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Acquired Ichthyosis with Systemic Sclerosis: A Rare Case Report

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Abstract: ***Acquired ichthyosis (AI) is a condition accompanying many systemic illnesses such as lymphoma, sarcoidosis, systemic sclerosis, dermatomyositis and systemic lupus erythematosus (SLE). Systemic Sclerosis (SS) is an unknown autoimmune connective tissue disease of unknown etiology, characterised by symmetric hardening of the skin of the fingers, hands and face that may generalize. Internal organ involvement is frequent and affects the lungs, gastrointestinal tract, heart and kidneys. The co - existence of systemic sclerosis and AI has been very rarely reported worldwide. We describe a 27 - year - old male with systemic sclerosis who developed ichthyosis on his extremities and trunk. We also review the world literature on AI & SS.***

Keywords: AI, Acquired ichthyosis, Systemic sclerosis, Autoimmune connective tissue disease, Coexistence

1. Introduction

Ichthyoses are disorders of cornification in which abnormal differentiation and desquamation of the epidermis results in a defective cutaneous barrier. Acquired ichthyosis (AI) is an uncommon disease that usually appears in adulthood and is clinically and histologically indistinguishable from ichthyosis vulgaris. Systemic sclerosis (SSc, scleroderma) is an autoimmune connective tissue disease (AI - CTD) of unknown etiology that affects the skin, blood vessels and internal organs. It has two major clinical subtypes: cutaneous limited and cutaneous diffuse. Limited SSc is characterized by fibrotic skin changes that are limited to the fingers, hands and face. In diffuse SSc, generalized cutaneous sclerosis is seen that usually begins in the fingers and hands but eventually extends to the forearms, arms, face, trunk, and lower extremities.

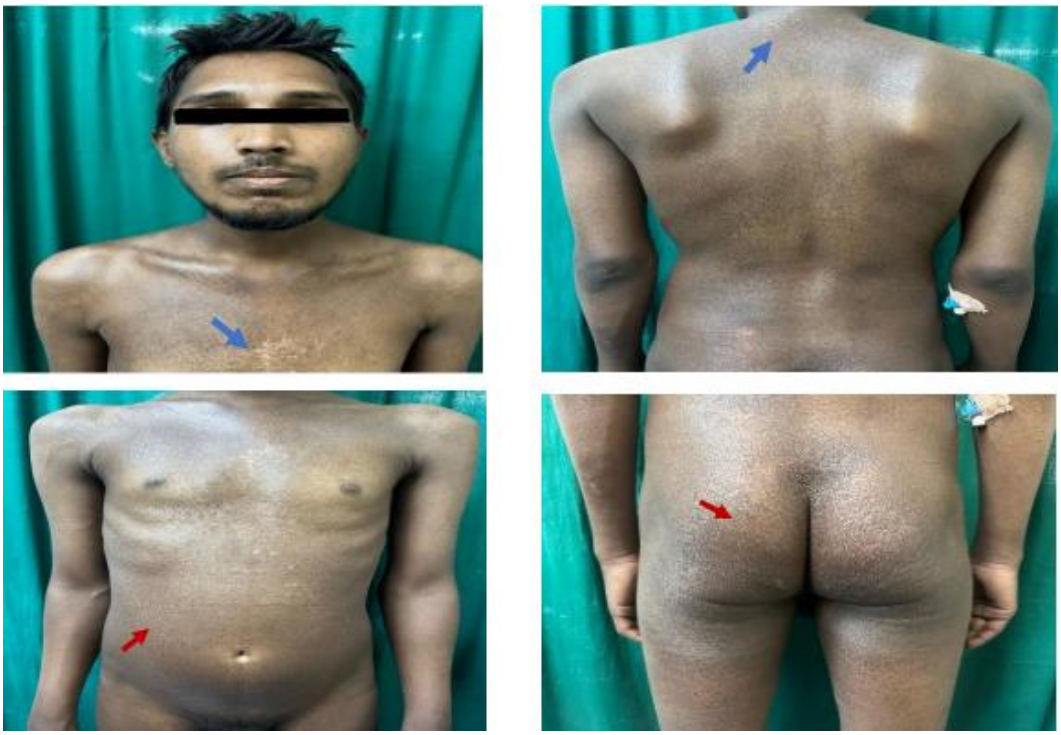
History

A 27 year old male presented with complaints of dryness and tightening of skin all over the body for past 2 months.

