

## Case Series

# Salt-and-pepper dyspigmentation as a distinctive feature in systemic sclerosis: A case series

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## ABSTRACT

Systemic sclerosis (SSc) is a rare autoimmune disorder characterized by tissue fibrosis, immune dysregulation, and vascular damage. SSc classically presents cutaneous features such as scleroderma, Raynaud's phenomenon, salt-and-pepper dyspigmentation, and telangiectasia. Anticentromere, anti-Scl-70, and anti-RNA III antibodies are well-established biomarkers of SSc. This study aimed to illustrate the clinical profile of SSc patients from western Assam in northeast India. The American College of Rheumatology/European League Against Rheumatism classification criteria of 2013 were used for diagnosing SSc. Arthralgia, myopathy, and a history of Raynaud's phenomenon were commonly observed, whereas sclerodactyly and puffy fingers were observed in seven and three cases, respectively. Half of the cases were found to have interstitial lung disease (ILD). This case series identified salt-and-pepper skin dyspigmentation as a distinctive cutaneous feature of SSc. One case also exhibited a unique presentation of serpentine supravenuous hyperpigmentation (SSH) along the paths of superficial veins, against a backdrop of salt-and-pepper dyspigmentation. All the cases were found to be positive for antinuclear antibody using indirect immunofluorescence assay and for anti-Scl-70 antibody using line immunoassay (LIA). This study highlights salt-and-pepper dyspigmentation as a distinctive clinical feature of SSc, alongside anti-Scl-70 antibody positivity in all cases. Further research could provide a comprehensive explanation of these findings.

**Keywords:** Antinuclear antibody, Salt-and-pepper, Scl-70, Systemic lupus erythematosus, Systemic sclerosis

## INTRODUCTION

Systemic sclerosis (SSc) is a rare and complex progressive autoimmune disorder characterized by events of tissue fibrosis, immune dysregulation, and vascular damage.<sup>[1]</sup> The initial trigger of SSc is thought to be autoimmune-mediated attacks on endothelial cells in genetically predisposed individuals upon exposure to specific SSc-associated environmental factors.<sup>[2]</sup> Raynaud's phenomenon often serves as the earliest indicator of the disease.<sup>[3]</sup> SSc is principally classified into limited cutaneous SSc (lcSSc), characterized by slowly progressing localized skin manifestations and diffuse cutaneous SSc (dcSSc), marked by rapidly progressing widespread skin manifestations.<sup>[4]</sup> Classically, the SSc patients present with cutaneous manifestations, such as abnormal skin thickening, puffy fingers, sclerodactyly, and salt-and-pepper dyspigmentation. Other clinical manifestations include Raynaud's phenomenon, restricted mouth opening, fingertip ulcerations and scarring, telangiectasia, calcinosis cutis, hypothyroidism, pulmonary fibrosis, esophageal dysmotility, pulmonary arterial hypertension, and scleroderma renal crisis.<sup>[5]</sup> Salt-and-pepper dyspigmentation of the skin showing perifollicular pigmentary retention can offer a valuable clue for the early diagnosis of SSc.<sup>[6]</sup>

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The 2013 American College of Rheumatology/European League Against Rheumatism (ACR/EULAR) classification criteria are a valuable tool for diagnosing SSc. Under these criteria, cutaneous manifestations are considered reliable signs for SSc diagnosis.<sup>[7]</sup> Among the immunological features of the criteria, circulating anti-nuclear antibodies (ANAs), SSc-specific ANAs such as anticentromere, anti-topoisomerase I (anti-Scl-70), and anti-RNA polymerase III antibodies are recognized as key biomarkers of SSc.<sup>[8,9]</sup>

The study aims to present a case series of ten SSc cases from the western Assam region of India with the objective of exploring the spectrum of clinical and immunological manifestations associated with the disease. ANA was performed by the indirect immunofluorescence assay (IIFA), using the Hep-2 cell line to identify autoantibodies. The line immunoassay (LIA) (immunoblot) was additionally performed to detect various autoantibodies of nuclear, cytoplasmic, and mitochondrial antigens, including SSc-specific autoantibodies.

