

A case of overlap syndrome (scleroderma and polymyositis) associated with the development of sudden chest pain due to myocardial damage

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Summary Myocardial injury with systemic sclerosis (SSc) causes pericarditis and arrhythmia, and polymyositis-induced muscle inflammation causes myocarditis. We report a rare case of overlap syndrome (SSc and polymyositis) who presented with sudden chest pain secondary to myocardial fibrosis. Although the etiology of chest symptoms in collagen disease was difficult to identify, cardiac magnetic resonance imaging (MRI) revealed not myocarditis but myocardial fibrosis in our case. Synthetic judgement of serum brain natriuretic peptide/troponin T levels and cardiac MRI is useful in the search for the cause of chest symptoms even in patients with collagen diseases.

Keywords: Scleroderma, polymyositis, myocardial fibrosis, myocarditis

1. Introduction

Cardiac involvement is rare in patients with collagen diseases; however, this is a serious condition when it does occur. Myocardial injury with systemic sclerosis (SSc) causes pericarditis and arrhythmia, and polymyositis-induced muscle inflammation causes myocarditis (1,2). We report a rare case of overlap syndrome (SSc and polymyositis) who presented with sudden chest pain secondary to myocardial fibrosis.

2. Case Report

A 20-year-old man presented with a 3-year history of Raynaud's phenomenon and fingertip ulceration in winter. He noticed muscle weakness 3 months prior to presentation to our hospital. Physical examination revealed pitting scars and skin sclerosis (modified Rodnan's total skin thickness score, 16). Manual muscle testing revealed normal results; however, we observed decreased grip strength (10 kg [right], 8 kg [left]) and myalgia in both thighs. Laboratory investigations revealed the following results: white blood cell count

5,200 cells/mm³, serum creatine kinase 763 IU/L (59-248 IU/L), aldolase 15.3 IU/L (2.7-7.5 IU/L), anti-Smith antibody 3.6 IU/mL (< 10 U/mL), anti-U1 ribonucleoprotein antibody > 550 IU/mL (< 10 U/mL). Biopsy of the skin of the forearm showed edematous and increased collagen fibers in the dermis, and these findings were compatible with SSc (Figure 1A). Muscle biopsy of the biceps brachii revealed dense and diffuse perivascular lymphocytic infiltration (Figure 1B). Chest computed tomography (CT) revealed mild interstitial pneumonia in the basal segments of the lungs. We diagnosed him with overlap syndrome (diffuse cutaneous SSc and polymyositis) and treated with oral prednisolone (0.5 mg/kg/day, 35 mg/day). On the first day of oral steroid therapy, he developed sudden chest pain and dyspnea. Electrocardiography (ECG), echocardiography (UCG), and coronary CT revealed no abnormalities. However, the serum troponin T level was 0.0427 ng/mL (< 0.014 ng/mL), and serum brain natriuretic peptide (BNP) was 55.9 pg/mL (< 18.4 ng/mL). Cardiac magnetic resonance imaging (MRI) revealed that the extracellular volume fraction (ECV) was 35% (normal range 20-30%), and the value of native T1 mapping at the left ventricular wall was 1,320 ms (normal range approximately 950 ms). These findings indicated significant myocardial fibrosis (not myocarditis). Based on the aforementioned findings, we concluded that his chest symptoms were secondary to myocardial fibrosis caused by SSc. This symptom

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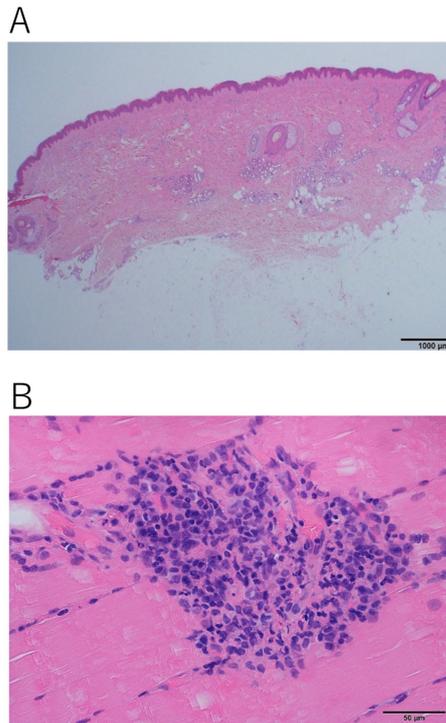


Figure 1. (A) Swollen and increased collagen fibers in the dermis. (B) Dense and diffuse infiltration of lymphocytes around vascular wall in the muscle.

disappeared spontaneously within 4 hours. Follow-up cardiac MRI performed 9 months later showed improved ECV and native T1 values.

3. Discussion

Serum BNP and troponin T levels are useful predictors

of myocardial injury in patients with SSc (3,4). Our patient showed the elevation of both serum BNP and troponin T levels. To our knowledge, this is a rare case report that describes overlap syndrome (SSc and polymyositis) with sudden onset of chest pain secondary to myocardial fibrosis. The exact etiology of chest symptoms (SSc and/or polymyositis) was difficult to identify because ECG and UCG may not accurately identify abnormalities in conditions that may be clinically indistinguishable, as was observed in our patient. We dermatologists must recognize that synthetic judgement of serum BNP/troponin T levels and cardiac MRI is useful in the search for the cause of chest symptoms even in patients with collagen diseases.

References

1. Gabrielli A, Avvedimento EV, Krieg T. Scleroderma. *N Engl J Med.* 2009; 360:1989-2003.
2. Zhang L, Wang GC, Ma L, Zu N. Cardiac involvement in adult polymyositis or dermatomyositis: a systematic review. *Clin Cardiol.* 2012; 35:686-691.
3. Ivanovic BA, Tadic MV, Zlatanovic MM, Damjanov NN, Ostojić PM, Bonaci-Nikolic BN. Which factors impact myocardial function in systemic sclerosis? *Echocardiography.* 2012; 29:307-317.
4. Bosello S, De Luca G, Berardi G, Canestrari G, de Waure C, Gabrielli FA, Di Mario C, Forni F, Gremese E, Ferraccioli G. Cardiac troponin T and NT-proBNP as diagnostic and prognostic biomarkers of primary cardiac involvement and disease severity in systemic sclerosis: A prospective study. *Eur J Intern Med.* 2019; 60:46-53.

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