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Original Research Article

A Study of Ocular Manifestations of Scleroderma in Tertiary Care Centre

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Abstract

Aim: This study was aimed at determining the ocular manifestations of patients presenting with clinical features suggestive of scleroderma in dermatology department in a tertiary care centre.

Settings and Design: Tertiary care referral centre in Tamilnadu, Prospective, Single institution, interventional case series.

Materials and Methods: A prospective cross sectional review of patients presenting with clinical features suggestive of scleroderma, in dermatology department, in a tertiary care centre in Tamilnadu was conducted between Nov 2017 to Feb 2019. Fifty patients with systemic clinical features suggestive of scleroderma were included. Investigations done and systemic associations were confirmed. Ocular investigations including visual acuity, intra ocular pressure, Slit lamp examination, colour vision, visual fields, Schirmer's test, Tear Film Break Up Time and fundus examination were done in all cases.

Results: Among fifty cases 45 were females (90%) and 5 were males (10%) and the mean age was45±15 years. Slit lamp examination and oblique examination revealed inability to retract the lower eyelid (Ingram's sign) in 40 out of fifty cases due to tightening of lid skin. Schirmer's test and TBUT revealed moderate to severe dry eye in 25 out of 50 cases. Colour vision, fields and fundus were normal in all cases.

Conclusion: Eyelid abnormalities and defective tear secretion were the most common ocular findings in patients with scleroderma.

Keywords: Scleroderma, Systemic Sclerosis, Lid Retraction, Dry Eye.

Introduction

Systemic sclerosis (scleroderma) is a connective tissue disorder of unknown etiology with varied manifestations. The incidence is about 9 to 19 cases per million per year in the united states[1] .Due to the rare nature of the disease most papers have been single case reports or small case series. The disease affects multiple organ systems in the body. It is of two types.

Diffuse systemic sclerosis and limited systemic sclerosis. The various ocular manifestations reported are eyelid skin changes, keratoconjunctivitis sicca and primary SSc retinopathy. Eyelid skin changes were most frequent in patients with the diffuse type of systemic sclerosis .The purpose of this study was to evaluate the frequency and characteristics of ocular manifestations in patients with systemic sclerosis.

Inclusion Criteria

- Patients with systemic features of scleroderma
- Patients with ocular manifestations of scleroderma
- Patients with both systemic and ocular features of scleroderma

Exclusion Criteria

- Patients without systemic features of scleroderma
- Patients without ocular features of scleroderma

Materials and Methods

A prospective cross sectional study was conducted in a tertiary care centre in Tamilnadu from November 2017 to Feb 2019 on fifty patients diagnosed with scleroderma visiting Dermatology department. Data regarding age, gender, disease duration, age at diagnosis, systemic corticosteroid or chloroquine use, ocular symptoms and detailed ophthalmic history were recorded.

Investigations: Distance visual acuity was measured with Snellen's chart and near vision with Jaeger's chart. Intra ocular pressure was measured with Goldmann's applanation tonometer. Dry eye evaluation was done in the following order: 1. Tear Film Break Up Time 2.Schirmer's test. Tear Film Break up Time was measured after Fluorescein instillation into the conjunctival sac. Schirmer's test was performed by keeping a 5x35 mms Schirmer's strip for 5 minutes without applying topical anaesthesia in the lower fornix at the junction of medial 2/3 and lateral 1/3 of lower eyelid.

Indirect ophthalmoscopy was performed with an indirect ophthalmoscope and +20 dioptre lens after both pupils were dilated with 1% tropicamide and 5% phenylephrine.

Result Analysis

Age Distribution: The age of patients ranged from 30-60 years.

Table 1: Age distribution

Age Group	No of Patients	Percentage
30-40 years	17	34%
40-50 years	17	34%
50-60 years	16	32%

