

## Case Report

# LIMITED CUTANEOUS SCLERODERMA WITH AN UNUSUAL PRESENTATION -A CASE REPORT

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## ABSTRACT

Systemic sclerosis or scleroderma (SSc) is a chronic multisystem disease of unknown etiology characterized by skin induration and thickening, accompanied by fibrosis and chronic inflammatory infiltration of internal organs, microvascular damage and dysfunction, and immune dysfunction. Systemic sclerosis is an uncommon acquired connective tissue disorder characterized by an abnormal thickening of the skin. Long history of Raynaud's phenomenon, Limited skin involvement (peripheral only), Calcification, telangiectasia, digital ischemia, pulp loss, pitted scars, paronychia, late onset of pulmonary hypertension, Capillary dilatation visible in nail folds and Anticentromere antibody positive are seen. We report this case of Limited Cutaneous Scleroderma with an unusual presentation.

**Keywords:** *Limited Cutaneous Scleroderma*

## INTRODUCTION

Systemic sclerosis is now divided into two forms, limited and diffuse disease, based on the extent of skin involvement. Limited disease is defined as skin involvement confined to areas distal to the elbows and knees. Patients with limited cutaneous scleroderma generally have a long duration of Raynaud's phenomenon before they develop puffy fingers and other systemic symptoms. Other common presenting features are of esophageal dysmotility or reflux. Patients are often anticentromere antibody positive and have a high frequency of internal organ involvement, including gastrointestinal tract problems, pulmonary fibrosis, and pulmonary hypertension late in the course of their illness.

## CASES

A 30yr old lady Painful ulcer over the tip of right middle finger since 1 month and Painful swelling of the left index finger since 2 weeks. History of trauma 1 month back. No history suggestive of reynaud's phenomenon or drug intake prior to onset or fever/ joint pain/ breathlessness /difficulty in swallowing of food/ abdominal pain or photosensitivity or tightening of skin.

Right middle finger is swollen and hyperpigmented with finger tip showing ulcer of size 2x2 cms covered with necrotic crusting Left index finger was swollen with showing subungual hyperkeratosis with onycholysis with collection of pus below the nail. Mild swelling of all fingers of both hands Depigmented macules present over knuckles of both hands and both shins. All peripheral pulses were palpable normally Peripheral nerve examination –normal No hypopigmented anaesthetic patches No sclerosis or binding down of skin

**Systemic Examination-**WNL

## DISCUSSION

Systemic sclerosis is an uncommon acquired connective tissue disorder characterized by an abnormal thickening of the skin with associated clinical features such as Long history of Raynaud's phenomenon, Limited skin involvement (peripheral only), Calcification, telangiectasia, digital ischemia, pulp loss, pitted scars, paronychia, late onset of pulmonary hypertension, Capillary dilatation visible in nail folds and Anti-centromere antibody-positive.

Investigations to establish diagnosis and prognosis are Skin biopsy for diagnosis, ESR –raised, Rheumatoid factor –positive 30% of cases, Antinuclear antibodies positive in 78% of cases, Anticentromere antibodies- strongly positive.

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Treatment consists of General measures such as advice the patient to keep as warm as possible, Avoid even minor trauma of the hands and electrically heated gloves are helpful Disease modifying agents immunomodulators such as steroids, cyclosporin A, methotrexate, Cyclophosphamide, Mycophenolatemofetil, infliximab. Therapies for reynaud phenomenon & digital ischemia in systemic sclerosis are Calcium channel blockers like nifedipine, diltiazem, ACE inhibitors, Angiotensin receptor blockers, Anticoagulants and Phosphodiesterase type 5 inhibitors.



**Figure 1 (A): Left Index Finger was Swollen with Showing Subungual Hyperkeratosis with Onycholysis with Collection of Pus below the Nail; Right Middle Finger is Swollen and Hyperpigmented with Finger Tip Showing Ulcer of Size 2x2 cms Covered with Necrotic Crusting**

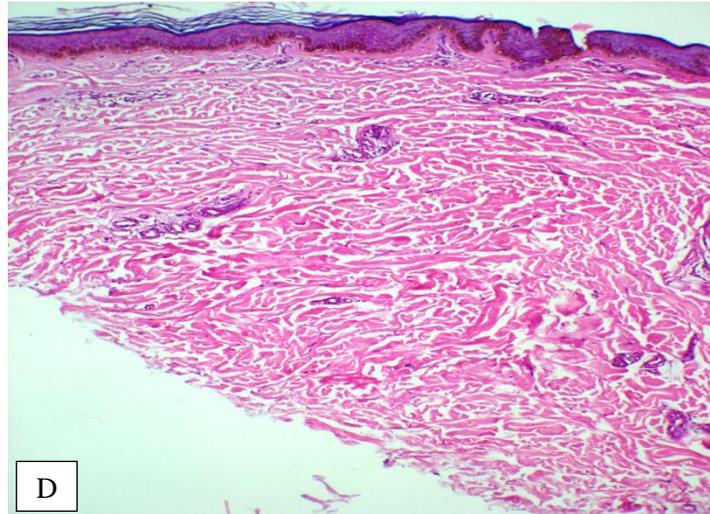


**Figure 1 (B): X ray of Both the Wrists (AP & Oblique X ray of Both the Wrists- Shows Evidence of Acro-Osteolysis of Terminal Tuft of Bilateral Index Fingers, Subtotal Resorption of Terminal Tuft of Terminal Phalynx of Bilateral Index Finger, Middle Finger Resorption**



**Figure 1(C): Fungal Culture: Left Index Finger Nail Clippings Send for Culture which Yielded *Trychophyton ajelloi***

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**Figure 1 (D): HPE Shows Dermis- Hypocellular Homogenous Collagen in papillary Dermis; Eccrine Glands Show Atrophy and are Lying Higher in Dermis Surrounded by Thick Sclerotic Eosinophilic Collagen Bundles. There is Sparse Perivascular & Periadnexal Inflammation**

### Conclusion

The main point of interest in this case is Limited Cutaneous Scleroderma with an unusual presentation of skin thickening distal to the metacarpophalangeal joint.

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