

# Cutaneous manifestations of systemic sclerosis

Veeresh Dyavannanavar, Shashikant Malkud\*

Department of Dermatology, Gadag Institute of Medical Sciences, Mallasamudra, Gadag-582103, Karnataka, India.

\* Department of Dermatology, Mahavir Institute of Medical Sciences, Vikarabad-501102, Telangana, India.

## Abstract

**Background** Systemic sclerosis (SSc) is a multi-system disorder that affects skin and other organ systems. It is a disease of unknown etiology with an interplay between vascular and collagen maturation factors. It has varied mucocutaneous manifestations and immunological features that are helpful in initial diagnosis and subsequent classification of the disease.

**Objective** To study the mucocutaneous manifestations and immunological profile in patients of SSc.

**Methods** Thirty one patients of SSc attending to dermatology department over a period of 2 years were included in the study. Detailed history regarding age, sex, occupation, duration of complaints and evolution of the lesions were noted. Routine laboratory investigations, biochemical and immunological profile were done.

**Results** The common presenting features were Raynaud's phenomenon in 30 (96.7%), pigmentary changes in 29 (93.5%), sclerodactyly in 28 (90.3%), microstomia 25 (80.6%), fingertip ulceration and scarring in 18 (58.1%) and flexion contractures of the fingers in 10 (32.2%) patients. Mucosal and nail changes were observed in 8 (25.8%) and 10 (32.2%) patients respectively. Diffuse cutaneous SSc was noted in 17 (54.8%) and limited cutaneous SSc in 14 (45.2%) patients. Twenty (64.5%) patients tested positive for ANA.

**Conclusion** The study showed the spectrum of mucocutaneous and immunological profile of SSc patients in south Indian population.

## Key words

Diffuse, limited, sclerosis.

## Introduction

Systemic sclerosis (SSc) is a multisystem disorder that affects mainly skin, blood vessels, and muscles. The disease is more in females with a female to male ratio of 3:1.<sup>1,2</sup> It is a disease of unknown etiology and appears to be an interplay between vascular, autoimmune and

collagen maturation factors.<sup>3</sup> There are various cutaneous features like Raynauds phenomenon, pigmentary changes, fingertip scarring, sclerodactyly etc. which are helpful in making diagnosis. Based on the extent of skin involvement and immunological features it is subdivided into diffuse cutaneous systemic sclerosis (dcSSc) and limited cutaneous systemic sclerosis (lcSSc).<sup>4,5</sup> Till today only few studies have been conducted in India on SSc. Hence it was thought worth while to analyse the various cutaneous features and immunological profile in patients of SSc.

## Address for correspondence

Dr. Shashikant Malkud, Associate Professor,  
Department of Dermatology,  
Mahavir Institute of Medical Sciences,  
Vikarabad-501102, Telangana, India.  
Ph: +917353663101  
Email: smalkud@yahoo.com

## Materials and Methods

It was a hospital based cross-sectional study conducted in outpatient department of Dermatology, Venereology and Leprosy of Government Medical College hospital, Kottayam, Kerala. The study was conducted from January 2010 to January 2012. Study approval was taken from institutional ethical committee. Thirty one patients of SSc were included in the study. Patient selection was based on the American College of Rheumatology (ACR) criteria for SSc. Detailed history regarding age, sex, occupation, duration of complaints, precipitating factors, evolution of the cutaneous lesions, family history and drug intake were noted. Detailed cutaneous examination was also done. Laboratory investigations including complete hemogram, Erythrocyte Sedimentation Rate (ESR), C-reactive protein, random blood sugar, renal function tests, and liver function tests were done in all patients. Other tests like antinuclear antibody (ANA), anti Scl-70 antibody anti-centromere antibody, imaging of hand joints and lungs were done wherever necessary. Other investigations like histopathology and barium swallow were done wherever needed. Data was analysed using SPSS 16.0 version and presented as proportions.

## Definition

Patients were selected according to the ACR criteria for SSc.

### Major criteria

1. Skin thickening and induration proximal to the metacarpophalangeal or metatarsophalangeal joints.

### Minor criteria

1. Sclerodactyly
2. Digital pitting scars
3. Bibasilar pulmonary fibrosis

To fulfill the diagnosis of SSc, patient should have either one major or two minor criteria.<sup>6</sup>

## Results

Thirty one cases of SSc were included in the study. Out of 31 patients, dcSSc was noted in 17 (54.8%) patients and lcSSc in 14 (45.2%) patients. Most of the patients were in the fourth (32.3%) and fifth (41.9%) decades. The youngest patient was 24 years old girl and the oldest was 68 years old woman. The average age at diagnosis was 38.4 years. All patients were females. Twenty patients (64.5%) were engaged in household work, 5 (16.1%) patients in manual labour and rest 6 (19.4%) in other miscellaneous jobs. None of them were engaged in any occupation that predisposes to SSc. None had a family history of similar illness. Out of 31 patients, 27 (87%) were married and 4 (13%) were unmarried. The maximum duration at presentation was 15 years and minimum was 4 months.

Most of the cases had more than one symptom. Sclerosis of skin was noted in all 100% patients and Raynaud's phenomenon in 30 (96.7%) patients. Raynaud's phenomenon was preceded skin tightness in 26 (83.8%) patients and in 4 (12.9%) patients both appeared simultaneously. Pigmentary changes were noted in 29 (93.5%) patients of which, diffuse hyperpigmentation was seen in 8 (25.8%), localized pigmentation in 2 (6.4%) however salt and pepper pigmentation was in 19 (61.2%) patients (**Figure 1**). Swelling of hands and feet was noted in 21 (67.7%) patients, sclerodactyly in 28 (90.3%) patients, microstomia in 25 (80.6%) patients (**Figure 2**), mat-like telangiectasia in 11 (35.4%) patients,



**Figure 1** Salt and pepper pigmentation.



**Figure 2** Characteristic facies with microstomia.

fingertip ulcer/ scar in 18 (58.1%), digital gangrene in 2 (6.4%), cutaneous small vessel vasculitis in 2 (6.4%) patients and cutaneous calcinosis in 1 (3.2%) patient (**Table 1**).

