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Demographic Profile and Cutaneous Manifestations of Systemic Sclerosis: A Study on 35 Patients of Jharkhand State

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Abstract: ***Background: Systemic sclerosis (SSc) is a disorder of connective tissue with an unknown cause affecting multiple systems of the body. The clinical presentation often features significant skin manifestations that are important for diagnosis and prognosis. This study aimed to determine demographic profile, frequency & characteristics of various cutaneous manifestations of SSc in patients from tertiary care centre. Additionally, the study compared the pattern & frequency of these features in between the diffuse subtype form and limited subtype of this pathological disease. Material & methods: This was a cross - sectional, observational study. Cases of systemic sclerosis visiting department of dermatology in tertiary care centre of Jharkhand over 2 years were included in our study. Results: Total 35 patients, 30 females and 5 males; mean age of systemic sclerosis cases were evaluated. Among cutaneous features, Sclerodactyly and Raynaud's Phenomenon were most common finding. Conclusion: This study gives an overall picture of spectrum of demographic profile along with clinical profile of systemic sclerosis in patients in population of Jharkhand.***

Keywords: Demographic profile, clinical profile, systemic sclerosis, Jharkhand

1. Introduction

Systemic sclerosis (SSc) is a long - term, multi - organ pathological disorder of unknown cause, marked by skin thickening and abnormalities in various organs. Extent & severity of skin and different internal organ involvement can vary. Changes in blood vessels and tissue fibrosis are thought to be crucial in the development of SSc. Numerous cutaneous findings are present in Systemic Sclerosis, and recognizing these features is necessary in order to get earlier diagnosis.

There are two main types of systemic sclerosis: limited cutaneous systemic sclerosis (lcSSc), which involves distal extremities and face while spares the proximal extremities & diffuse cutaneous systemic sclerosis (dcSSc), which affects both proximal and distal extremities. These forms differ significantly in disease progression. In dcSSc, visceral involvement can occur within weeks, whereas lcSSc patients usually remain static for a long period of time but may eventually develop delayed complications of many visceral organs. Differences in clinical features and laboratory findings of SSc among different ethnic groups have been well documented.

Jharkhand, a state of eastern India, is characterised by diverse demographics and a mixture of rural as well as urban population. Rajendra institute of Medical Sciences (RIMS), Ranchi, is the main tertiary care centre in Jharkhand and provides a distinctive perspective on the epidemiology of systemic sclerosis on different regional basis.

Aim

To assess clinico - demographical profile in patients of systemic sclerosis in tertiary care centre of Jharkhand.

2. Material and Methods

- **Study setting**: Department of Dermatology, venereology and Leprosy, Rajendra institute of Medical Sciences (RIMS), Ranchi
- **Study design**: This was cross - sectional, clinical observational study.
- **Study period**: This study was performed over duration of 18 months, starting from January 2023 to June 2024.
- **Study population**: 35 patients (30 female and 5 male) of systemic sclerosis during a specified period of time. Informed consent was obtained from all the patients (and guardian if needed). Detailed history about demographic profile, onset of disease, duration, as well as progression of cutaneous lesions, drug intake, family history of cases was taken. All cases were examined properly keeping main target to cutaneous findings.
- **Inclusion criteria** - Cases satisfying "American College of Rheumatology (ACR) criteria" for systemic sclerosis were involved in our series. "Major criterion being sclerosis proximal to the metacarpophalangeal joints. " "Minor criteria for SSc were sclerodactyly, digital pitting scars, and bibasilar pulmonary fibrosis". To establish a condition of Systemic Sclerosis, either one major or two minor criteria were necessary.
- **Exclusion criteria** - We excluded patients with other sclerotic conditions like eosinophilic fasciitis, scleromyxoedema, scleredema of Bushke or patients on drugs causing fibrosis like bleomycin, pentazocine etc.
- Routine laboratory tests, such as a complete blood count (CBC), C - reactive protein (CRP), and ESR levels evaluated. Specialized tests included measuring ANA profile with antinuclear antibody, anti - topoisomerase - 1, anticentromere antibody, and X ray imaging of hand joints and lungs. When necessary, histopathology of lesions,

imaging with barium swallow, and other relevant investigations performed.

Statistical analysis

Categorical data were analysed with a chi - square test. Continuous variables were compared by unpaired student t test. All tests were done by using SPSS software (version 20.0). A P value <0.05 = statistically significant.