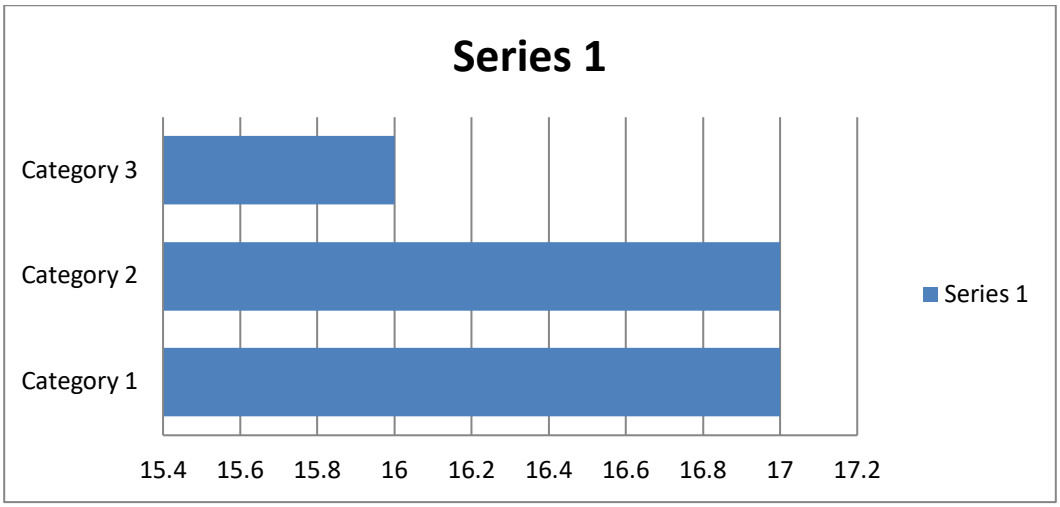

Figure 1: Series 1

Sex Distribution: Our study showed a female preponderance of 90%

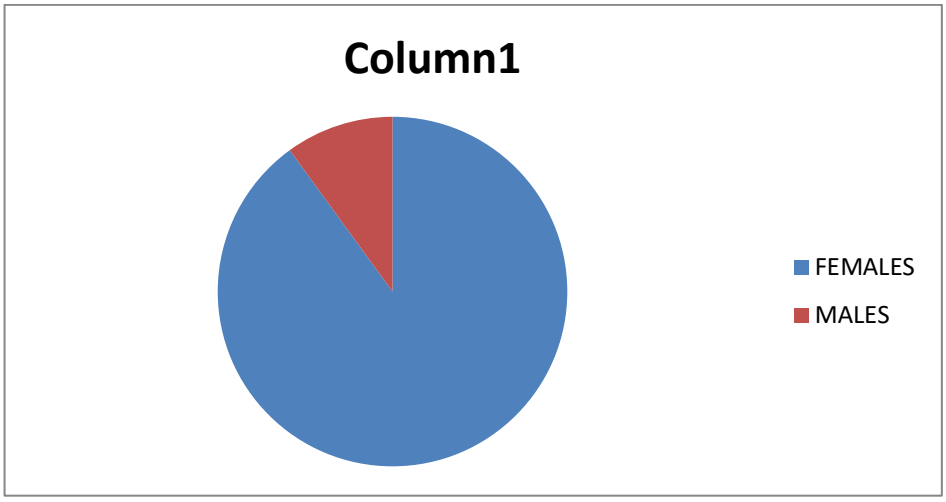

Figure 2: Column 1

Results

The most common systemic presentation was pain in hands or legs (100%) especially in cold climates.

The most common ophthalmic manifestation is inability to retract the lower eyelid (Ingram's sign)

(80%) followed by moderate to severe dry eye (50%).

Discussion

Scleroderma is a disorder of unknown cause in which there is localized sclerosis of the skin [1].

The condition has traditionally been subdivided clinically into the following types [2]:

1. Circumscribed plaques
2. Morphea profundus/subcutaneous (deep)
3. Bullous morphoea
4. Linear morphoea
5. Frontoparietal lesions (en coupe de sabre), with or without hemiatrophy of the face

Etiology: The cause of scleroderma is unknown[3]. It is an autoimmune condition, in which the body's immune system attacks healthy tissues [3]. Strong associations with certain mutations in HLA genes have been identified [4,5]. Strong environmental influences have also been implicated in the etiology of scleroderma [6,7].

Pathology: 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 [8-11].

Incidence: All ages are affected, the peak incidence occurring between 20 and 40 years of age, although 15% begin below the age of 10 years [12,13] in which age group the linear lesions predominate. The female to male ratio is around 3:1 in most studies.

Clinical Features

Plaque lesions: These occur as indurated areas of skin, which at first are faintly purplish or mauve in colour. After some weeks or months they lose their colour, especially in the centre, and appear as thickened wavy areas, which are ivory in colour, with a characteristic lilac-coloured edge. The surface is usually smooth and shiny but may be nodular14.

Linear lesions: The limbs are frequently affected, the legs more than the arms. They may also occur on the anterior aspect of the thorax, and sometimes the abdomen or buttocks are affected.

Frontoparietal lesions: 'en coupe de sabre' (from its resemblance to a sabre cut), with or without hemiatrophy. These lesions usually start with contraction and firmness of the skin over the affected area. The corresponding side of the tongue may be atrophic although sometimes the lesion is in the midline of the tongue.

Not infrequently, there is atrophy of the corresponding part of the face and cheek, with facial asymmetry, and this usually occurs within a year. A variety of ocular lesions occur [15], including enophthalmos, involvement of the lids, oculomotor muscles, iris and fundus, myopathy of external eye muscles [16] and vasculitis [17].

Atrophy of the nasal part of the iris and loss of cilia on the upper eyelid followed exactly the line of the skin lesions in one case [18]. Heterochromia of the iris also occurs [19]. 'En coup de sabre' morphoea has presented as unilateral eyelid edema [20]. Ossification occasionally occurs [21]. Other ocular manifestations are eyelid abnormalities like stiffness and telengiectasia, defective tear secretion and conjunctival abnormalities.

The organs affected are skin, lungs, gastro intestinal tract, kidneys, heart, joints (synovium), tendons, muscles, thyroid and minor salivary glands.

American college of rheumatology diagnostic criteria for systemic sclerosis

Major Criteria

1. Proximal sclerodermatous skin changes (Proximal to the metacarpophalangeal joints)

Minor Criteria

1. Sclerodactyly
2. Digital pitting scars of fingertips
3. Bibasilar pulmonary fibrosis

Patient should fulfill the major criterion or two of the minor criteria [22].

Most striking ocular manifestation of systemic sclerosis is the fibrotic change in the eyelids.Skin fibrosis is the hallmark of SSc. It is defined as excess deposition and accumulation of extra cellular matrix mainly type 1 collagen in the dermis [9]. Tailor et al. have shown that skin fibrosis in SSc patients can result in a spectrum of clinical alterations ranging from lid stiffness or tightness to blepharophimosis or lagophthalmos [23] Findings regarding eyelid skin changes in previous studies showed prevalence ranging from 29-65%. In our study it was 100%.

Conclusion

Eyelid abnormalities and defective tear secretion were the most common ocular findings in patients with systemic sclerosis.

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