Mucosal changes were noted in 8 (25.8%) cases, among these 6 (19.3%) patients had mucosal hyperpigmentation and 2 (6.4%) had oral candidiasis. Diffuse alopecia was noted in 11 (35.4%) patients. Nail changes were noticed in 10 (32.2%) patients of which shiny nails in 8 (25.8%), increased convexities in 4 (12.9%), clubbing of the fingers in 2 (6.4%), and periungual telangiectasia were seen in 1 (3.2%) patient. Flexion contracture of the fingers was noted in 10 (32.2%) patients and amputation of the digits in 2 (6.4%) patients.

Positive ANA was noted in 20 (64.5%) patients. Scl-70 antibody was positive in 12 (38.7%) patients and anti-centromere antibody in 8 (25.8%) patients (**Table 2**).

**Table 1** Distribution of various presenting features of systemic sclerosis (n=31).

Variables	Number	Percentage
Raynaud's Phenomenon	30	96.7
Cutaneous Pigmentation	29	93.5
Sclerodactyly	28	90.3
Microstomia	25	80.6
Finger tip ulcer/scar	18	58.1
Hand & feet swelling	21	67.7
Puffy hands	03	9.6
Nail changes	10	32.2
Mat like telangiectasia	11	35.4
Flexion contracture of digits	10	32.2
Diffuse alopecia	11	35.4
Mucosal changes	08	25.8
Amputation of digits	02	6.4
Small vessel vasculitis	02	6.4
Digital gangrene	02	6.4
Calcinosis cutis	01	3.2

**Table 2** Immunological features of patients (n=31).

Variables	Number	Percentage
ANA	20	64.5
Anti-centromere antibody	8	25.8
Anti- Scl -70 antibody	12	38.7

## Discussion

Cutaneous manifestations of systemic sclerosis (SSc) are important for the initial diagnosis of the disease and subsequent classification of disease into limited and diffuse category. All the thirty one patients satisfied the ACR (1980) criteria for the diagnosis of SSc.<sup>6</sup> The patients' age ranged from 24 to 68 years with majority of patients in the fourth (32.3%) and fifth decades (41.9%) of life. This is in concordance with study by Bhagavandas *et al.*<sup>7</sup> and Malviya *et al.*<sup>8</sup>

In our study all the patients were females. The female preponderance was noted in various Indian and Western studies.<sup>5,9</sup> Most of the patients in our study were housewives. It may be due to constant wetting of fingers. No relation to drugs and chemicals was seen in our study although some studies have indicated the role of certain chemicals in the evolution of SSc.

The skin is involved almost universally in SSc. Puffiness of hands, swelling and decreased flexibility of the joints are considered as the earliest finding in SSc. Puffiness of hands was found in 9.6% of patients in the present study. This is similar to the study by Sharma *et al.*<sup>1</sup> Raynaud's phenomenon was present in 96.7% of patients. This is slightly more than the study in Afro-Caribbean population.<sup>10</sup> However, a much lower frequency has been reported by Krishnamurthy *et al.*<sup>11</sup> and Gupta *et al.*<sup>9</sup> The higher incidence in our patients may be due to the fact that majority of patients were house wives or manual labours who had history of constant wetting of fingers in water.

Pigmentary changes constitute 93.5% in our study. Earlier studies have shown pigmentary changes ranging from 73.1 to 91%.<sup>1,11</sup> In our study salt and pepper pigmentation was more common, this is in concordance with study by Kumar *et al.*<sup>12</sup> In our study fingertip ulceration and scarring was seen in 58.1% of the patients. The prevalence of fingertip ulceration in previous studies was 52-70%.<sup>1,13</sup> Cutaneous calcinosis was seen in 3.2% of the patients but no case of CREST (calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia) syndrome was noted in our study. In our study flexion contractures of the hands were noted in 32.2% patients while Sharma *et al.*<sup>1</sup> found a higher incidence. Telangiectasia was reported in 35.4% of patients in our study this is similar to study by Gupta *et al.*<sup>9</sup> Nail changes were seen in 28.2% and mucosal changes in 21.7% of our patients.

Cutaneous sclerosis was seen in all patients. Similar finding was noted in previous Indian and Western studies.<sup>1,14</sup> In our study dcSSc was noted in 54.8% patients and lcSSc in 45.2% of the patients. This is in concordance with study by Teh *et al.*<sup>15</sup>, were dcSSc was more common (70%) than lcSSc (30%) but in contrast to study

by Malaviya *et al.*<sup>8</sup> 94.4% had lcSSc and 5.6% had dcSSC.

In present study positive ANA was noted in 64.5% of the patients. Previous studies have reported positive ANA in 56.8-89% of the patients.<sup>1,11,15</sup> Antibody Scl-70 was positive in 38.7% of cases & anticentromere antibody in 25.8% cases. Similar findings were noted in other studies.<sup>16,17</sup>

Small sample size is the limitation of the present study. A larger study is needed in Indian population to further highlight the mucocutaneous manifestations in SSc patients.

## Conclusion

Systemic sclerosis predominantly affects females. The most common presenting symptom was Raynaud's phenomenon followed by cutaneous pigmentation.

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