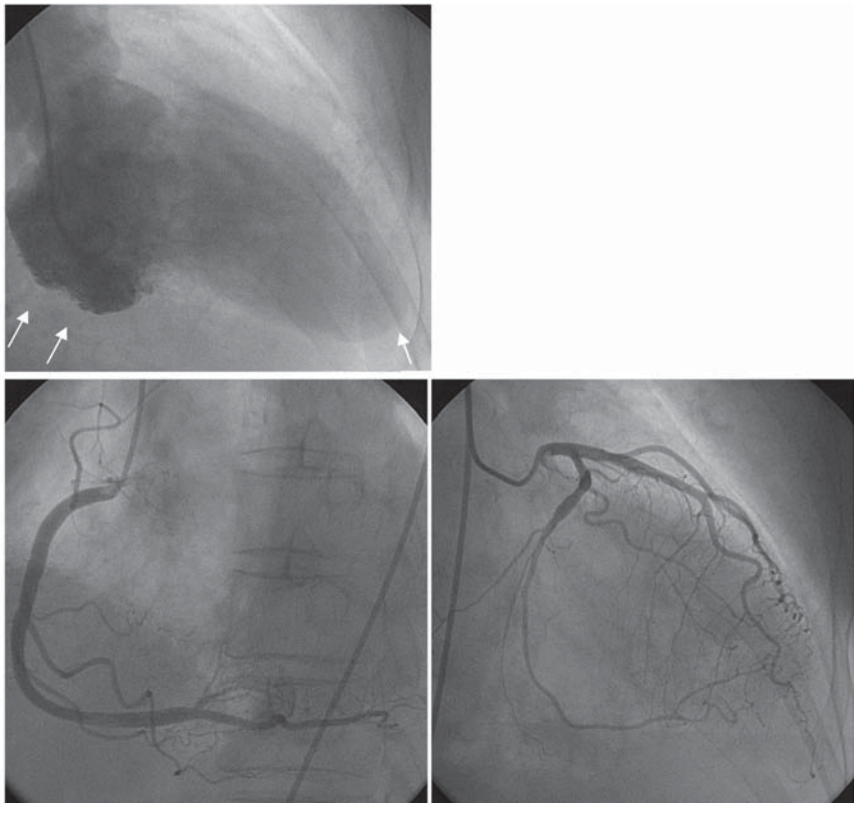

Fig. 2. **A** Left ventricular angiography revealed ventricular aneurysms in the apex and the posterior wall of the left ventricle (*arrows*), where the wall motion decreased. **B** Coronary angiography showed no significant stenosis of the coronary arteries

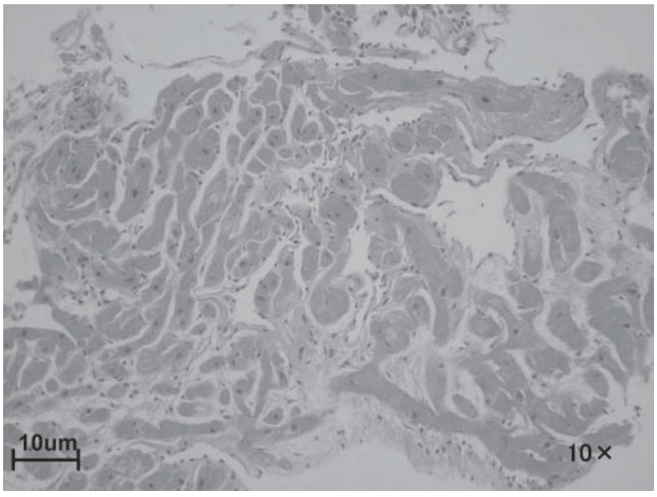

Fig. 3. A biopsy specimen of myocardium from the right ventricle, which was echo- and radiographically normal. There existed a diffuse random arrangement of small focal fibrosis in the myocardium. There was no other abnormality suggestive of sarcoidosis or cardiomyopathy (×100)

sclerosis caused lowering of blood perfusion and hypokinesis of the ventricular wall[3] and the duration of Raynaud's phenomenon correlated with heart failure,[4] repeated interruptions of myocardial blood perfusion may be responsible for the heart failure in patients with systemic sclerosis.

It was reported that about 80% of the autopsy cases had myocardial fibrosis that had gone without recognition along with the patchy fibrotic change spread over the myocardium that had no connection with the coronary artery distribution.[5,6] The development of myocardial fibrosis may be caused by microvascular spasms and reperfusion in the myocardium, which was named myocardial Raynaud's phenomenon.[2,5]

Cardiac aneurysms are usually caused by myocardial infarction, myocardial sarcoidosis, or idiopathic dilated cardiomyopathy. Our patient had no significant coronary artery stenosis, and a biopsy specimen from the right ventricle showed no special abnormality, such as granulomatous lesion, except fibrotic change. In spite of the intact coronary artery, the low uptake of radioisotope in myocardial scintigraphy suggests that she had myocardial fibrosis in parts of the left ventricular posterior and anterior walls as a result of myocardial Raynaud's phenomenon. There was only one previous report of a case of systemic sclerosis with a left ventricular aneurysm.[7] A 37-year-old man with systemic sclerosis died from heart failure, and the postmortem examination revealed a left ventricular aneurysm and massive myocardial fibrosis. The authors suggested that the aneurysm had been caused by the myocardial fibrosis.

In our patient, the cause of the left ventricular aneurysm is unknown. One possible explanation is that the myocardial Raynaud's phenomenon caused patchy myocardial fibrosis, and her high blood pressure caused the weak and fibrotic parts of the left ventricle wall to expand over the long term. Although patients with systemic sclerosis seldom develop ventricular aneurysms, we should note that underlying myocardial fibrosis is common and that it could lead to a fatal injury. Because it was reported that the duration of Raynaud's phenomenon correlated with heart failure,[4] patients with Raynaud's phenomenon should avoid cold stimulation and should be medicated with a vasodilator, which may prevent the spreading of myocardial fibrosis.

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