DOI: 10.14260/jemds/2014/1928

CASE REPORT

LOCALIZED SCLERODERMA ASSOCIATED WITH KERATOCONJUNCTIVITIS SICCA: A RARE CASE REPORT

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HOW TO CITE THIS ARTICLE:
Sundip Shenoy, R. Jayaram, Reagan Madan, Venita J Noronha, Basavaraj Zalaki. "Localized Scleroderma Associated with Keratoconjunctivitis Sicca: A Rare Case Report". Journal of Evolution of Medical and Dental Sciences 2014; Vol. 3, Issue 04, January 27; Page: 941-946, DOI: 10.14260/jemds/2014/1928

ABSTRACT: Scleroderma is autoimmune connective tissue disorder which destroys healthy body tissues that causes widespread systemic and ocular features. A buildup of collagen in the skin and other organs leads to symptoms of the disease. It is of two types localized and diffuse. Systemic sclerosis has tendency to affect internal organs. It shows a non-mendelian pattern of inheritance with gene polymorphism.[1]Incidence of the disease is 9-19 cases per million[2]. Mortality is 30% with life threatening and visual threatening complications.[3] Occurrence of keratoconjunctivitis sicca in localized form of scleroderma is a quite a rare abnormality. We have presented a 38 year old female patient exhibiting characteristic features of localized scleroderma with keratoconjunctivitis sicca and then the literature is reviewed.

KEYWORDS: SCLERODERMA, KERATOCONJUNCTIVITIS SICCA (KCS), RAYNAUD'S PHENOMENON

INTRODUCTION: Scleroderma is a relatively rare connective tissue disorder of unknown etiology, heterogeneous clinical manifestations, chronic and progressive course. This is a disorder in which body destroys own healthy tissues leading to skin thickening, spontaneous scarring, blood vessel disease, various ocular manifestations and varying degrees of inflammation. There is hardening and sclerosis of the skin due to hypertrophy and subsequent atrophy of connective tissue of the dermis with fibrous tissue replacing subcutaneous fat. There is fibrinoid degeneration and perivasculitis of the blood vessels. Localized form of scleroderma has absent visceral involvement.

This disorder is more common in blacks, females in age group of 30-50 years. Cases of skin disease similar to scleroderma can be found in the writings of Hippocrates as far back as 460–370 B.C. Oribasius (325–403 A.D.) and Paulus Aegineta (625–690 A.D.) also wrote on the subject. The first definite description of the disease, by Carlo curzio was published in Naples in 1753.In 1964 Winterbaur applied CREST term to this localized scleroderma

CASE REPORT: A 38 year old married female patient presented to our OPD with symptoms of tightening sensation around the eyelids and face for last 9 months along with generalized stiffness of the skin elsewhere. She also complains of inability to open the mouth fully during the same period. Symptoms pertaining to dry eye like grittiness, foreign body sensation and eye tiredness were evaluated using MacMonnies's and OSDI dry eye questionnaire.[4]

In the case described above following changes were seen.

ON GENERAL AND SKIN EXAMINATION revealed shiny waxy appearance of the face with obliteration of skin folds, bird like facies(photo 1a), pinched beak appearance of the nose(photo 1b), decreased oral aperture(photo 1c), abnormal tautness of the skin with hyperpigmentation (photo 1d), Raynaud phenomenon of the extremities with nail fold infarcts(photo 2a, b, 3a).Other clinical

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signs pertaining to systemic involvement of internal organs was unremarkable thus pointing towards localized scleroderma.[5]

OCULAR EXAMINATION (BOTH EYES)

1. Visual acuity (without glasses):	Right Eye	Left Eye
	6/6	6/6

Anterior segment:

1. Loss of plasticity of the lids with difficulty in lid retraction due to thickening of the lids.
2. Absence of lagopthalmos.
3. Punctal ectropion
4. Decreased tear meniscus
5. Conjunctival filaments (dry eye)(photo 4)
6. Corneal filaments(dry eye) (photo 4)
7. Shallow fornices revealing foreshortening
8. Lens normal
9. Fundus examination features suggestive of hypertensive retinopathy was not found thereby ruling out systemic involvement of the disease (photo 5a, 5b).