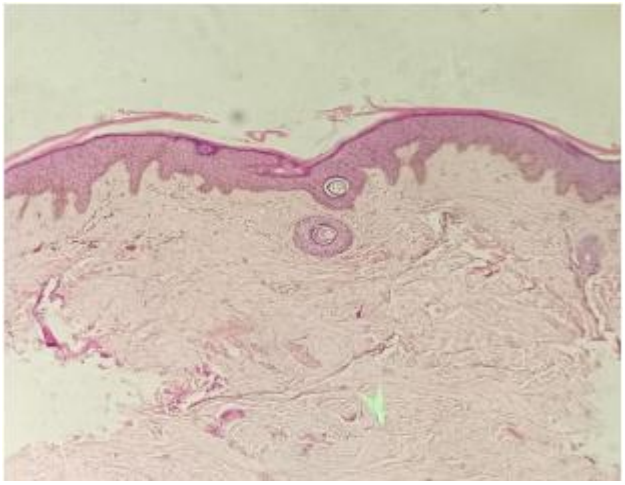
2. On examination

Inspection shows shininess and taughtness of skin all over the body, indicative of hideboundness with generalised xerosis along with multiple, ill defined ichtyotic patches in symmetric distribution over trunk, bilateral upper and lower limbs. Multiple, Ill defined patches of salt and pepper pigmentation were also observed over chest and upper back. Palpation revealed diffuse tightening of skin with Modified Rodnan's Skin Score calculated to be around 11. ANA Blot showed Strong Positivity (126) for Antibodies against Scl - 70. Histopathology of skin punch biopsy showed features suggestive of both Scleroderma and Ichthyosis. Pulmonary function test was suggestive of restrictive pattern of lung disease.

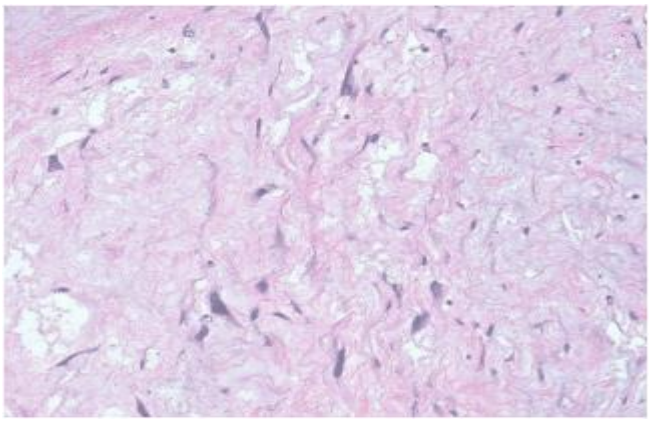
Patient was treated with Methyl Prednisolone pulse along with Pirfenidone and supportive treatment for 2 months, but he lost to follow up and had succumbed due to ARDS.



Clinical images showing generalised xerotic changes, Salt and pepper pigmentation (blue arrows) and ichthyotic patches (red arrows)



10x microscopic view of hpe slide shows:
Dermis showing excess collagen bundles (Suggestive of sclerotic changes)



40x microscopic view of hpe slide shows:
Magnified View of dermis showing highly packed eosinophilic collagen fibres

3. Discussion

Few papers have been published in the past regarding the relationship of Autoimmune Diseases and Acquired Ichthyosis, though the exact pathogenic mechanisms have not been successfully elucidated.

1) Williams et al (2010) reported three children with Ichthyotic - appearing Skin Changes Associated with Childhood Morphea, Systemic Sclerosis, and Systemic Lupus Erythematosus⁄Scleroderma Overlap and inferred that it might be an early feature reflecting underlying dermal sclerosis, predictive of more widespread cutaneous involvement, ultimately prompting earlier treatment.[1]
2) Lee at al (2006) described a 33 - year - old woman with an overlap syndrome consisting of systemic sclerosis and SLE who developed ichthyosis on her extremities and observed that this association of autoimmune diseases with Acquired Ichthyosis suggests that an abnormal immune response may be involved in its pathogenesis.[2]
3) Feola et al (2019) reported a patient who presented with two pathologies associated with Acquired Ichthyosis (hypothyroidism and MCTD) and postulated that MCTD is the main cause as the histological study revealed underlying sclerosis, and the patient developed synchronic MCTD symptomatology.[3]

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