Journal of Taibah University Medical Sciences (2016) 11(1), 77–81

Taibah University

Journal of Taibah University Medical Sciences

www.sciencedirect.com

Case Report

Scleroderma overlap syndrome with interstitial lung disease and pulmonary artery hypertension

Mohan S. Jangala, MD (General Medicine) [a], Venkata P.K.R. Lomati, Pharm-D [b,*] and Arvinda R. Purma, PhD [c]

[a] *Department of Medicine, Rajiv Gandhi Institute of Medical Sciences, Kadapa, India*
[b] *Department of Pharm-D, Rajiv Gandhi Institute of Medical Sciences, Kadapa, India*
[c] *Talla Padmavati College of Pharmacy, Warangal, India*

Received 9 May 2015; revised 7 October 2015; accepted 14 October 2015; Available online 25 November 2015

الملخص

تعتبر متلازمة تداخل تصلب الجلد حالة نادرة تظهر على شكل اضطرابين أو أكثر من اضطرابات النسيج الضام لنفس المريض. يرتبط بعض أمراض النسيج الضام، بأجسام مضادة محددة الملامح، بينما الأخرى لا يظهر معها أي ارتباطات مصلية. نعرض لحالة متلازمة تداخل تصلب الجلد تعاني من حمى، وضيق التنفس، وتليف رئوي، وارتفاع ضغط الدم الرئوي، ومرض الرئة الخلالي، وفقر الدم والعلامات السريرية للتصلب الجهازي والتهاب المفاصل الروماتيزمي. في نفس الوقت، أظهرت النتائج المخبرية إيجابية قوية للأجسام المضادة للنواة والأجسام المضادة توبوايزوميريز-1 وإيجابية مصلية للحمى الروماتيزمية والأجسام المضادة.

الكلمات المفتاحية: كالسينوزيز؛ تصلب الجلد الجلدي المنتشر؛ متلازمة التداخل؛ التهاب المفاصل الروماتيزمي؛ ارتفاع ضغط الدم للشريان الرئوي

Abstract

Scleroderma overlap syndrome is a rare condition that presents as two or more connective tissue disorders in the same patient. Some of these connective tissue diseases are associated with a specific autoantibody profile, while others do not show any serological association. We report a case of a patient with scleroderma overlap syndrome who presented with fever, breathlessness, pulmonary fibrosis, pulmonary hypertension, interstitial lung disease, anaemia and clinical manifestations of systemic sclerosis and rheumatoid arthritis. The patient's laboratory results were strongly positive for antinuclear antibody (ANA) and anti-topoisomerase-1 antibody and seropositive for rheumatic fever and CCP antibodies.

Keywords: Calcinosis; Diffused cutaneous scleroderma; Overlap syndrome; Pulmonary artery hypertension; Rheumatoid arthritis

Abbreviations: DcSSc, Diffused Cutaneous Scleroderma; RA, Rheumatoid Arthritis; ANA, Anti-Nuclear Antibody; RF, Rheumatoid Factor; SSc, Scleroderma; PF, Pulmonary Fibrosis; ILD, Interstitial Lung Disease; PAH, Pulmonary Arterial Hypertension; LcSSc, Limited Cutaneous Scleroderma

Introduction

Overlap syndrome is defined as entities satisfying the classification criteria of at least two connective tissue diseases (CTDs) occurring at the same or at different times in the same patient. CTDs include systemic lupus erythematosus, rheumatoid arthritis (RA), scleroderma (SSc), polymyositis/dermatomyositis and Sjögren syndrome.[1] The most common combinations are scleroderma with systemic lupus erythematosus, Sjogren's syndrome, rheumatoid arthritis and dermatomyositis.[2] Here, we discussed about a case of

* Corresponding address: Department of Clinical Pharmacy, Rajiv Gandhi Institute of Medical Sciences, Kadapa 516003, India.
E-mail: pavankumar.lomati@gmail.com (V.P.K.R. Lomati)
Peer review under responsibility of Taibah University.

ELSEVIER Production and hosting by Elsevier

 http://dx.doi.org/10.1016/j.jtumed.2015.10.004

overlapping features of rheumatoid arthritis and scleroderma along with interstitial lung disease and pulmonary artery hypertension.

