Scleroderma and Mitral Stenosis: A New Cardiac Involvement or an Association by Chance

Ahmadreza Zarifian,¹ Alireza Sepehri Shamloo,¹ Mohammad Sobhan Sheikh Andalibi,¹ Hamid Hoseinkhah,² Freshteh Ghaderi,³ Morteza Khaki,⁴ Aliasghar Moeinipour²

1. Mashhad University of Medical Sciences, Mashhad, Iran
2. Department of Cardiovascular Surgery, Atherosclerosis Prevention Research Center, Imam Reza Hospital, Mashhad University of Medical Sciences, Mashhad, Iran
3. Department of Cardiology, Atherosclerosis Prevention Research Center, Imam Reza Hospital, Mashhad University of Medical Sciences, Mashhad, Iran
4. Department of Internal Medicine, Mashhad University of Medical Sciences, Mashhad, Iran

Corresponding author:
Aliasghar Moeinipour (Assistant Professor of Cardiovascular Surgery, Imam Reza Hospital, Mashhad University of Medical Sciences, Mashhad, Iran)
Postal code: 9137913316
Fax: (+98) 511-38525307
Mobile: (+98) 915310827
Email: moinipoora1@mums.ac.ir

Introduction
Systemic Sclerosis (SSc) is an autoimmune disorder with unknown etiology, which presents with vascular lesions and fibrosis of the skin and internal organs. Cardiac, renal, and pulmonary involvements are major causes of death in systemic sclerosis. Although a major cause of scleroderma deaths is cardiac failure, scleroderma rarely causes valvular disease. We report the case of a 48-year-old woman with severe mitral stenosis who was diagnosed with scleroderma 6 months before. Echocardiographic assessments had revealed no clinical manifestation of heart involvement in her previous visits. We used cardiac surgical treatment with successful outcome. The present case confirms the data from the currently available literature, indicating that mitral valve involvement in systemic sclerosis is a rare occurrence. Awareness of this uncommon association and adequate management to prevent complications of the underlying disease are prerequisites for successful mitral valve repair or replacement in such patients.

Keywords: Systemic sclerosis, Mitral stenosis, Cardiac involvement, Case report

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Highlights
Systemic Sclerosis (SSc) is an autoimmune disorder with unknown etiology, which presents with vascular lesions and fibrosis of the skin and internal organs. Cardiac, renal, and pulmonary involvements are major causes of death in systemic sclerosis. Although a major cause of scleroderma deaths is cardiac failure, scleroderma rarely causes valvular disease. We report the case of a 48-year-old woman with severe mitral stenosis who was diagnosed with scleroderma 6 months before. Echocardiographic assessments had revealed no clinical manifestation of heart involvement in her previous visits. We used cardiac surgical treatment with successful outcome. The present case confirms the data from the currently available literature, indicating that mitral valve involvement in systemic sclerosis is a rare occurrence. Awareness of this uncommon association and adequate management to prevent complications of the underlying disease are prerequisites for successful mitral valve repair or replacement in such patients.

Case Presentation
A 48-year-old homemaker was referred to the Rheumatology ward of Imam Reza Hospital, due to arthralgia, fatigue, myalgia, and dyspnea. She had 6 months previously presented to the Rheumatology ward with symptoms of fatigue, arthralgia in both wrists, and myalgia, and after a complete work-up, she was diagnosed with SSc. She also had a history of diffuse thickening and hardening of the skin, frequent digital ulcers, pitting of the fingertips, and a history of Raynaud’s phenomenon. Furthermore, test results for antinuclear antibody (ANA), antitopoisoioserase I (anti-Scl-70), anticientromere (ACA), and anti-RNA polymerase III have been reported, all of which have been published in the last three decades [5-8]. Herein, we aimed to present the first Iranian case of severe mitral stenosis in a patient with scleroderma, who had no clinical manifestation of cardiac involvement.

* Corresponding author. E-mail: moinipoora1@mums.ac.ir

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antibody were positive. Physical examinations and paraclinical tests for simultaneous gastrointestinal, pulmonary, neuromuscular, renal, cardiac, and musculoskeletal disease were negative at the time of diagnosis. She was discharged with daily prednisolone tablet (PO, 30 mg), and monthly cyclophosphamide (IV) for 6 months. The patient stopped using her medications 4 months after discharge due to relative feeling of well-being and after a month, her symptoms recurred. A month after she stopped her medications, she came to the emergency department with a relapse of SSc (arthralgia in both wrist and ankles, fatigue, and myalgia). She underwent a thorough diagnostic work-up again. New positive items in her history sheet were her recent admission, a 3% weight loss, and recurring symptoms of SSc during the month before. She gave no history of joint stiffness, loss of strength or sleep disorders, but pruritus, thickening and hardening of the skin, digital ulcers, and Raynaud phenomenon were positive. Although she had no sign of gastrointestinal, neuromuscular, renal, or musculoskeletal involvement, she complained of dyspnea (NYHA function class II) during the previous month. Her vital signs were stable and no fever was detected (blood pressure= 125/75 mmHg, heart rate=90, temperature=37.3°C axillary, and respiratory rate=14). Hyperpigmentation of the skin, especially over the face, legs and dorsal surface of the hands was discovered on physical exam. The skin was thin and pigmented over the face and lips were thin as well. She had several scars on the hands with blunted terminal phalanges and deformed nails. The skin was fixed over the fingers and thumbs, making them tightly stretched (Figure 1).