## CASE SERIES

### Case 1

A 28-year-old male reported experiencing joint pain, body aches, skin pigmentary changes, and shortness of breath during exertion for 3 years, accompanied by a 3-week history of an ulcer on the second toe of his left foot.

Examination revealed rigidity and restricted movement in the interphalangeal and metacarpophalangeal joints. Ulceration with blackish discoloration on the second toe of the left foot had progressed to gangrene. In addition, there was skin thickening and sclerodactyly of the fingers on both hands, along with salt-and-pepper-like dyspigmentation on the upper limbs.

Complete blood count (CBC) report revealed hemoglobin level of 6.9 g/dL, a white blood cell (WBC) count of 9,300/cumm, red blood cell (RBC) count of 2.85 million/cumm, and a platelet count of 3.1 lakh/cumm. Additional investigations showed random blood sugar (RBS) at 92 mg/dL, total serum bilirubin at 0.4 mg/dL, serum glutamic oxaloacetic transaminase of 46 U/L, SGPT (serum glutamic pyruvic transaminase) of 27 U/L, serum urea of 41 mg/dL, serum creatinine of 0.8 mg/dL, thyroid-stimulating hormone (TSH) of 2.63 mIU/L, with normal serum electrolytes. Urine analysis revealed albuminuria and the presence of granular casts. X-ray of both hands with wrists and forearms detected osteoporotic changes and ulnar deviation of fingers. High-resolution computed tomography (HRCT) of the thorax revealed interstitial lung disease (ILD) with pericardial effusion. A Doppler study of the left lower limb detected ischemic changes affecting the second left toe.

IIFA showed the presence of a homogenous nucleolar ANA pattern, and LIA confirmed the presence of anti-Scl-70 antibody. With an ACR-EULAR score of 12, a diagnosis of SSc with ILD and digital gangrene was established. He received conservative treatment during his hospital stay.

### Case 2

A 42-year-old female presented with easy fatigability, dyspnea with moderate exertion, occasional non-productive cough, arthralgia, skin dyspigmentation, and skin tightening around the mouth for around 3 years, with a history of hypothyroidism and Raynaud's phenomenon during winters. Examination revealed skin tightening around the perioral region and upper limbs, along with restricted mouth opening. Dyspigmented salt-and-pepper-like lesions were noted on her scalp and ears. On palpation, the thyroid gland was found to be normal in shape and contour.

The CBC report indicated lower levels of hemoglobin, WBC count, and RBC count. The absolute lymphocyte count was found to be low at  $0.39 \times 10^3/\mu\text{L}$ . The details of blood parameters are shown in Table 1. HRCT thorax revealed ILD, while both echocardiography and abdominal ultrasonography (USG) results were normal.

IIFA showed a nuclear homogeneous ANA pattern, and LIA detected the presence of anti-Scl-70 antibody. With an ACR-EULAR score of 10, she was diagnosed as a case of SSc with ILD and managed conservatively.

### Case 3

A 60-year-old female presented with a 2-year history of skin thickening and abnormal pigmentation, and finger-tip scars for 1 year. On examination, skin tightening and fibrosis were observed on her face and upper limbs. She exhibited microstomia with significantly limited mouth-opening capacity. Dyspigmented salt-and-pepper-like lesions were observed on her hands and around her eyebrows. She also had nail clubbing, puffy fingers, fingertip scars, and finger shortening.

The CBC report revealed low hemoglobin levels but normal levels of WBC, RBC, and platelet counts [Table 1]. RBS, liver function test (LFT), and kidney function test (KFT) were also normal [Table 1]. Chest X-ray, electrocardiogram (ECG), and USG abdomen did not reveal any abnormality. IIFA showed a homogenous nucleolar ANA pattern, and LIA demonstrated anti-Scl-70 antibody. With an ACR-EULAR score of 11, she was diagnosed with SSc and treated conservatively.