3. Results

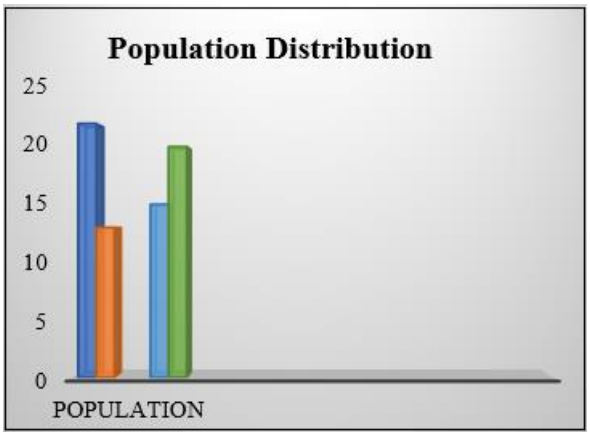
Total 35 patients (female: male F: M = 6: 1; mean age = 27.2±14.7 years) of SSc were evaluated

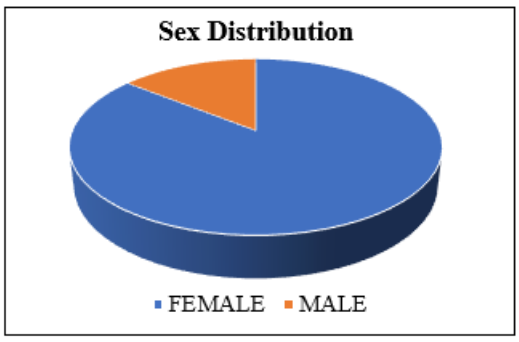
Family history of Systemic Sclerosis was significant in none of the cases. The mean duration of Systemic Sclerosis was around 2 years.

Among total study population, 13 patients (37%) were belonging to tribal population and rest (63%) to non - tribal areas.

Among total 35 patients in our study, 15 (42%) patients were from rural areas and 20 (57.1%) were from urban areas.

23 (65.7%) cases had history of swollen hands along with feet as their primary symptom. Among all the cutaneous features, Raynaud's Phenomenon (RP) was seen in 30 (85.7%) cases. Out of all the 30 cases, it started with tightening of skin in 86.6% (26) of cases and in the rest (4, 13.4% cases), both the symptoms were appeared simultaneously.





Sclerodactyly was seen in 31 (88.5%) patients and microstomia (restriction in opening the mouth) in 28 (80%) cases.

Ulceration & scarring of fingertip were present in 22 (62.8%) patients.

Dyspigmentation was seen in 29 (82.8%) patients, salt and pepper pigmentation was noted in 19 (54.2%) patients, diffuse type of hyperpigmentation was noted in 12 (34.2%) cases, and localized types of pigmentation in 3 (8.5%) cases.

Flexion contractures in fingers were observed in 11 (31.4%) cases & digital amputation seen in 3 (8.5%) patients.

Mat - like telangiectasia in 7 (20 %) patients, cutaneous calcinosis observed in only 1 (2.8%) case of SSc, and gangrene of digits was present in only 1 (2.8%) patient, generalized pruritus in 3 (8.5%) cases. Diffuse alopecia affected 5 (14.2%) cases. Nail changes noted in 10 (28.5%) cases. The nail in these SSc patients were shiny nails in 6 (17.1%) patients, increased convexities in 3 (8.5%), clubbing of the fingers in 2 (5.7%), periungual telangiectasia in one (2.8%).

ANA profile was performed in all the cases.28 (80%) cases were ANA positive. Analysis shows Anti Scl - 70 Antibody positive in 20 cases and Anti - centromere antibody was found in 8 patients.

Finding	No. of Patients	Percentage
Sclerodactyly	31	88.5%
Raynaud Phenomenon	30	85.7%
Cutaneous dyspigmentation	28	80%
Microstomia	28	80%
Swollen hands	23	65.7%
Fingertip ulceration	22	62.8%
Flexion contracture of digits	11	31.4%
Nail changes	10	28.5%
Mat like telangiectasia	7	20%
Diffuse alopecia	5	14.2%
Generalised pruritus	3	8.5%
Digit amputation	3	8.5%
Gangrene of digits	1	2.8%
Calcinosis cutis	1	2.8%

4. Discussion

Systemic sclerosis (SSc) presents a wide range of mucocutaneous symptoms. Nearly all SSc patients exhibit skin involvement, making these manifestations key to early diagnosis and classification into disease subsets. SSc is categorized as diffuse cutaneous (dcSSc) or limited cutaneous (lcSSc) based on the extent of skin involvement (proximal or distal to the elbows and knees). Limited cutaneous disease typically follows Raynaud's phenomenon and features skin thickening distal to the elbows, along with gastrointestinal and pulmonary fibrosis, and anticentromere antibody positivity. In contrast, diffuse cutaneous disease is characterized by simultaneous Raynaud's phenomenon, skin involvement proximal to the elbows, and fibrosis affecting the gastrointestinal, pulmonary, renal, and cardiac systems.