Other features like peripheral ulcerative keratitis, cataract, glaucoma, vitreous body frost, papilledema were absent.

	Right Eye	Left Eye
TEAR MENISCI	0.2mm	0.2mm
TBUT	4sec	4sec
SCHIRMER'S	<10	<10
RBT	positive	positive (staining of filaments) (PHOTO 4)

INVESTIGATIONS:

1. ROUTINE BLOOD INVESTIGATIONS.(TLC, DLC ESR)
2. ECG- NORMAL
3. URINE ROUTINE- NORMAL
4. RENAL FUNCTION TESTS- NORMAL
5. AUTOANTIBODY(ANTI SCL 70)- NEGATIVE
6. SKIN BIOPSY- Section studied shows epidermis and dermis (photo 6a, 6b).
   A. Epidermis lined by stratified squamous epithelium with mild hyperkeratosis.
   B. Dermis composed of structure collagen bundles and adipose tissue.
   C. Mild lymphoplasmacytic infiltrate is seen between collagen bundles and around adnexal structures.

DISCUSSION: Though etiology remains inconclusive still genetic, environmental factors and occupational risk factors have been quoted like human cytomegalovirus, human parvovirus B19, toxic oil syndrome, drugs pentazocine, bleomycin, cyclophosphamide.[6- 9]

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Autoantibodies to extracellular matrix microfibrillar protein fibrillin 1 has been found in patients of localized scleroderma leading to skin collagen changes with thickening.[10]Further studies have implicated fibroblast responsible for increased collagen production in localized scleroderma. Studies have proved telangiectasia with capillary abnormalities and Raynaud's phenomenon playing a role in scleroderma.[11]

Scleroderma is classified as localized and widespread (systemic sclerosis with a tendency to affect internal organs).Two forms diffuse and limited cutaneous based on the skin involvement.

Diffuse is associated with progressive skin induration which starts in the fingers ascending from the distal to proximal involving face and trunk. It causes early fibrosis.

Limited has associated with long standing Raynaud's phenomenon. It is slowly progressive limited to fingers (sclerodactyly), distal extremities and face, not involving trunk. Subset of patients have CREST syndrome.

Cardinal features of the syndrome involve vasculopathy, cellular and humoral autoimmunity, progressive visceral and vascular fibrosis in multiple organs.

Pathology includes autoantibodies causing vascular injury with release of endothelin. This leads to obliterative vasculopathy, tissue hypoxia and ultimately fibrosis.[12, 13]

Role of CD4+ CD8+, B and T cells, activated monocyte, macrophages leading to release of cytokines and chemokines that leads to abnormal fibroblast activation which causes fibrosis of the organs, fibrosis and inflammation of conjunctival glands that leads to defective vasculogenesis.[14]

Pathology hallmarks include combination of widespread capillary loss and obliterative vasculopathy of small arterioles with fibrosis of skin and internal organs. Multiple genetic loci is suspected.[15, 16]

SYSTEMIC EFFECTS: Cardiac effusions and serositis, lung fibrosis, esophageal fibrosis, kidney nephritis were absent pointing to the localized nature of the disease.

MANAGEMENT: Diagnosis and management of keratoconjunctivitis sicca (KCS) is a must in all cases.[17, 18] Till date there is no proven therapy for the treatment of scleroderma. Symptomatic based management depends upon which organ is involved which constitutes the following.

MEDICAL

1. DISEASE MODIFYING AGENTS.—cyclophosphamide and steroids.
2. ANTIFIBROTICS---D-penicillamine, interferon gamma
3. VASCULAR THERAPY—calcium channel blockers
4. GIT-broad spectrum antibiotics

SKIN –antihistaminic, prednisolone

OCULAR–primarily tear substitutes

In addition to above modalities use of hypomellose, hydroxyl methyl cellulose, polyvinyl alcohol, sodium hyaluronidase is prescribed.

