

# Systemic Sclerosis: A Clinical Analysis of Foot Findings in 52 Cases

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## Abstract

**Introduction:** The diagnosis of systemic sclerosis is mostly clinical and is identified by the various cutaneous changes. The cutaneous changes in systemic sclerosis are already well established and supported by a large number of studies and textbooks. Very few literatures describe foot findings in patients with systemic sclerosis. This study was aimed at identifying the various changes in plantar aspect of foot of patients with systemic sclerosis.

**Methods:** All patients diagnosed with systemic sclerosis were recruited. Apart from clinical examination, X – ray of both feet was done in all the patients.

**Results:** A total of 52 patients were recruited in the study period. Most of the patients (86.54%) were females and the mean age at presentation was 34.19 years. A history of Raynaud's phenomenon was present in 47(90.38%) patients. The foot findings were digital pitted scars in 44(84.61%) patients, macular hyperpigmentation in 40(76.92%) patients and corns in weight bearing areas were found in 40 (76.92%) cases.

**Conclusion:** In our study corn was a common finding in the foot of patients of systemic sclerosis which has not been routinely documented.

## Introduction

Systemic sclerosis is a multisystemic connective tissue disorder. Incidence of the disease is between 2.3 and 10 per million population<sup>1-3</sup> and the ratio of females to males is between 3 and 6:1.<sup>3-5</sup> Systemic sclerosis has been divided into the Limited cutaneous and Diffuse cutaneous subsets which are basically differentiated by the extent of skin sclerosis.<sup>6</sup>

The cutaneous changes are characteristic and are often first to manifest. The sclerosis of hands and face are most frequent, but it may extend to involve the forearms, upper arm, neck, trunk and lower limbs. The other commonly encountered cutaneous changes are the presence of

hide bound skin, finger tip pitted scars,<sup>7</sup> telangiectasia, hyperpigmentation and mottled pigmentation, calcinosis, dilated nail fold capillaries, ragged cuticles and sclerodactyly.<sup>[8]</sup> Rare findings include, pterygium inversum unguis like changes,<sup>9</sup> livedo reticularis<sup>10</sup> and lesions resembling acrokeratoelastoidosis.<sup>11</sup>

As seen above, the skin involvement in systemic sclerosis has been described very vividly in various literatures and textbooks but limited text is available focusing the sole findings. One report by Sari-Kouzel et al.<sup>12</sup> described various

changes in feet of systemic sclerosis patients. Apart from other common changes like colour changes in feet in response to temperature changes, pain related to cold, foot ulceration, pre-ulcerative lesions, toenail changes and calcinosis, they observed callosities in 80% of patients.

Our study was aimed to identify the various clinical findings in sole of patients with systemic sclerosis.

## Methods

A cross-sectional study was undertaken in the department of Skin & VD, in a tertiary care teaching hospital in Bhubaneswar, Odisha, for a period of six years from 2011 to 2016. All the cases diagnosed with systemic sclerosis according to the American College of Rheumatology criteria<sup>13</sup> were examined. After obtaining the informed consent a detailed history regarding the age of onset, duration of disease, occupation, family history and presence or absence of Raynauds phenomenon was obtained with detailed dermatological examination giving more importance to findings on the plantar aspect of feet. All cases with disease overlap syndromes, history of any lower limb bone or joint orthopaedic surgery within past 1 year, diagnoses of diabetes and loss of sensation on sole were excluded. A total of 52 cases were included in the study. Routine X – ray feet and fasting

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**Table 1: Clinical outcome of the study**

Parameters	No. of Patients (%) n=52
Age	
20 – 30 yrs	19 (36.54%)
30 – 40 yrs	18 (34.61%)
40 – 50 yrs	15 (28.85%)
Sex	
Male	7 (13.46%)
Female	45 (86.54%)
Associated Raynaud's phenomenon	47 (90.38%)
Foot findings*	
Digital pitted scar	44 (84.61%)
Macular pigmentation	40 (76.92%)
Corn	40 (76.92%)
Corn distribution n = 40	
Heel only	21 (52.5%)
Metatarsal heads only	5 (12.5%)
Both	14 (35%)

blood sugar was done in all patients along with neurological consultation to rule out the presence of any form of neuropathy.

## Results

Out of the total 52 cases of systemic sclerosis included in the study (Table 1), 19(36.54%) patients presented in their third decades of life, 18(34.61%) in fourth decade and 15(28.85%) in fifth decade. The mean age at presentation was 34.19 years. Most of the cases [45(86.54%)] were females. The mean duration at presentation of the disease was 26.9 months i.e. around 2 years in 50 patients except 2 patients who presented at 13 and 12 years of disease manifestation.