Multiple healed scars were observed on both of the lower limbs, up to the knee. Moreover, polyarthritis was seen in the wrists, ankles, proximal and distal interphalangeal joints in both sides along with decreased range of motion in both active and passive movements.

In cardiopulmonary evaluations, there was no deformity in the chest wall, jugular venous pressure was normal, and no bruit was auscultated over the carotid arteries. In cardiac auscultation, a loud mid-diastolic heart murmur was present in fifth intercostal space, at mid-clavicular line amplifying upon expiration. Fine crackles were detected at the base of lungs and there were no other abnormal findings on chest auscultation.

Laboratory findings showed normal values for serum electrolytes, except for serum iron=11 µmol/L and total iron-binding capacity (TIBC) = 401 µg/dl which were low and suggestive of iron deficiency. Other tests indicated normal levels of fasting plasma glucose, urea, and creatinine, besides low total bilirubin (0.5 mg/dl) with relatively high direct bilirubin (0.2 mg/dl). The measured concentration of transaminase enzymes was normal; however, alkaline phosphatase was increased (127 u/l). Elevated levels of lactate dehydrogenase (LDH), aldolase and creatine phosphokinase (CPK) (638, 10.6 and 858 u/l, respectively) might be suggestive of a myopathy.

Urine analysis and 24 hour urine testing revealed no abnormality and complete blood count (CBC) showed normal levels of white blood cell (WBC) count (6700/ml), but a low red blood cell (RBC) count (4.04 *106/µl) and a low hemoglobin content (9.3 g/dl) were detected, due to anemia.

Chest X-ray was normal, pulmonary function tests (PFT) showed a restrictive pattern, suggestive for scleroderma and chest computed tomography (CT) revealed a mild pleural effusion in left hemithorax and diffuse ground-glass opacities at the base of both lungs. In order to rule out cardiac involvement, a transthoracic echocardiography was requested, which showed severe MS with enlargement of the left atrium (figure-2), left ventricular ejection fraction of 45-55%, and pulmonary arterial pressure of 50 mmHg. Electrocardiography confirmed left atrial enlargement. The patient was referred to Cardiac Surgery ward for mitral valve replacement (MVR). She underwent MVR and
the excised mitral valve was sent for pathological evaluation, which showed a hypertrophic valvulopathy. No post-operative complication was observed after a year of serial follow-ups.

Discussion
The cardiovascular system is commonly involved in scleroderma. Cardiac involvement is recognized as a poor prognostic factor in patients with SSC [4]. Pericardial disorders, myocardial fibrosis, and vascular defects such as microvascular stiffness and vascular vasoactivity, which lead to focal ischemia in myocardium, are reported to be the major etiologies for cardiac involvement in scleroderma [9,10]. Cardiac involvement in scleroderma can present as an occult disorder or with potentially fatal cardiac changes, which result in increased morbidity and mortality [11]. Vasospasm of the microvasculature may reversibly impair perfusion and function of the myocardium. However, the following structural coronary arteriolar lesions lead to irreversible abnormalities. Although scleroderma rarely causes primary valvular disease, small L-S vegetation like masses, aortitis, aortic regurgitation (AR), and mitral valve prolapse (MVP) have been reported [8].

We found left atrial enlargement, pulmonary arterial hypertension (PAH) and Raynaud phenomenon in our case. In a cohort study, it was suggested that the presence of PAH is a poor prognostic factor and is associated with higher mortality rate in patients with scleroderma, compared with those who had idiopathic PAH [12]. In a recent study, it has been reported that the prevalence of PAH ranges from 8 to 12% in patients with SSC [13].

Mitral stenosis or LV dysfunction results in augmented LA afterload which leads to LA enlargement, which is a prognostic factor for the severity of diastolic dysfunction [14]. It should be noted that age is also an important determinant of atrial volume. Diastolic dysfunction is associated with the severity of cutaneous involvement and it was seen in about half of patients who suffered from scleroderma, even in the absence of myocardial ischemia [15]. However, the prevalence of ventricular hypertrophy has been reported to be more than that of atrial enlargement [8, 16, 17].

Mitral stenosis is a rare event in patients with systemic sclerosis [11]. Autopsies have revealed that 38% of SSC cases have nodular thickening of the mitral valve. In addition, regurgitation and vegetation of the mitral and aortic valves, shortening of the chordae tendineae at the mitral valve and mitral valve prolapse have been reported [9]. Many studies have reported myocardial Raynaud’s phenomenon (MRP) in patients with scleroderma. Many studies have examined the effect of cold presser provocation on myocardial function and perfusion in SSC patients to investigate the hypothesis of cold-induced coronary vasospasm, but their results are not definite and the clinical significance of this hypothesis requires further investigation [18].

In conclusion, the relative shortage of published studies makes the current knowledge indicating that involvement of mitral valve in scleroderma is a rare condition. Awareness of this uncommon association and adequate management of the patients is of utmost importance in prevention of complications and preparation of prerequisites for a successful MVR surgery.

Conflicts of Interest
The authors declare that they have no conflicts of interest to declare.

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References