Case Report

A case of sine scleroderma with parenchymal lung disease

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Abstract

Systemic sclerosis sine scleroderma is a subtype of scleroderma, which is characterized by involvement of visceral organs, but no characteristic skin alteration. The involved organs could be kidneys, heart, gastrointestinal system, and lungs. Interstitial lung disease (ILD) is one of the pulmonary manifestations of sine scleroderma. We report a 38-year-old woman presenting with chill, fever, generalized malaise, dyspnea on exertion, and dry cough with a history of Raynaud's phenomenon, who was evaluated by physical examination, spirometry, and computed tomography scan, that all lead to the diagnosis of ILD. Combination of high-titer positive anti-nuclear antibody, high erythrocyte sedimentation rate, positive C-reactive protein, and ILD could be explained by sine scleroderma.

Key Words: Interstitial lung disease, systemic sclerosis sine scleroderma, rituximab

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INTRODUCTION

Systemic sclerosis (SSc), also known as scleroderma, is an autoimmune disease that is characterized by inflammatory process and exuberant fibrosis that involves skin and internal organs. It is divided into subgroups by extension of cutaneous and visceral involvement.[1] SSc sine scleroderma is a rare form of SSc without cutaneous alteration, but may involve heart, lungs, and gastrointestinal system disease, which make the diagnosis difficult.[2] Interstitial lung disease (ILD) is a group of heterogeneous disease that is characterized by chronic inflammation and fibrosis of the interstitium and lung parenchyma, it can be associated with sine scleroderma.[3,4] We describe here a patient with ILD and Raynaud's phenomenon as the initial and sole presentation of sine SSc.

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CASE REPORT

A 38-year-old, house keeper woman was admitted in Alzahra hospital because of chills, fever, and generalized malaise. She also complained about exertional dyspnea and nonproductive cough of 2 years duration and had a history of Raynaud's phenomenon that started 8 months prior to admission. She was a nonsmoker with no documented lung disease and denied any other symptom. Physical examination revealed Velcro-like rales at the bases of both the lungs, but no sign of skin nail involvement or heart failure. Tuberculosis and other infections were ruled out. Laboratory tests showed a high positive titer of anti-nuclear antibody, erythrocyte sedimentation rate of 73, and C-reactive protein of 3 + and a normocytic normochromic anemia, whereas test for anti-Scl70 antibody and anti-centromere antibody was negative [Table 1].

Computed tomography scan showed bilateral ground-glass pattern suggestive of ILD [Figure 1].

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Table 1: Laboratory data of our patient

WBC: 8000 (4000-10,000)	ESR: 73 mm/h (3-15)
Hb: 8.4 g/dl (12–16)	CRP:+++
Plt: 373,000 (150-450,000)	ANA: 40 (10)
AST: 24 U/L (12-38)	Anti-SCL70: WNL
ALT: 20 (7-41)	Anti-centromere: WNL
ALK-P: 173 IU/L (64-306)	U/A: Normal
Cr: 0.8 mg/dl (0.5-1.5)	
BUN: 21 mg/dl (8-24)	

ALT: Alanine aminotransferase, ALK-P: Alkaline phosphatase, ANA: Anti-nuclear antibodies, AST: Aspartate transaminase, BUN: Blood urea nitrogen, CRP: C-reactive protein, ESR: Erythrocyte sedimentation rate, Hb: Hemoglobin, Plt: Platelets, WBC: White blood cells, WNL: Within normal limit

Spirometry was done, which showed a moderate restrictive pattern.

She was treated with prednisolone (1 mg/kg) and pulse of cyclophosphamide (1 g/month) for 6 months, and then put on Cellcept for another 6 months, but ILD did not improve. Thus, the patient was treated with rituximab 2 g (1 gram every 14 days) and showed gradual improvement, then she was subsequently treated with 100 mg of azathioprine daily, prednisolone 5 mg *bid*, and calcium plus vitamin-D. She has been followed up for 2 years and has not shown any other manifestation.

DISCUSSION

Sine scleroderma, a form of SSc without cutaneous involvement, has a highly variable presentation and course, which makes the disease unpredictable and difficult to diagnose and treat.[3]

Poormoghim *et al*., have suggested that the disease should be considered in the presence of the following features: (1) Raynaud's phenomenon or a peripheral vascular equivalent, such as digital pitting scars, digital tip ulcers, digital tip gangrene, or abnormal capillaries. (2) Positive ANA results (more convincing if the ANA specificity was due to an SSc-associated autoantibody). (3) Any one of the following: Distal esophageal hypomotility, small bowel hypomotility, pulmonary fibrosis, pulmonary arterial hypertension without fibrosis, cardiac involvement typical of scleroderma, or renal failure consistent with sclerodermic renal crisis, (4) No other connective tissue disorder or disease that would explain these findings.[5]

ILD is a wide range of acute and chronic pulmonary disorders capable of diffusely affecting the lung parenchyma with variable amounts of inflammation, fibrosis, and architectural distortion.[4] Fifteen percent

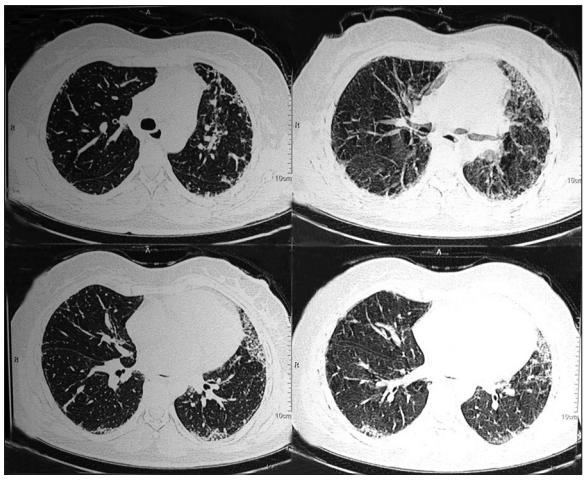

Figure 1: High-resolution CT scan showing lung fibrosis

of patients with ILD have an occult connective tissue disorder.[6] ILD may occur in > 80% of patients with sine scleroderma,[3] as the first or sole manifestation.[7]

The case presented here is an interesting one, because it is important to think about sine scleroderma in a patient with ILD and Reynaud's phenomenon, which can be confirmed by serologic tests, so that a proper diagnosis could be made and a proper treatment could be started.

CONCLUSION

Rituximab in resistant patients with sine scleroderma and ILD acts as an anti-fibrotic agent.

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