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Case report

Scleroderma-related interstitial lung disease

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ABSTRACT

Scleroderma-related interstitial lung disease (SSc-ILD) is a pulmonary fibrosing disorder characterized by systemic inflammation and progressive scarring of the lungs that leads to respiratory failure. Although certain immunosuppressive therapies may slow disease progression, current treatment strategies are not curative; consequently, SSc-ILD continues to be a major cause of morbidity and mortality. We present four cases of SSc-ILD that emphasize the importance of early screening and detection, close follow-up, and aggressive management. We also highlight the need for well-conducted clinical trials designed to identify new and effective treatments.

1. Introduction

Progressive Systemic Sclerosis or scleroderma is a systemic inflammatory disorder characterized by vascular damage, inflammation, and fibrosis of the skin and internal organs [1]. It was first described in 1752 by Dr Carlo Curzio in Italy as a disease that "turned the skin to wood" [2]. Since then, the prevalence and incidence of the disease has continued to rise with current literature estimating the prevalence of scleroderma at 18.4 per 100,000 [3]. In addition to its involvement of the skin and joints, scleroderma can also affect several organs including the lungs, kidneys and gastrointestinal system. Pulmonary hypertension and interstitial lung disease (ILD) are the most widely reported pulmonary complications that arise in scleroderma patients with a third of patients having pulmonary fibrosis [4]. The 5-year mortality for patients with progressive systemic sclerosis and ILD have a survival of 82–90% in 5 years, with ILD or pulmonary hypertension being responsible for 60% of the mortality observed in such cases [5]. Since the 1980's, successful treatment of scleroderma renal crisis has rendered pulmonary complications (e.g., pulmonary hypertension) the most common cause of death in these patients [6], with severe restrictive lung disease being present in 13% of patients with SSc [7].

Herein, we present four cases of SSc-ILD that emphasize the need for early screening and detection, close follow-up and aggressive management, and the desperate need for the development of safe and effective treatments capable of improving outcomes.

2. Case reports

2.1. Case 1

A 64-year-old African-American female carrying a diagnosis of scleroderma since 2007 was referred to our clinic in 2011 after she began to exhibit dyspnea on exertion and decreased exercise capacity. She denied cough and fevers, and had no significant weight loss. Her physical examination revealed a prominent P2 heart sound and coarse crackles at the left base. Initial pulmonary function tests (PFTs) showed a FEV1 of 2.18L (97% of predicted), FVC 2.53L (88% of predicted), TLC of 3.69L (71% of predicted), and a DLCOHb 37% of predicted. She underwent a high-resolution chest computer tomogram that showed reticulation, ground-glass opacifications, and traction bronchiectasis consistent with non-specific interstitial pneumonitis (Fig. 1A). She also had a transthoracic echocardiogram showing changes consistent with moderate pulmonary hypertension and tricuspid regurgitation, but preserved left heart function. Given her symptoms and objective findings of lung involvement, she was started on cyclophosphamide therapy