### Case 4

A 35-year-old female presented with a 2-year history of abnormal skin pigmentation and breathing difficulty, along

**Table 1:** Showing the clinical findings and relevant investigations of all the SSc cases of the study.

Clinical findings	Case No.				
	Case 1	Case 2	Case 3	Case 4	Case 5
Puffy fingers	X	Present	Present	X	X
Sclerodactyly	Present	X	X	Present	Present
Fingertip ulcer	X	X	X	Present	X
Fingertip pitting scar	X	X	Present	X	Present
Digital gangrene	Present	X	X	X	X
Raynaud's phenomenon	Present	Present	Present	Present	Present
Interstitial Lung Disease (ILD)	Present	Present	X	Present	X
Pericardial effusion	Present	X	X	X	X
Salt-and-pepper lesion	Present	Present	Present	Present	Present
Restricted mouth opening	Present	Present	Present	Present	Present
Calcinosis	X	X	X	Present	X
Oesophageal dysmotility	X	X	X	X	X
Immunological tests					
Antinuclear Antibody (ANA)	Positive	Positive	Positive	Positive	Positive
ANA pattern	Homogenous nucleolar	Nuclear homogenous	Homogenous nucleolar	Nuclear homogenous	Homogenous nucleolar
Anti Scl-70 antibody	Positive	Positive	Positive	Positive	Positive
Blood Tests					
Hb (gm/dl)	6.9	8.4	7.5	11.7	13
WBC (cells/cumm)	9300	3840	3770	19970	8600
RBC (million cells//cumm)	2.85	2.31	4.12	4.9	4.6
Platelet count (lakh/cumm)	3.1	1	3.7	4	2.4
RBS (mg/dl)	92	84	101	92	132
TSB (mg/dl)	0.4	0.8	0.4	0.5	0.8
SGOT (mg/dl)	46	49	39	39	43
SGPT (mg/dl)	27	58	27	32	26
Urea (mg/dl)	41	20	21	18	29
Creatinine (mg/dl)	0.8	0.8	0.7	0.6	0.7
TSH (mIU/L)	2.63	2.63	-	-	3.5
Urine Tests					
Sugar	Nil	Nil	Nil	Nil	Nil
Proteinuria/Albuminuria	Present	Nil	Nil	Present	Nil
Cast	Present	Nil	Nil	Nil	Nil
Clinical findings	Case No.				
	Case 6	Case 7	Case 8	Case 9	Case 10
Puffy fingers	X	X	Present	X	X
Sclerodactyly	Present	Present	X	Present	Present
Fingertip ulcer	X	X	X	X	X
Fingertip pitting scar	X	X	Present	X	X
Digital gangrene	X	X	X	X	X
Raynaud's phenomenon	Present	Present	Present	Present	Present
Interstitial Lung Disease (ILD)	X	Present	X	X	Present

(Contd...)

**Table 1:** (Continued).

	Case no.				
	Case 6	Case 7	Case 8	Case 9	Case 10
Salt-and-pepper lesion	Present	Present	Present	Present	Present
Restricted mouth opening	Present	Present	Present	Present	Present
Calcinosis	X	Present	Present	Present	X
Oesophageal dysmotility	X	X	X	X	Present
Immunological tests					
Antinuclear Antibody (ANA)	Positive	Positive	Positive	Positive	Positive
ANA pattern	Homogenous nucleolar	Smooth nuclear envelope	Homogenous nucleolar	Nuclear fine speckled	Few nuclear dots
Anti Scl-70 antibody	Positive	Positive	Positive	Positive	Positive
Blood Tests					
Hb (gm/dl)	9.6	7.9	12.1	7.4	9.3
WBC (cells/cumm)	7190	11000	10250	8100	9240
RBC (million cells//cumm)	3.86	3.57	4.68	2.6	3.93
Platelet count (lakh/cumm)	1.5	1.7	1.6	1.5	1.5
RBS (mg/dl)	108	86	159	100	93
TSB (mg/dl)	0.9	0.7	0.6	0.9	0.5
SGOT (mg/dl)	28	25	29	44	21
SGPT (mg/dl)	21	19	20	32	13
Urea (mg/dl)	20	30	33	39	12
Creatinine (mg/dl)	0.7	0.7	0.8	1.1	0.6
TSH (mIU/L)	-	1.0	3.8	1.87	2.5
Urine Tests					
Sugar	Nil	Nil	Nil	Nil	Nil
Proteinuria/Albuminuria	Nil	Present	Nil	Nil	Nil
Cast	Nil	Nil	Nil	Nil	Nil

