International Journal of Current Advanced Research
ISSN: O: 2319-6475, ISSN: P: 2319-6505, Impact Factor: SJIF: 5.995
Available Online at www.journalijcar.org
Volume 7; Issue 2(B); February 2018; Page No. 9722-9723
DOI: http://dx.doi.org/10.24327/ijcar.2018.9723.1620

Research Article

A RARE CASE OF SCLERODERMA IN MALE

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A R T I C L E I N F O

Article History:
Received 14th November, 2017
Received in revised form 5th December, 2017
Accepted 3rd January, 2018
Published online 28th February, 2018

Key words:
Scleroderma, Esophageal dyskinesia, Connective tissue disorder

A B S T R A C T

Scleroderma is an autoimmune connective tissue disorder. This condition can be localized or systemic. It mainly involves skin, blood vessel, and visceral organs. The term "sclera" meaning hard "derma" meaning skin in Greek. The manifestations are due to diffuse deposition of collagen in the skin and internal organs along with vascular injury and immunologic abnormalities. Its estimated prevalence is 250 cases in a million and occurs rarely in males. Salt and pepper and hyde leather skin are typical findings for a patient with scleroderma. The aim of this case report is to present a 29-year-old male patient who came to our hospital with complaints of difficulty in swallowing since 2 weeks. On examination all systems were normal, and he had salt and pepper type of skin pattern post auricular region, hyde leather like skin, shiny and dry, pinched up nose. He was worked up for Connective tissue disorder and was found to have scleroderma with esophageal dyskinesia. However, atypical about this case is patient is a male and anti topoisomerase and anti centromere were negative.

INTRODUCTION

Systemic sclerosis (SSc) is a clinically heterogeneous generalized disorder affecting the connective tissue of the skin, blood vessel wall, and internal organs such as the gastro-intestinal tract, lungs, heart, and kidneys. It is characterized by alterations of the microvasculature, disturbances of the immune system, and by massive deposition of collagen in the connective tissue. Due to the complexity of the internal organ involvement, SSc has attracted attention from many disciplines, including rheumatology, pulmonology, cardiology, nephrology, gastroenterology, and dermatology. Many aspects of diagnosis and therapy require close cooperation between several disciplines.

CASE REPORT

29 year old male with no comorbidities came with complaints of difficulty in swallowing since 2 weeks. On examination he had salt and pepper type of skin pattern post auricular region, hyde leather like skin, shiny and dry, pinched up nose. All systems were within normal limits.

On examination**,** Skin over face, trunk, distal part of upper and lower extremities are taut-Hyde leather like skin, Salt and pepper skin appearance post auricular area bilaterally, trunk and left mid thigh, Pinched up nose, Scalp and hair normal, Pitting pattern in nail bed were seen. Other systemic examinations were normal.

