Original Research Paper Ophthalmology

Severe DRY Eye Disease in A Case of Scleroderma- A Case Report

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ABSTRACT

Scleroderma is a generalized connective tissue disease of unknown origin with heterogeneous manifestations. ocular manifestations of scleroderma are characterized by marked heterogeneity. Eyelid skin abnormalities and kerato-conjunctivitis sicca were the most common SSc-related ocular findings. A 50 years old married female patient presented to our OPD with symptoms of grittiness, foreign body sensation and eye tiredness in both her eyes for 1 year. Her Tear film Break up time in right and left eye were 3 sec, 2 sec ;Schirmer's test I were 5 min, 3 min respectively. Skin biopsy showed extensive collagenisation within skin tissue.

KEYWORDS Scleroderma, Kerato-conjunctivitis sicca, Tear film Break up time, Schirmer's test I, Skin biopsy

INTRODUCTION

Systemic sclerosis (SSc) is a generalized connective tissue disease of unknown origin with heterogeneous manifestations.1

In the pathophysiology of SSc, three abnormalities have been distinguished: 1) a fibroblast dysfunction leading to increased deposition of extracellular matrix; 2) a vascular abnormality resulting in tissue hypoxia; and 3) an immune response, manifested as altered T- and B-lymphocyte function and autoantibody production.2-5

Systemic involvement in form of cardiac effusions and serositis, lung fibrosis, esophageal fibrosis, nephritis along with generalised stiffness of skin.6

The ocular manifestations of SSc are characterized by marked heterogeneity. Eyelid skin abnormalities and KCS were the most common SSc-related ocular findings.7

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 and inflammation of conjunctival glands.8 This leads to qualitative and quantitative alteration of tear production leading to severe dry eye disease.

CASE REPORT

A 50 years old married female patient presented to our OPD with symptoms of grittiness, foreign body sensation and eye tiredness in both her eyes for 1 year. She also complained of tightening sensation around the eyelids and face for last 10 months along with difficulty in respiration for same period.

OCULAR EXAMINATION (BOTH EYES)

Visual acuity:

Unaided → 6/12, 6/12

BCVA → 6/6, 6/6

B. Ocular adenexa:

1. Decreased plasticity of the lids with difficulty in lid retraction , woody texture on palpation

2. Absence of lagopthalmos.

3. Shallow fornices

C. Ocular surface assessment:

Parameter	R/E	L/E
TBUT	3 sec	2 sec
TEAR MENISCI HEIGHT	0.3 mm	0.2 mm
SCHIRMER'S I	5 mm	3 mm

D. Others: WNL

Fig-I: Schirmer's Test

INVESTIGATIONS:

1. ROUTINE BLOOD-
TLC- 9,000 /cumm
DLC-N85L08M06E01B00
ESR-85 mm in 1st Hr
Hb- 8.4 mg/dl
Others- WNL

2. ECG- NORMAL
3. URINE ROUTINE- NORMAL
4. RENAL FUNCTION TESTS- NORMAL
5. SKIN BIOPSY- Section from the tissue shows extensive collagenisation within skin tissue.

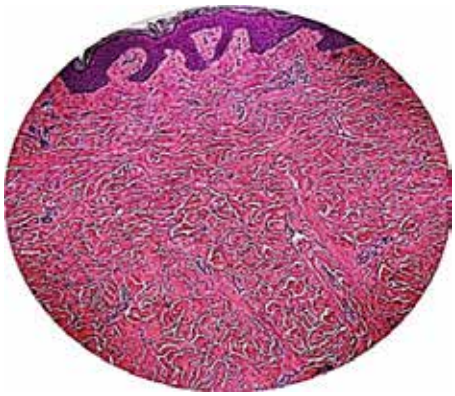

Fig-II: Skin biopsy

6. HRCT THORAX:
a. Interstial septal thickening with few tiny cystic spaces in sub-pleural location in anterior segment of right upper lobe-suggestive of early signs of interstial lung disease

b. consolidation with air bonchogram in bilateral basal segment with ground glass appearance of both upper lobe with bilateral pleural effusion.

TREATMENT:
Ocular:
1. Polyvinyl alcohol eye drop – frequently B/E

2. Loteprednol Etabonate eye drop- 4 times daily B/E

3.Hydroxy propyl methyl cellulose eye ointment- at bed time B/E

Systemic: Infusion Cyclophosphamide
Intravenous Antibiotic
Nebulization

DISCUSSION:
The most strikingly apparent ocular manifestation of scleroderma is the fibrotic change seen in the eyelids.9 Skin fibrosis is the hallmark of scleroderma and is defined as excess deposition and accumulation of extracellular matrix, mainly type I collagen in the dermis.3 Eyelid skin changes show a prevalence ranging from 29 to 65% of scleroderma patients.10 Depending on the study, the prevalence of scleroderma patients with KCS varies considerably (37 to 79%).11 The observed variation may be partly due to the KCS definition used and partly due to demographic differences in the patient populations. Studies have shown correlations between sicca syndrome and anti-Ro/SSa antibodies in patients with scleroderma.12 Anti-Ro/SSA antibodies were not investigated in our case due absence of proper infrastructure. The relation between anti-Ro/SSA antibodies and KCS in scleroderma patients should be investigated further.

CONCLUSIONS:
Scleroderma is an auto-immune disease involving multi-organ including lacrimal gland and conjunctival glands. Conservative treatment in the form of lubricating agents are treatment of choice. Although no cure has been found for scleroderma, the disease is often slowly progressive and manageable, and people who have it can lead healthy and productive lives.

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