Hb: Haemoglobin; WBC: White Blood Cells; RBC: Red Blood Cells; RBS: Random Blood Sugar; TSB: Total Serum Bilirubin; SGOT: Serum Glutamic Oxaloacetic Transaminase; SGPT: Serum Glutamate Pyruvate Transaminase; TSH: Thyroid-Stimulating Hormone; X: Absent; SSc: Systemic Sclerosis

with ulcerations on the elbows, left knee, and buttocks for the past 4 months.

Extensive depigmented salt-and-pepper-like lesions were observed on her neck, chest, upper limbs, and lower limbs. Sparing of serpentine supravenuous hyperpigmentation (SSH) over the anterior surface of the lower limbs was also noticed. Gross ulcerations were seen over her elbows and buttocks. A smaller ulceration was present on her left knee. She had alopecia and sclerodactyly of the fingers. Calcinosis, appearing as whitish-yellowish papules and nodules, was visible across various parts of her skin. Over time, she became bedridden due to decreased muscular strength. Gross muscular fibrosis and atrophy of her limb muscles were also noticed. She previously had episodes of Raynaud's phenomenon.

Blood investigations revealed low hemoglobin level and high WBC count; however, RBS, LFT, and KFT were normal [Table 1]. Proteinuria was detected by 24-h urinary

protein analysis [Table 1]. Other investigations, such as serum electrolytes and lipid profile, were normal. HRCT of the chest revealed ILD, and USG abdomen suggested B/L nephrocalcinosis and renal calculi. Spirometry indicated severe impairment of lung function. Echocardiography and barium swallow X-ray were normal. A nuclear homogeneous ANA pattern was detected by IIFA, and anti-Scl-70 antibody was detected by LIA. Her ACR-EULAR score was 12. She was diagnosed with SSc with ILD and treated conservatively.

#### Case 5

A 35-year-old male presented with an 8-month history of diffuse skin thickening, abnormal skin pigmentation, and fingertip scars. Thickened skin was observed on his hands and face, with visible scars on his fingertips, with a history of Raynaud's phenomenon. Depigmented, salt-and-pepper-like lesions were noted on his scalp, forehead, and hands.

CBC, LFT, and KFT were within normal ranges [Table 1]. Serum electrolytes and lipid profile were also normal. No abnormalities were detected in the ECG, chest X-ray, or USG abdomen. IIFA demonstrated the presence of a homogenous nucleolar ANA pattern, and LIA detected anti-Scl-70 antibody. His ACR-EULAR score was 13, hence diagnosed as a case of SSc and treated conservatively in the hospital.

#### Case 6

A 35-year-old female presented with a 2-year history of skin tightening, skin dyspigmentation, multiple joint pains, and hypertension. On examination, her blood pressure was elevated at 160/100 mmHg. Skin thickening was observed on her face and fingers. Swelling and tenderness were noted in the metacarpophalangeal joints. Depigmented salt-and-pepper-like lesions were evident on her forehead and hands, and she reported a history of Raynaud's phenomenon.