Systemic Sclerosis can be seen at any given age & can occur in both sexes. In accordance to our data, our series showed a higher prevalence of SSc in females (male to female ratio of 0.14). Similar findings were reported in 'North Indian study' (M: F = 0.19).

Approximately 85% of SSc occur in people belonging to age group of 20 - 60, though disease are also seen in children as well as in elderly population.

The average age of onset in the present study was 27.2±14.7 years. Comparable results were found in a 'North Indian study', where the mean for age of onset was 32.75 ± 11.62 years, and in a study from Dakar, where the average age was 33 years. In a recent study reported that 8.9% of cases occurred in children.

In our study, we have seen systemic sclerosis more in non - tribal population and in patients belonging to urban areas.

ANA positivity was found in 80% patients in this study. Previous studies suggested variable finding ranging between 50 % to 90 % of ANA positivity.

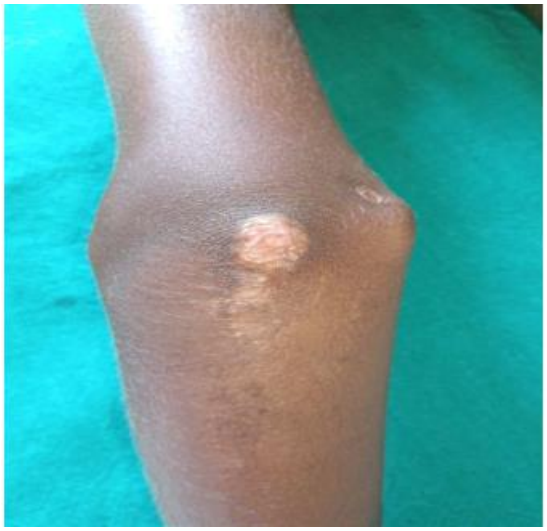
In SSc, the skin is almost always affected. Early symptoms may include puffiness of face, swelling of extremities, and reduced flexibility of tendons and joints. In the present study, 65.7 % of patients had puffy hands, and all patients exhibited cutaneous sclerosis.

Raynaud's phenomenon (RP) is almost always present in Systemic Sclerosis, presenting as episodes of blanching of digits (whitish discoloration), followed by cyanosis (bluish discoloration), and then finally rubor (reddish discoloration). In this present study, 85.7% of cases experienced Raynaud's phenomenon. RP usually precedes other symptoms in limited form of SSc and appears along with skin tightness in diffuse SSc. In our study, RP either preceded or occurred simultaneously with skin tightness.

Ulceration of Fingertip was seen in 65% cases. Similar frequencies of lesions were observed among the Afro - Caribbean population (93%) and in North Indian study (91%). However, lower frequencies were reported in a South Indian study (28%) These discrepancies likely result from ethnic and climatic differences.

Numerous forms of pigmentary abnormalities, such as hyperpigmentation including both localized and diffuse, as well as salt - and - pepper pigmentation, observed commonly in SSc. In our study, dyspigmentation was a prominent finding, affecting 85.7% of patients. Previous studies reported hyperpigmentation incidences ranging from 73.1% to 91%. Cutaneous calcinosis on bony prominences noted in 2.8% of cases.


These pictures showing salt & pepper pigmentation over neck, clavicular region and upper chest. Also overcrowding of teeth and restricted mouth opening




Calcinosis cutis and finger- tip scars are visible in the pictures

5. Conclusion

This study offers an overview about the range of cutaneous manifestations of Systemic Sclerosis in the population of Jharkhand. Several cutaneous findings of systemic sclerosis are known to be very helpful in making quick diagnosis. limitation of this study was its relatively small sample size.

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Declaration of Interest: "The authors report no conflicts of interest. The authors alone are responsible for the content and writing of this paper. "

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