DENTAL –mouth rinses to replace minerals, floss, visiting dentist for cleaning.

SUMMARY: A case of 38 year old female patient having localized scleroderma with keratoconjunctivitis sicca has been described. One must look at the psychosocial aspects as

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scleroderma is a disfiguring disease that alters all aspects of patient's quality of life and thus has a deep psychological impact in the patient. Depression and distress about the disfigurement is common and require psychological counseling and intervention.

Photo 1A- BIRD FACIES

Photo 1B- PARROT BEAK APPEARANCE OF NOSE

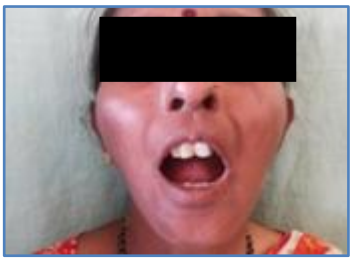

Photo 1C- MOUTH WITH REDUCED ORAL APERTURE

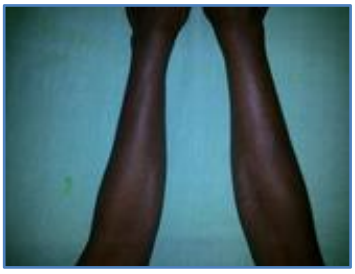

Photo 1D- FOREARM SHOWING HYPERPIGMENTATION

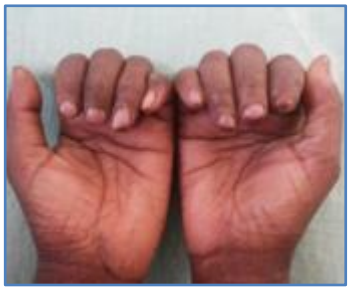

Photo 2A and 2B- HANDS SHOWING NAIL INFARCTS

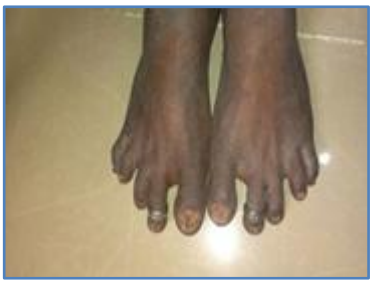

Photo 3A- FEET SHOWING NAIL INFARCTS

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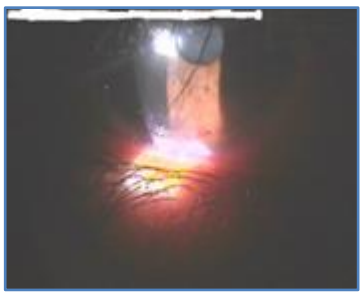
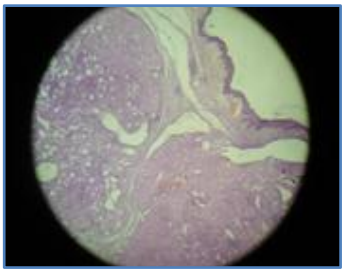


Photo 4- SLIT LAMP SHOWING CONJUNCTIVA AND CORNEAL FILAMENTS

Photo 5A- RIGHT EYE FUNDUS

Photo 5B- LEFT EYE FUNDUS



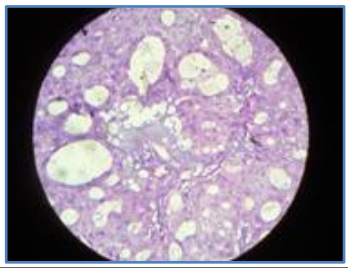


Photo 6A & 6B- HISTOPATHOLOGY SLIDE SHOWING SCLERODERMA CHANGES

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Date of Submission: 03/01/2014.
Date of Peer Review: 04/01/2014.
Date of Acceptance: 16/01/2014.
Date of Publishing: 23/01/2014.