Her CBC report indicated a low level of hemoglobin, whereas WBC, RBC, and platelet counts were normal [Table 1]. Additional test results of RBS, LFT, and KFT have also been mentioned in Table 1. Serum electrolytes, ECG, and chest X-ray were normal. However, her USG abdomen report suggested the presence of gall bladder sludge. IIFA demonstrated a homogeneous nucleolar ANA pattern, and LIA confirmed the presence of anti-Scl-70 antibody. With an ACR-EULAR score of 10, she was diagnosed with SSc and managed conservatively in the hospital.

#### Case 7

A 21-year-old male presented with arthralgia, restricted finger movement, dyspigmentation of skin, restricted mouth opening, and a history of Raynaud's phenomenon triggered by cold exposure for the past 2 years. He also reported occasional shortness of breath over the past year. Examination revealed stiff skin on his face and fingers, along with proximal interphalangeal joint contractures that limited full extension of his fingers. Calcinosis was observed around some of his joints. Dyspigmented salt-and-pepper-like lesions were noted on his forehead, neck, and thighs.

His CBC report revealed low hemoglobin and normal WBC, RBC, and platelet counts [Table 1]. RBS, LFT, and KFT were within normal limits [Table 1]. C-reactive protein (CRP) was elevated at 42.80 mg/L, and the urine-albumin creatinine ratio of 9.86 indicated proteinuria. Serum electrolytes, TSH, and ECG findings were normal; however, spirometry indicated restrictive lung disease, and NCCT chest revealed ILD. Smooth nuclear envelope ANA pattern was observed by IIFA, and anti-Scl-70 antibody was detected by LIA. His ACR-EULAR score was 12, and diagnosed as SSc with ILD and treated accordingly.

#### Case 8

A 40-year-old male presented with a decade-long history of weakness, pigmentary changes in skin, multiple joint pains, and skin tightening. He reported color changes in his fingers upon exposure to cold water, indicative of Raynaud's phenomenon.

On examination, skin tightening was evident on his face, fingers, and forearms. Puffy fingers were noted along with widespread scaling and peeling of skin from the fingers. Calcinosis was observed in the subcutaneous tissue of his right elbow. Dyspigmented salt-and-pepper skin-like lesions were present on his face, ears, neck, chest, forearms, back, and along his spine. In addition, he exhibited mild perioral skin tightening and oral mucosal ulcerations.

His blood test results (CBC, LFT, KFT, RBS, and TSH) have been summarized in Table 1. Lipid profile, electrolytes, chest X-Ray, and ECG were normal. ANA detection by IIFA revealed a homogenous nucleolar pattern, and the presence of anti-Scl-70 antibody was detected by LIA. His ACR-EULAR score was 11, diagnosed as SSc, and managed conservatively.

#### Case 9

A 59-year-old female presented with multiple joint pains, skin tightening, and abnormal skin pigmentation for the past 6 months. She often suffered from color changes of the digits upon exposure to cold water.

On examination, patches of alopecia were noted, along with dyspigmented salt-and-pepper-like lesions on her scalp, forehead, and face. Swelling and reduced mobility were observed in multiple joints of her upper limbs, particularly in the fingers, where she had developed sclerodactyly. Calcinosis was evident in various finger joints. In addition, the perioral region displayed skin tightening, resulting in limited mouth-opening capacity.

Her CBC report revealed a low hemoglobin level. Details of the blood parameters, such as CBC, RBS, LFT, and KFT, have been summarized in Table 1. Additional tests, such as lipid profile, TSH, electrolytes, X-ray chest, USG abdomen, and ECG reports, were normal. A nuclear, fine speckled ANA pattern was detected by IIFA, and LIA detected anti-Scl-70 antibody. Her ACR-EULAR score was 10, thereby she was diagnosed with SSc; her treatment was initiated accordingly.

#### Case 10

A 45-year-old female presented with a 4-year history of skin dyspigmentation, skin tightening affecting her face and fingers, difficulty in swallowing, and exertional shortness of breath over the past 6 months. She had a history of Raynaud's phenomenon. Skin thickening was observed around the face, perioral region, forearms, and fingers of both hands. Alopecia

was noted on her scalp, while areas of dyspigmentation, salt-and-pepper-like lesions were seen on her scalp, forehead, and face. Her blood pressure was recorded at 148/94 mmHg.

