Acute Right Heart Failure and Achalasia-like Syndrome in a Patient with Limited Cutaneous Systemic Sclerosis and Primary Biliary Cirrhosis

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Abstract

We report a case of a 63-year-old woman who developed acute right heart failure and an achalasia-like syndrome with limited cutaneous systemic sclerosis (lcSSc) and primary biliary cirrhosis. Intravenous administration of diuretics improved her acute heart failure. Anti-centromere antibodies and anti-mitochondria antibodies were present. A coronary angiogram and a Swan-Ganz catheter revealed no abnormalities. Thallium-201 scan at rest demonstrated mild perfusion defects in both the apex and the anteroseptal and the inferior myocardium. A cine-esophagram revealed an achalasia-like syndrome. Though rare, physicians should be aware that some patients with lcSSc may develop acute right heart failure or achalasia-like syndrome.

Key words: systemic sclerosis, primary biliary cirrhosis, acute right heart failure, achalasia-like syndrome, limited cutaneous systemic sclerosis, thallium scan

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Introduction

Systemic sclerosis (SSc) is categorized into two subsets based on the extent of cutaneous involvement (1, 2). Diffuse cutaneous SSc (dcSSc) is characterized by generalized skin involvement and by more extensive visceral organ involvement early in its course and a poor prognosis. Patients with limited cutaneous SSc (lcSSc) have skin changes confined to the fingers or face and a protracted illness with a better prognosis, but later develop visceral organ involvement, such as pulmonary arterial hypertension or primary biliary cirrhosis (PBC). Anticentromere antibodies (ACA) are most frequently seen in patients with lcSSc, whereas antitopoisomerase I (Scl-70) antibodies are associated with dcSSc.

Cardiac involvement occurs frequently in Caucasian patients with SSc (3), but not so often in Japanese patients with SSc (4). Recently a case of acute congestive heart failure was reported in a Japanese patient with lcSSc (5). The esophagus is the most commonly affected internal organ in patients with SSc (6). However, achalasia-like syndromes are rarely seen. In this report, we describe a patient with lcSSc and PBC who developed acute right heart failure and an achalasia-like syndrome.

Case Report

A 63-year-old woman was admitted to our hospital with edema of the face and the extremities in February 2009. Ten years earlier, she first noticed Raynaud’s phenomenon in her fingers during the winter time, but took no medication. She had been diagnosed with diabetes at the age of 14 and was treated with insulin therapy for a few years, but had discontinued the treatment due to hypoglycemia. She had been diagnosed with diabetes at the age of 14 and was treated with insulin therapy for a few years, but had discontinued the treatment due to hypoglycemia. She had been diagnosed with diabetes at the age of 14 and was treated with insulin therapy for a few years, but had discontinued the treatment due to hypoglycemia. The marriages of her grandparents and parents were between first cousins. Her son was diabetic and her daughter had acute leukemia. On admission, her height was 151.8 cm; body weight 47 kg. Blood pressure was 150/90 mmHg; pulse rate 92/min with an irregular rhythm. Facial edema as well as pitting edema on the lower and upper extremities was observed.

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Figure 1A. Chest radiograph on admission showing a pleural effusion on the right side.

Figure 1B. Electrocardiogram on admission showing tachycardia and supraventricular premature beats.

however neither rales nor heart murmur could be detected during auscultation. Her bilateral fingers and hands were sclerotic and edematous and nail-fold hyperkeratosis was observed. Intravenous administration of diuretics improved her right heart failure, while insulin treatment lowered her blood glucose. After discharge her diuretics were changed to β-blockers because of tachycardia and premature beats. In April she complained of nausea, vomiting and an abdominal distension and visited an emergency clinic two times. Thereafter, she suffered recurring diarrhea and constipation. In May her facial edema and pitting edema developed again and she was readmitted to our hospital.

A chest radiograph showed a pleural effusion on her right side (Fig. 1A). An electrocardiogram revealed tachycardia and supraventricular premature beats (Fig. 1B). A coronary angiogram revealed no organic stenosis. A left ventriculogram and an echocardiogram showed no abnormalities. A Swan-Ganz catheter was intravenously inserted and revealed the following findings: mean right arterial pressure, 2 mmHg; pulmonary arterial pressure, 15 (25/8) mmHg; and pulmonary wedge pressure, 5 mmHg. Single photon emission computed tomography (SPECT) using thallium-201 at rest demonstrated a mild perfusion defect in the apex, the anteroseptal and the inferior myocardium of the left ventricle (Fig. 2). The right ventricular wall was normally visualized. There were no findings which suggested pulmonary congestion. A Gastrografin cine-esophagram revealed an achalasia-like syndrome despite the absence of dysphagia (Fig. 3). Five days later Gastrografin still remained in the colon, suggesting severely delayed intestinal peristalsis.