Case report

A 55-yr-old agriculture labourer from the rural area of the Kadapa district of Andhra Pradesh was admitted to RIMS Medical College Hospital with the complaints of fever, cough and breathlessness for two weeks before admission as well as joint pains and stiffness of fingers from the previous 6 months. She had been diagnosed with scleroderma (lcSSc) for 2 yrs and had inconsistently been on medication. On appearance, she had a mild pallor, typical face (Figure 1), sclerodactyly (Figure 2), calcinosis cutis was present on the dorsum of the right foot (Figure 3) and wasting of the distal phalanges of the hand. Loss of joint range of motion, pain and stiffness of the metacarpophalangeal joints of her feet, as well as small joints (proximal interphalangeal and metacarpophalangeal) of both hands were effected.

On examination, her pulse rate was 82 beats/min, Blood pressure 150/90 mmHg and S_1 and S_2 were positive. Her respiratory rate was 20 cycles/min. Her oxygen saturation was SpO_2 90, and she had basal lung crepitations. On abdominal examination, she had mild hepatomegaly. Her urine output was normal. On cutaneous examination, skin thickening and stellate healed scars on the fingers and ankle were present, as well as stiffness, pain on motion and deformity changes of her hands.

Patient's laboratory and diagnostic data

Her reports showed nonreactive HIV, and the HBsAg was negative. A 2D-ECHO cardiogram report showed pulmonary artery hypertension with a mean pulmonary artery pressure of 30 mmHg. A chest X-ray showed a linear and reticular pattern, superimposed upon ground-glass attenuation in the basal region of the lung and mild cardiomegaly (Figure 4). The high-resolution computed tomography sections closer to the lung bases showed evidence of interstitial lung disease (predominantly subpleural in location) with septal thickening, bronchiectasis and fibrotic changes (Figure 5). Her sputum was negative for AFB. An abdominal ultrasound showed mild hepatomegaly, minimal ascites and renal parenchymal changes. A barium swallow examination showed moderate dilatation of 1/3 of the oesophagus with a poor progression of peristalsis (Figure 6). A renal Doppler showed that the main renal artery and segmental renal arteries were a normal coloured filling and exhibited a normal spectral wave pattern. Her

Figure 1: Photograph of the patient showing pinched nose and forte ring appearance of mouth.

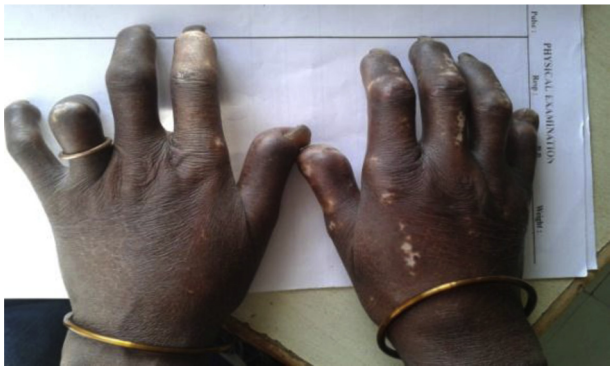

Figure 2: Photograph of hands showing stellate heeled scars on fingers and ankle.

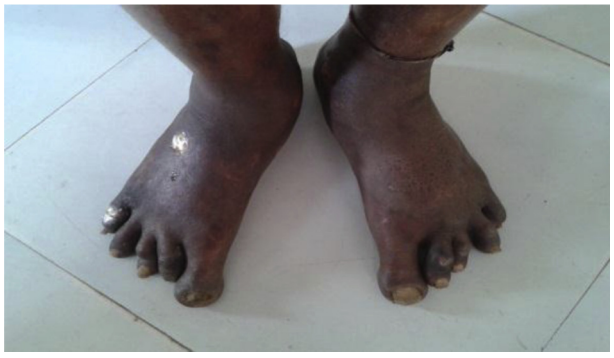

Figure 3: Photograph of the patient showing calcinosis cutis on legs.

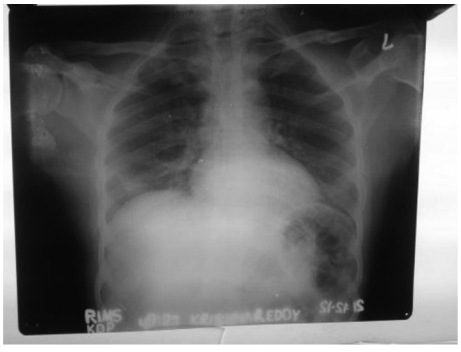

Figure 4: Chest X-ray shows that interstitial lung disease shows linear and reticular pattern, superimposed upon the ground-glass attenuation in the basal region of lung and mild cardiomegaly.