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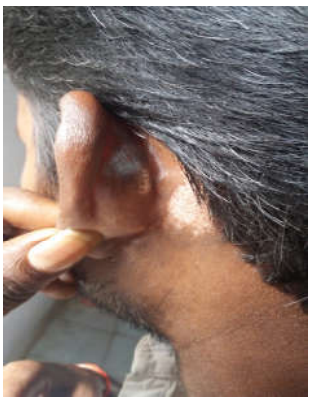
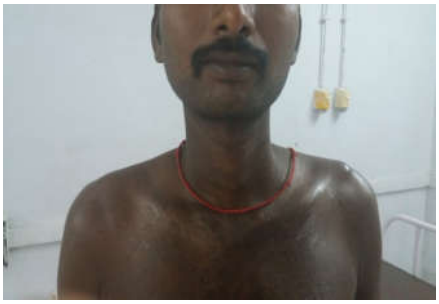
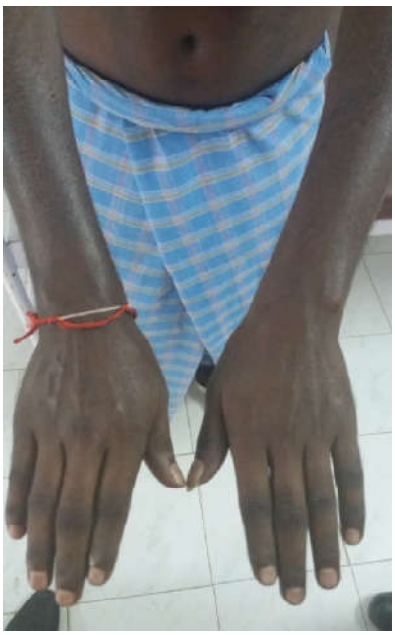
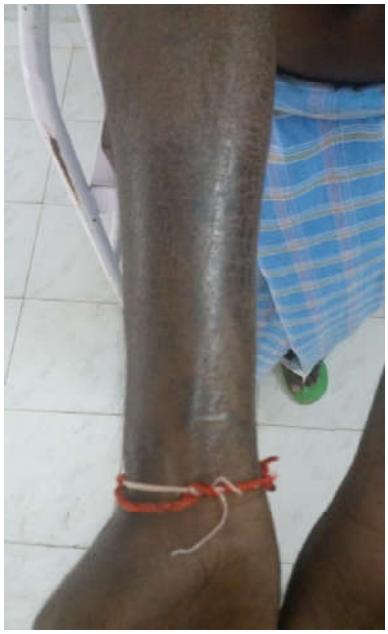
We went on to investigate this patient and he was found to have elevated ESR and CRP with Positive ANA (anti nuclear antibody); However anti topoisomerase and anti centromere were negative. RFT was normal. According to criteria of ACR we diagnosed him to have Systemic sclerosis.

He was treated with DMARD's (Methotrexate 15mg/week), PPI's, Topical emollients.









DISCUSSION

Scleroderma is an autoimmune disorder involving multiple systems thus making the course of this disease unpredictable. Though the correct nature of the disease is not known, high prevalence of circulating auto antibodies in serological investigations points it toward autoimmune mechanism.[1]

This condition can be localized or systemic. Systemic sclerosis (SSc) is a clinically heterogeneous generalized disorder affecting the connective tissue of the skin, blood vessel wall, and internal organs such as the gastro-intestinal tract, lungs, heart, and kidneys. It is characterized by alterations of the microvasculature, disturbances of the immune system, and by massive deposition of collagen in the connective tissue.

Due to the complexity of the internal organ involvement, SSc has attracted attention from many disciplines, including rheumatology, pulmonology, cardiology, nephrology, gastroenterology, and dermatology. The pathogenesis of scleroderma remains unclear but it is characterized by endothelial activation, immune system dysfunction and enhanced fibroblast activity. The endothelium controls the contraction and relaxation of vascular smooth muscle cells, leading to vasospasm and smooth muscle hypertrophy. Eventually it leads to obliteration of the lumen of small arteries and capillaries which leads to ischemia. There is extravasation of inflammatory cells initially predominated by monocytic lineage and later by lymphocytes.[2,3]

Diagnosis is usually clinical based on clinical manifestations. According to American college of rheumatology, Criteria for diagnosing is as follows[5]:

Criterion	Definition
major criterion	proximal scleroderma
or two or more of the following:	
minor criteria	1) sclerodactyly 2) digital pitting scars of fingertips or loss of substance of the distal finger pad 3) bilateral basilar pulmonary fibrosis
The proposed criteria had a 97% sensitivity for definite systemic sclerosis and 98% specificity.	

ANA, Anti centromere, Anti topoisomerase antibodies and their respective sensitivity and specificity for diagnosing systemic sclerosis is as given below.

	Sensitivity	specificity
Anti centromere	32%	>98%
Anti topoisomerase	40%	>95%
ANA	85%	54%

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How to cite this article:

Sankar R.N.K *et al* (2018) 'A Rare Case of Scleroderma In Male', *International Journal of Current Advanced Research,* 07(2), pp. 9722-9723. DOI: http://dx.doi.org/10.24327/ijcar.2018.9723.1620