A history of Raynaud's phenomenon could be elicited in 47(90.38%) patients. Apart from other cutaneous findings of systemic sclerosis, the sole findings were digital pitted scars (Figure 1) in 44(84.61%) patients, macular hyperpigmentation in 40(76.92%) and corn (Figure 2) in 40(76.92%) patients. Corn was diagnosed based on absence of bleeding on pairing and eliciting tenderness on vertical pressure. The corns were distributed over weight bearing areas of soles like heel only in 21(52.5%) patients, metatarsal heads only in 5(12.5%) and both sites were affected in 14(35%) patients. There was no family history of systemic sclerosis or any familial tendency for corns and callosities in any of the patients. X-ray feet of all the patients having corn did not show any bony deformities predisposing to corn formation. Two patients had blood sugars in

**Fig. 1: Digital pitted scars**

higher borderline range, but were not associated with any neuropathy. The patients having multiple corns were advised local application of topical keratolytics (salicylic acid 17% + lactic acid 17%) and those with single corn were excised. No major complications were found in the foot due to the corns apart from the routine morbidity of pain while walking.

## Discussion

In 1975, Jablonska first gave the concept of scleroderma. Scleroderma literally means sclerosis of the skin. Systemic sclerosis otherwise also known as systemic scleroderma has in addition to sclerosis of skin, other multiple system involvement. The diagnosis of systemic sclerosis depends mainly on the presence of characteristic clinical features along with laboratory investigations. The cutaneous changes discussed earlier are more prominent on face and hands. Similar changes of digital ulceration and pitted scars, hyperpigmentation, and sclerodactyly have been documented in feet but in less severity.<sup>8</sup> A study in 2002<sup>14</sup> showed that compared to hand involvement, the foot involvement in systemic sclerosis has a later onset and less frequent but can be disabling to the patient. Corns and callosities in the feet of systemic sclerosis patients have not been routinely documented apart from two studies from UK in 2001 and 2010 respectively.<sup>12,15</sup> Recently corns in scleroderma have been reported from India by Chathra N et. al.<sup>16</sup>

While analysing the sole findings of our patients we found digital pitted scars in 44(84.61%), corn in 40(76.92%) and macular hyperpigmentation in 40(76.92%) patients as compared to the findings by Sari-Kouzel et al.,<sup>12</sup> where the podiatry assessment of fifty patients

**Fig. 2: Multiple corns in the sole of a systemic sclerosis patient**

of systemic sclerosis found the presence of callosities in 80%, toenail changes in 62%, pre-ulcerative lesions in 34%, calcinosis in 18% and ulcerations in 10% of patients. Similar findings were seen by Alcacer-Pitarch B et al.<sup>15</sup> where 95% of patients of systemic sclerosis attending foot health clinic received interventions for corns and calluses. Other foot interventions were toe nail care (100%), insole provision (52%), ulcer care (76%), nail surgery (14%) etc.

Excess collagen production has been seen in systemic sclerosis leading to microvascular and macrovascular damage and fibrosis of the skin as well as internal organs. Digital pitted scar in systemic sclerosis has been associated with Raynaud's phenomenon, skin sclerosis and articular involvement in the form of swelling, pain or stiffness.<sup>7</sup>

Corns and callosities are areas of hyperkeratosis on weight bearing parts or bony prominences on the palms and soles.<sup>17</sup> Deformity in the foot associated with unsuitable footwear many a times contribute to the condition.<sup>18</sup> Some intrinsic abnormalities of foot predisposing to callosities include bony prominences, a prominent condylar projection or malunion of a fracture. In patients with rheumatoid arthritis there are distinctive patterns of callosity formation.<sup>19</sup> Diabetic patients with neuropathy are prone to callus formation.<sup>20</sup> As a whole faulty foot biomechanics lead to callosities.<sup>21-23</sup> An inherited predisposition to callosities has been described, with an autosomal dominant inheritance.<sup>24,25</sup> In systemic sclerosis there is increased fibroblast activity and collagen synthesis leading to skin fibrosis. Increased plantar pressure in systemic sclerosis<sup>26</sup> patients

along with skin fibrosis makes the pressure bearing areas of soles more prone for corn formation as found in our study.

There are various patterns of pigmentation described in scleroderma patients like Addisonian type, vitiligo type, streaky hyperpigmentation over blood vessels on a background of depigmentation and reticulate type.<sup>27-29</sup> The underlying pathogenesis has been proposed as increased keratinocyte-derived endothelin-1 (ET-1), increased melanin synthesis and increased secretion of melanocytic growth factors by fibroblasts and endothelial cells.<sup>30-32</sup> The macular hyperpigmentation seen in soles in our patients can be described as a part of the generalized hyperpigmentation in the disease process.

Studies on foot ailments in systemic sclerosis patients are limited. Corns and callosities have not yet been documented as a clinical finding in systemic sclerosis patients in any of the standard text books, to the best of our knowledge. Corns and callosities are harbingers of deeper ulcer formation if not intervened timely. So, the importance of presenting this study is to highlight the fact that, apart from other commonly described findings of soles, corns are a common ailment in patients with systemic sclerosis and needs a careful dermatological examination and necessary early interventions. Further studies with larger sample size are needed with regards to foot problems in patients with systemic sclerosis.

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