The CBC report indicated a low hemoglobin level of 9.3 g/dL. Details of WBC, RBC, platelet count, liver enzymes, and KFT values have been shown in Table 1. ECG and abdominal USG were unremarkable. Barium swallow X-ray revealed esophageal dysmotility and distal esophagus stricture, and the HRCT thorax findings suggested ILD. Further, IIFA detected the presence of a few nuclear dots, an ANA pattern, and the presence of anti-Scl-70 antibody. The ACR-EULAR score was 12, diagnosed as SSc with ILD, and treated accordingly.

## DISCUSSION

The study was conducted in a tertiary care hospital in the northeast Indian population. After screening 280 patients with suspected autoimmune disorders over a span of 2 years, 10 patients were identified with SSc. The cases consisting of six females and four males had a mean age of 37 ( $\pm 13.4$ ) years. All the cases met the ACR-EULAR 2013 classification criteria for SSc, achieving an average score of 12.

At the time of diagnosis, arthralgia, myopathy, and Raynaud's phenomenon were present in all the cases, especially during the winter months. Oropharyngeal manifestations, perioral skin tightening, and a reduced oral aperture that hindered eating and oral hygiene were consistently observed in all the cases.

Of the three common dyspigmentation patterns of SSc, i.e., generalized hyperpigmentation, focal pigmentary changes, and salt-and-pepper pattern,<sup>[10]</sup> all cases in the current study typically presented with salt-and-pepper dyspigmentation of the skin with perifollicular pigmentary retention, making the salt-and-pepper pattern a significantly unique characteristic observed in SSc cases from this region [Figure 1]. Perifollicular pigment retention is thought to result from a richer capillary supply near follicles, maintaining warmth and supporting

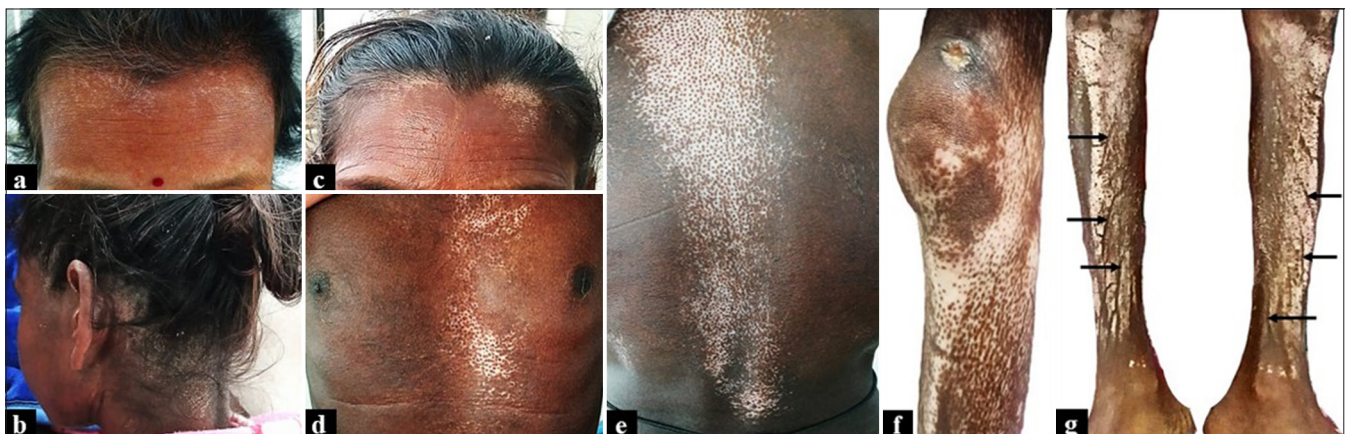
melanogenesis in those areas.<sup>[11]</sup> This study is concurrent with past findings of the prevalence of salt-and-pepper dyspigmentation in SSc.<sup>[12,13]</sup> A past study on the Eastern Indian population reported salt-and-pepper pigmentation in 33.3% cases of lcSSc and 66.67% cases of dcSSc.<sup>[14]</sup>