The laboratory findings were as follows: white blood cell count, 5.3×10^3/μL; hemoglobin level, 13.6 g/dL; erythrocyte sedimentation rate, 37 mm/h; C-reactive protein, 0.14 mg/dL; aspartate aminotransferase, 35 IU/L; alanine aminotransferase, 40 IU/L; lactate dehydrogenase, 185 IU/L; total bilirubin 0.71 mg/dL; alkaline phosphatase, 1,804 (normal: 120-340); γ-glutamyltranspeptidase, 350 IU/L; blood glucose 337 mg/dL; hemoglobin A1c, 9.0%; postprandial C-peptide, 1.15 ng/mL; total cholesterol, 179 mg/dL; high density lipoprotein cholesterol, 69 mg/dL; triglyceride, 156 mg/dL; BUN, 10.3 mg/dL; creatinine 0.29 mg/dL; immunoglobulin G, 1,946 mg/dL; immunoglobulin M, 380 mg/dL. No presence of HBs antigens and HCV antibodies were detected. γ-Aminobutyric acid antibodies were not present. Anti-nuclear
antibodies were present in a titer of greater than 1,280x with a centromere staining pattern, while ACA was present in a titer of 179 U/mL. Anti-Scl-70 and anti-RNP antibodies were both absent. Anti-mitochondrial antibodies were present in a titer of greater than 160x. There were no abnormal findings in urinalysis.

She was diagnosed with lcSSc and atypical PBC. Diuretics and 20 mg/day of prednisolone improved acute heart failure. PBC was treated with ursodeoxycholic acid and bezafibrate. She had no diabetic microangiopathies despite a long duration of diabetes.

**Discussion**

The overt cardiac manifestations in SSc, including pericarditis, conduction problems, and congestive heart failure, are recognized to be poor prognostic factors (3). Primary myocardial involvement without systemic or pulmonary hypertension and without significant pulmonary or renal disease is common in SSc and is related to repeat focal ischemic injury, causing irreversible myocardial fibrosis. The present case had a long history of diabetes, thus it is possible that coronary atherosclerosis due to diabetic complications caused acute right heart failure. However, it was not the case, as she had normal coronary arteriograms. Renal involvement or systemic and pulmonary arterial hypertension can also be excluded, as mentioned above. Since the Thalium-201 SPECT demonstrated mild perfusion defects, we suspect that this patient suffered intermittent microvascular dysfunction (myocardial Raynaud’s phenomenon) frequently at cold temperatures, primarily in the winter and early spring, thereby, developing cardiac dysfunction and acute heart failure. However, since we performed her echocardiography and ventriculography at room temperature after the improvement of her acute heart failure, we could not detect abnormal cardiac function. Tachycardic supraventricular
premature beats may have partly contributed to her heart failure. Calcium channel blockers and ACE inhibitors have been shown to improve cardiac microcirculation for some patients with lcSSc (7), but we could not prescribe them, because her systolic blood pressure dropped to around 100 mmHg after diuretic therapy.

Cardiac involvement occurs frequently in Caucasian patients with SSc (3), but rarely in Japanese patients (4). Early and widespread subclinical cardiac dysfunction was documented in many cases of either type of SSc in Caucasian patients using Thallium-201 scans (8). On the other hand, perfusion defects and large areas of induced ischemia was not so common among Japanese patients with SSc in gated SPECT studies (4), whereas significant diastolic abnormalities were shown even in patients with normal perfusion and systolic function. The severity of these abnormalities was related to the severity of SSc. Though rare, the present case suggests that some patients with lcSSc may experience cardiac manifestations, although for the majority, cardiac abnormalities, where present, will remain subclinical. Thus, a thallium scan should be performed even in early stages of lcSSc in order to detect cardiac abnormalities.

Gastrointestinal involvement occurs in most patients, and there is little difference in the frequency or type of problems between lcSSc and dcSSc (1). Distal esophageal motor dysfunction is the most frequent finding and leads to distal dysphagia. A cine-esophagram may show decreased peristalsis in the distal two-thirds of the esophagus. The lower esophageal sphincter musculature is similarly affected and reflux of gastric contents into the distal esophagus is common. However, an achalasia-like syndrome as observed in the present case is rarely seen and only one case has been reported in a patient with CREST syndrome (9). Thus, physicians should be aware of lcSSc as a rare cause of an achalasia-like syndrome.

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