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Case Report
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A BEAUTIFUL BETRAYAL: WHEN COLLAGEN BECOMES THE ENEMY

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ABSTRACT

Background: Systemic sclerosis (SSc) is a rare autoimmune connective tissue disease characterized by skin fibrosis, vasculopathy, and internal organ involvement. Early recognition is critical due to potentially life-threatening complications. **Case Presentation:** We report a 33-year-old female presenting with rapidly progressive skin thickening, Raynaud's phenomenon, and new-onset exertional dyspnea. Serology revealed anti-Scl-70 positivity. High-resolution CT (HRCT) of the chest showed early interstitial lung disease (ILD). **Treatment and Outcome:** The patient was managed with immunosuppressive therapy including mycophenolate mofetil and low-dose corticosteroids, showing stabilization of lung function and cutaneous symptoms over 6 months. **Conclusion:** This case highlights the importance of prompt diagnosis and early treatment initiation in systemic sclerosis to prevent irreversible organ damage.

KEYWORDS: Systemic sclerosis, Interstitial Lung disease, Anti Scl-70, Raynaud's phenomenon, vasculopathy.

INTRODUCTION

Systemic sclerosis is a complex multisystem autoimmune disease marked by immune activation, vascular dysfunction, and progressive fibrosis of the skin and internal organs. Its presentation can be variable and often delays diagnosis. Pulmonary complications, particularly interstitial lung disease and pulmonary hypertension, are leading causes of morbidity and mortality in SSc. We present a case of rapidly progressive diffuse cutaneous systemic sclerosis (dcSSc) with early pulmonary involvement, emphasizing the importance of early intervention.

CASE

A 33-year-old woman presented with a 3-month history of symmetric skin tightening over her hands, face, and forearms, associated with Raynaud's phenomenon and joint stiffness. Over the last 4 weeks, she developed progressive shortness of breath on exertion and dry cough. There was no history of smoking, occupational exposure, or previous autoimmune conditions.

Physical examination revealed.

- Puffy fingers and sclerodactyly
- Facial skin tightening
- Salt and pepper pigmentation on retroauricular area and back of trunk.
- Bibasilar fine inspiratory crackles on chest auscultation
- Capillary nailfold abnormalities (dilated loops, hemorrhages)

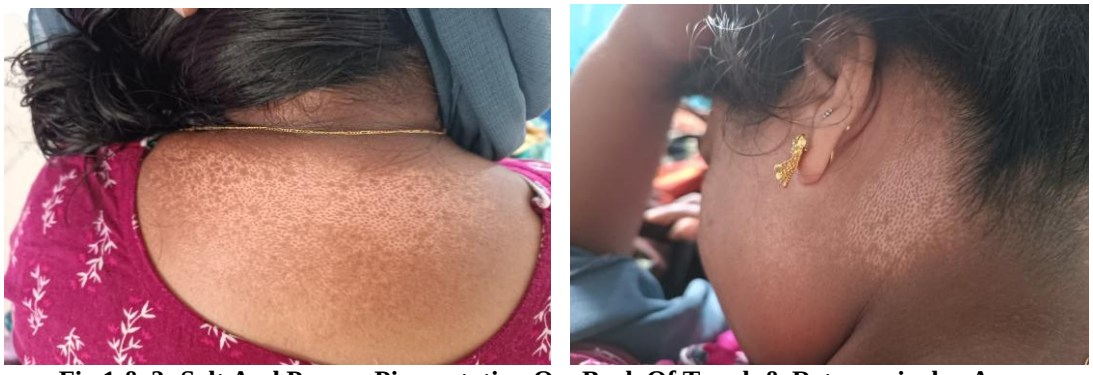

Fig 1 & 2: Salt And Pepper Pigmentation On Back Of Trunk & Retroauricular Area.

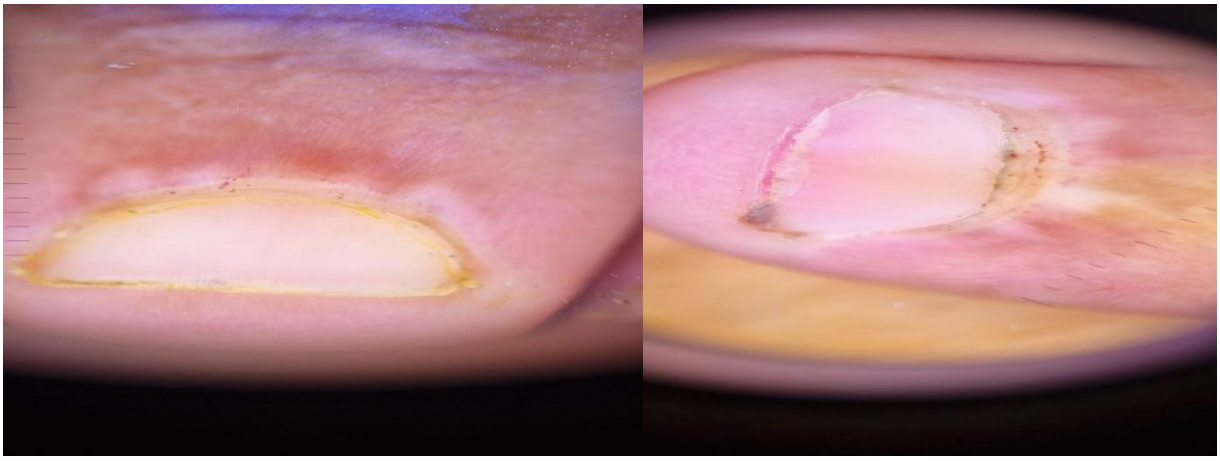

Fig 3&4: Nail Dermoscopy: Dilated Capillaries, Hemorrhagic Spots And Ragged Cuticles.