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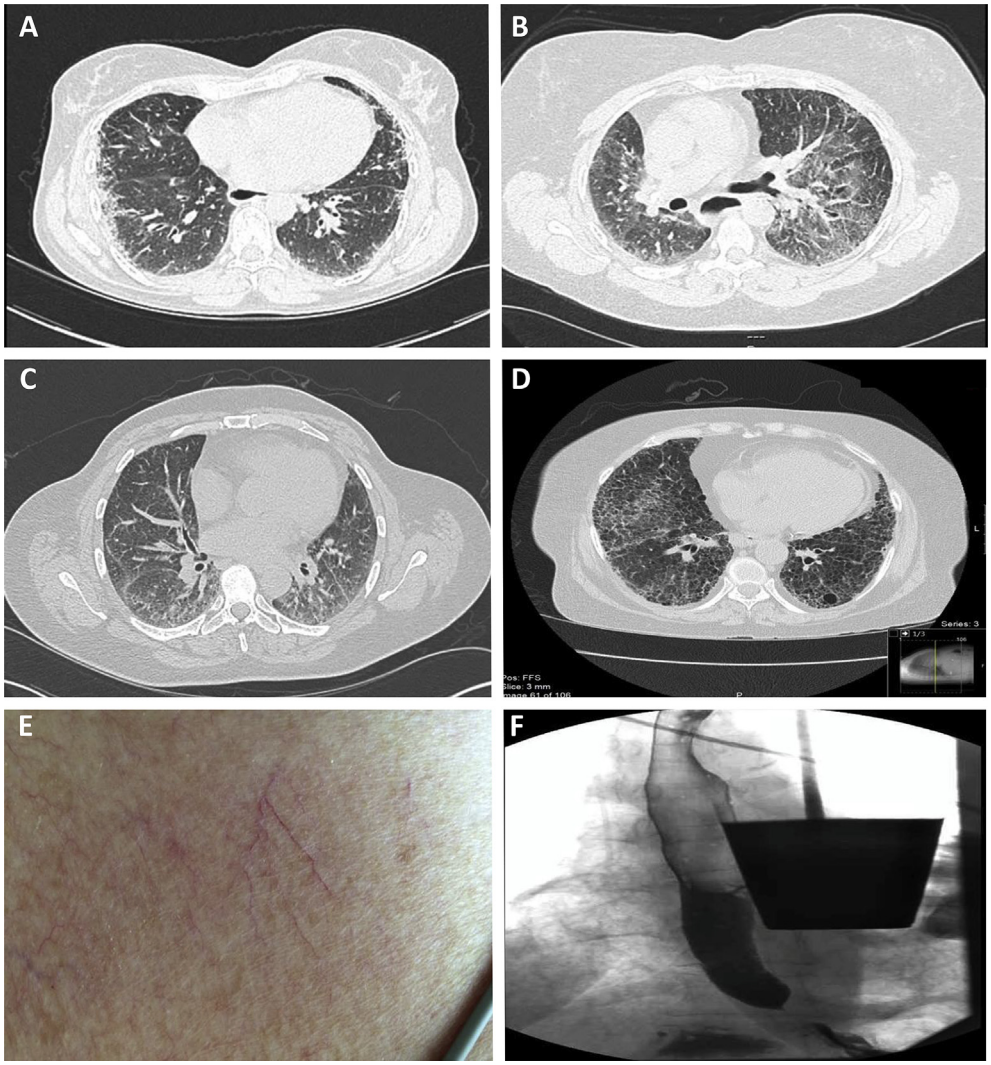

Fig. 1. Lung involvement in Ssc-ILD and related abnormalities. A, High resolution chest CT of patient of Case 1. B, High resolution chest CT scan of Care 2. C, High resolution chest CT scan of Care 3. D, High resolution chest CT scan of Care 4. E, Skin telangiectasia detected in Case 4. F, Barium swallow performed in Case 4 showing retention of fluid in esophagus.

and followed closely. She showed a mild improvement in her FEV1 over the next 2 years with little change in her FVC, and a gradual decline in her 6-min walk test (6MWT) decreasing from 391 to 330 m (74% predicted). During this time, she was also started on oxygen therapy at 2L per minute. Unfortunately, after 3 years of therapy she developed mild hematologic complications which triggered discontinuation of cyclophosphamide; this was followed by further decline in her lung physiology and symptoms. She is currently on prednisone with little effect on her clinical condition; other options are being considered.

2.2. Case 2

A 55-year-old Indian female presented with a 3-month history of dyspnea on exertion, cough, and skin tightness around her neck. Physical examination was positive for pulmonary bibasilar crackles. She underwent a high-resolution chest computer tomogram that was consistent with non-specific interstitial pneumonitis (Fig. 1B); initial serologies were negative with an ANA titer of 1:80. She was referred to Rheumatology for presumed scleroderma and the diagnosis was confirmed. In addition, this patient had a known history of uterine masses, and since scleroderma may represent a paraneoplastic syndrome [31], the immediate concern was the possibility of an underlying malignancy. She therefore underwent screening colonoscopy and mammography, which were normal, but was found to have uterine benign fibroids. A diagnosis of serology-negative Scleroderma was made and SSc-ILD. Further evaluation showed a decreased DLCOHb to 59% predicted and she was started on Mycophenolate Mofetil. Despite therapy, she continued to show a downward trend in lung physiology as well as worsening of her symptoms. She was then transitioned to cyclophosphamide, but continued to show deterioration of her lung physiology with worsening dyspnea on exertion. The patient was started on Tacilizumab (an interleukin-6 antagonist) and continues to be followed.

2.3. Case 3

A 60-year-old Caucasian female with a history of hypothyroidism presented with a 9-month history of worsening dyspnea on exertion following an upper respiratory tract infection. During her initial presentation, she also complained about swollen hands, skin discoloration, and symptoms consistent with Raynaud's phenomena. Her physical examination showed pulmonary bibasilar