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A Rare Case of Systemic Sclerosis Sine Scleroderma

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Abstract
Systemic sclerosis (SSc) is an uncommon connective tissue disorder characterised by multisystem involvement, heterogeneous clinical manifestations, a chronic and often progressive course, and significant disability and mortality. Patients with SSc can be broadly grouped into diffuse cutaneous (dcSSc) and limited cutaneous (lcSSc) subsets defined by the pattern of skin involvement, as well as clinical and laboratory features. The constellation of calcinosis cutis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia, seen in some lcSSc patients, is termed the CREST syndrome. In some patients, Raynaud's phenomenon and other characteristic features of SSc occur in absence of skin thickening. This syndrome has been termed SSc sine scleroderma.

Keywords: *Raynaud's phenomenon, sclerodactyly, CREST syndrome, lcSSC, dcSSC, SSc sine scleroderma.*

Introduction

Systemic sclerosis (SSc) is an uncommon connective tissue disorder characterised by multisystem involvement, heterogeneous clinical manifestations, a chronic and often progressive course, and significant disability and mortality. Patients with SSc can be broadly grouped into diffuse cutaneous (dcSSc) and limited cutaneous (lcSSc) subsets defined by the pattern of skin involvement, as well as clinical and laboratory features. The constellation of calcinosis cutis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia, seen in some lcSSc patients, is termed the CREST syndrome. In some patients, Raynaud's phenomenon and other characteristic features of SSc occur in absence of skin thickening. This syndrome has been termed SSc sine scleroderma.

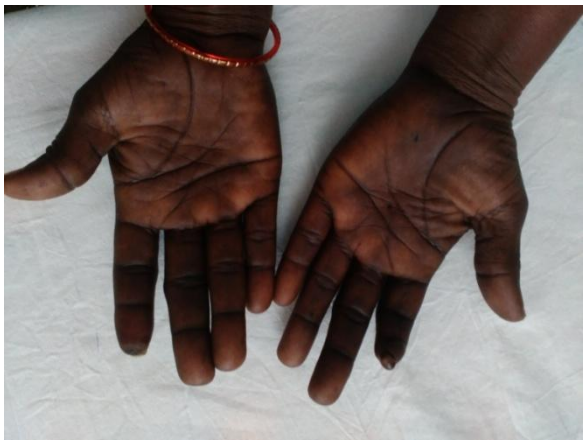
Case Report

A 40 year old female patient was admitted with complaints of ulcers at the tip of right index and middle fingers associated with pain and swelling over a period of 10 days. Past history of amputation of left distal phalanx 1 year back for similar complaint. She had a history suggestive of Raynaud's phenomenon. She was diagnosed as hypothyroid 5 years back with irregular treatment and with GERD 2 years back. General examination-normal except for presence of ulcers over right fingers and amputated distal phalanx of left index finger. Systemic examination respiratory system: Bilateral basal crepts on auscultation, CVS, CNS, GIT-normal. Her blood investigations revealed Hb-11g/dl,TLC-9800 cells/mm^3, normal hemogram, blood urea-24mg/dl, serum creatinine-1mg/dl, serum bilirubin-0.9mg/dl,T3-0.8ng/ml, T4-7.1µg/ml, TSH_10.63MIU/ml. urine analysis was normal, chest x-ray showed reticulo nodular

opacities, ECG was normal, 2D ECHO was normal, CT CHEST-showed bilateral ground glass opacities, paratracheal lymphadenopathy, mild thyromegaly, mild dilatation of esophagus. COLOUR DOPPLER of right upper limb showed normal study. ANA profile showed Anti scl-70 positive.

Discussion

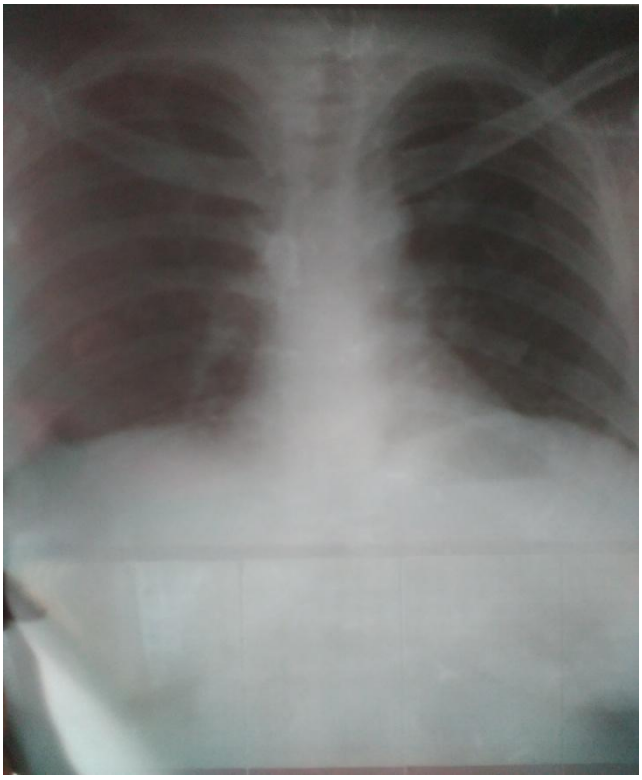
A 40 year old female who presented with painful digital ulcers with amputated distal phalanx without any skin thickening was investigated and found to have hypothyroidism, GERD, Interstitial lung disease with scl-70 positive antibodies comes under variant of SSC-SYSTEMIC SCLEROSIS SINE SCLERODERMA.



Patient was started on treatment with cyclophosphamide and bosentan. There was improvement in the condition with decreased pain.

Conclusion

Major organ involvement as part of SSc without the characteristic skin changes of scleroderma, defined as SSc sine scleroderma (ssSSc), was first described in 1954[1].



Systemic sclerosis sine scleroderma should be included in the spectrum of SSc with limited cutaneous involvement and should not be considered a distinct or separate disorder[2]. Clinical and serological features of ssSSc subjects were compared to limited (lcSSc) and diffuse cutaneous SSc (dcSSc) subjects[3]. Patients with ssSSc had a relatively mild disease with good prognosis[4].

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