Laboratory Tests

- ANA positive (speckled pattern, 1:640)
- Anti-Scl-70 antibody positive
- ESR and CRP elevated
- Normal renal function, CBC, and urinalysis

HRCT chest: Early ground-glass opacities in bilateral lower lobes
PFTs: Probable Restrictive Lung disease

MICRO BIOLOGY

ANA PROFILE

nRNP/SM	Negative
Sm	Negative
SS-A (Ro)	Negative
Ro-52	Negative
SS-B (LA)	Negative
Scl-70	Positive ++
PM-Scl	Negative
Jo-1	Negative
CENP-B	Negative
PCNA	Negative
Nucleosomes	Negative
Histones	Negative
Rib-P-Protein	Negative
AMA-M2	Negative
dsDNA	Negative

Fig 5: Ana Profile-Scl-70 Positive.

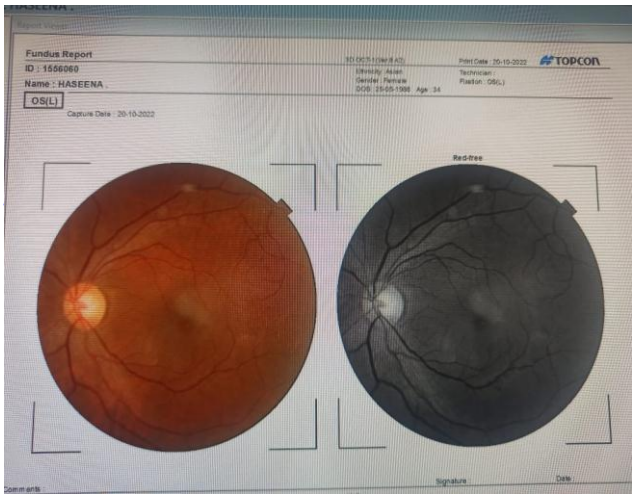

Fig 6: Fundus: Normal.

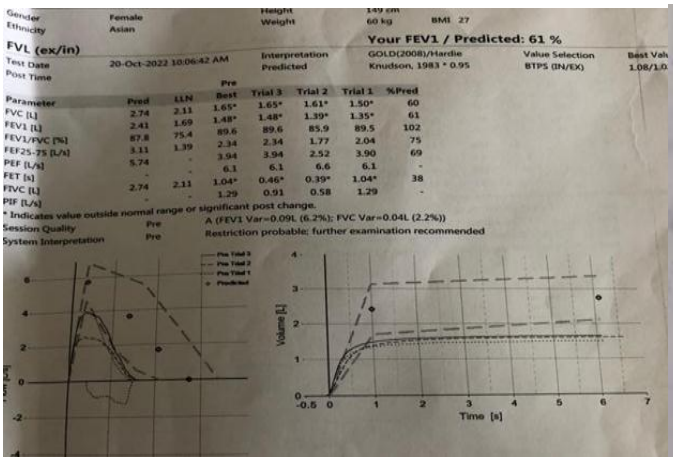

Fig 7: Pft-Probable Restrictive Lung Disease.

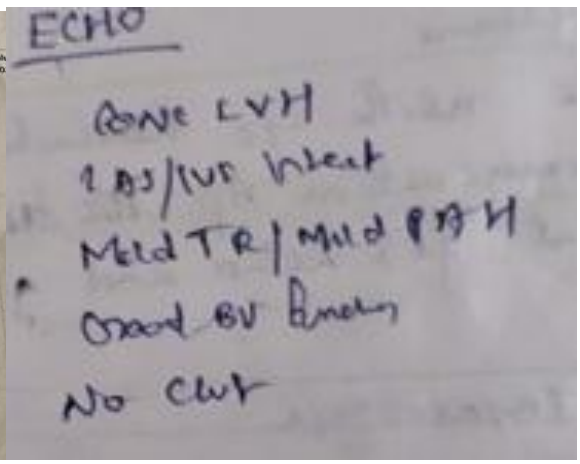

Fig 8: Echo-Mild Pah.

OBSERVATION

- Rapid progression of cutaneous symptoms over 3 months
- Early involvement of lung parenchyma on imaging and PFTs
- Serological profile consistent with diffuse systemic sclerosis (anti-Scl-70 positivity)
- No signs of renal crisis or cardiac involvement on initial assessment

TREATMENT

- **Immunosuppressive therapy:** Mycophenolate mofetil (1g BID)
- **Low-dose corticosteroids:** Prednisolone (10 mg/day) cautiously administered
- **Vasodilator therapy:** Amlodipine for Raynaud's phenomenon
- **Supportive care:** Physical therapy, skin emollients, pulmonary rehabilitation

Follow-up at 3months showed

- Stabilization of respiratory symptoms
- Improvement in skin tightness (modified Rodnan skin score reduced from 21 to 14)
- No further decline in pulmonary function
- No evidence of renal or cardiac involvement

CLINICAL SIGNIFICANCE

The case demonstrates the need for **heightened clinical suspicion** when patients present with **early skin tightening and Raynaud's phenomenon**. Timely diagnosis of dcSSc is essential because it often has **more aggressive organ involvement** than limited cutaneous forms. This case is clinically significant because it exemplifies a rapidly progressive, multi-organ autoimmune condition where **early diagnosis and targeted therapy** can alter disease trajectory. It emphasizes the need for **systematic screening**, **serological evaluation**, and **interdisciplinary coordination** in the management of systemic sclerosis.

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