Half of the cases in the present study were identified with ILD. Similarly, a recent study reported a predominance of ILD among the population, with overlapping features of salt-and-pepper dyspigmentation.<sup>[13]</sup> Indeed, a strong correlation between salt-and-pepper dyspigmentation and the presence of digital ulcers is also evident.<sup>[15]</sup>

In addition, in the current study, one case presented with a rarely described presentation of sparing the SSH along the path of superficial veins, on the background of salt-and-pepper depigmentation, possibly due to the protective effects of blood flow through the veins.<sup>[16]</sup> Thus, screening for salt-and-pepper dyspigmentation may serve as a reliable clinical sign in aiding early diagnosis of SSc and in identifying patients at heightened risk for developing digital ulcers and vascular complications.

Musculoskeletal manifestations such as thickened and fibrotic skin of the fingers distal to the metacarpophalangeal joint, presenting as sclerodactyly with limited mobility of the affected fingers, were observed in seven cases, while distal fingertip ulcer and fingertip pitting scar were also observed in some cases [Figure 2]. In addition, hypertension was detected in two cases, proteinuria in three cases, and one case demonstrated granular casts in urine. Esophageal dysmotility due to fibrosis of the esophagus and pericardial effusion as a cardiac manifestation was detected in two separate cases. Thus, the distinctive features of SSc, such as salt-and-pepper dyspigmentation, skin thickening, Raynaud's phenomenon, and restricted mouth opening, exhibited a strong association with SSc.

ANA testing by IIFA on HEp-2 cell line was found positive in all 10 SSc cases. The most common ANA pattern observed was homogeneous nucleolar, detected in half of the cases. The



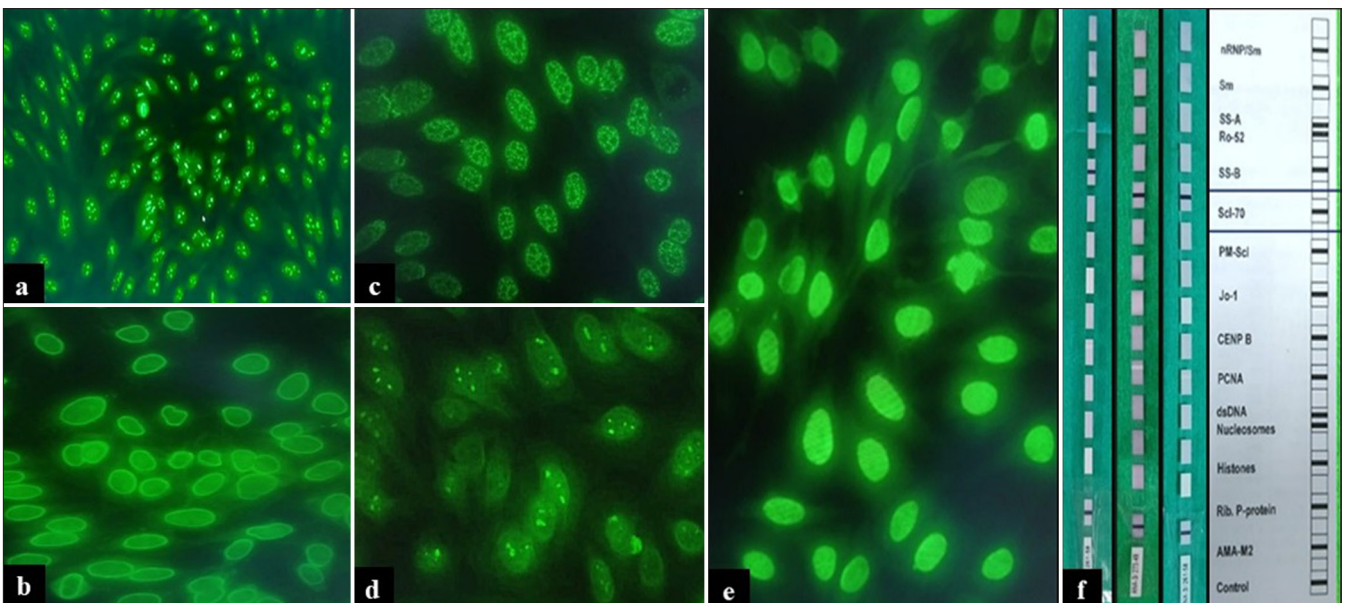
**Figure 1:** (a-f) Salt-and-pepper dyspigmentation over various body regions of different cases, (g) Supraventine sparing of salt-and-pepper dyspigmentation (indicated by black arrows).

remaining five cases showed nuclear homogeneous, smooth nuclear envelope, nuclear fine speckled, and a few nuclear dots patterns [Figure 3]. A previous study from western India and a separate study from the USA reported the speckled ANA pattern as the most common ANA pattern observed in SS.<sup>[17,18]</sup> Another study suggested centromere, nucleolar, and

speckled patterns to be less commonly associated with SS.<sup>[19]</sup> A recent study found the nuclear speckled pattern was most commonly associated with systemic lupus erythematosus (SLE), followed by the nuclear homogenous and mixed patterns.<sup>[20]</sup> However, the current study suggests and confirms the potential association of various ANA patterns with SS.



**Figure 2:** Other manifestations of systemic sclerosis as observed in the cases. (a) microstomia, (b) purse mouth, (c) restricted mouth opening, (d) sclerodactyly, (e) ulceration on elbow, (f) calcinosis cutis (indicated by white arrow), (g) puffy fingers, (h) acro-osteolysis of distal phalanx causing clubbing and finger shortening, (i) barium x-ray showing distal esophageal stricture (indicated by white arrows), and (j) chest-X ray (CXR) showing ground-glass opacities in lung fields.



**Figure 3:** Anti-nuclear antibody (ANA) patterns observed in the cases by indirect immunofluorescence assay (a-e): (a) Homogenous nucleolar, (b) Smooth nuclear envelope, (c) Nuclear fine speckled, (d) Few nuclear dots, and (e) Nuclear homogenous ANA patterns were observed in the study; (f) Anti Scl-70 antibody positivity as demonstrated in line immunoassay strips.

Furthermore, only SSc-specific anti-Scl-70 antibodies were detected by LIA in all SSc cases. In contrast, antibodies such as nRNP/Sm, Sm, SS-A, Ro-52, SS-B, PM-Scl, Jo-1, CENP B, PCNA, dsDNA, nucleosomes, histones, Rib P-protein, and AMA-M2 were not detected in them, effectively ruling out the presence of any other autoimmune conditions and overlap syndromes. Key SSc-specific antibodies include anti-centromere antibodies, primarily associated with lcSSc and occasionally with dcSSc, Sjögren's syndrome, and SLE; anti-Scl-70 (anti-topoisomerase I), associated with dcSSc, ILD, and cardiac issues; anti-RNA polymerase III seen in rapidly progressive dcSSc, and the presence of anti-U3-RNP (fibrillarin) indicates a poorer prognosis<sup>[5]</sup> whereas anti-Th/To, anti-fibrillarin, and anti-NOR90 antibodies are more rarely found in SSc.<sup>[21]</sup>

## CONCLUSIONS

Despite the wide-ranging features associated with SSc, this study identified salt-and-pepper dyspigmentation as a remarkable clinical manifestation in SSc, accompanied by SSc-specific anti-Scl-70 antibody positivity in all the cases from western Assam. A comprehensive study on SSc can corroborate these findings.

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