

Ocular Involvement in Connective Tissue Disorders in a Tertiary Hospital

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Abstract: Connective tissue disorders are systemic auto immune disorders. Ocular involvement is reported frequently in these diseases. **Aim of the study**—To assess and review the ocular involvement in Connective tissue disorders. **Methods**— This is a hospital based Retrospective observational case series analysis of patients. Clinical data regarding connective tissue diseases (CTD) and full Ophthalmic examination were extracted from medical records of the Medicine & Ophthalmic departments of Government general Hospital, Guntur. **Results**—In 76 Rheumatoid Arthritis patients examined, Dry Eye was 40%, Drug induced complications are also noted. In S.L.E (25) patients, Dry eye syndrome was 20%, Steroid induced Glaucoma 4%, steroid induced cataract 7% and retinopathy 7%.

Keywords: Collagen disease, Rheumatoid Arthritis (R.A), Systemic Lupus Erythematosus(S.L.E), Ocular manifestations, Dry eye syndrome.

1. Introduction

Connective tissue disorders are systemic autoimmune diseases. The term collagen disease was coined by **Paul Klemperer** at Mount Sinai hospital in New York city in 1941. Females are most likely to be affected than males and the prime age of onset is 30-50 years. People of all ages may be affected. Often connective tissue disorders are associated with conjunctivitis, dry eye syndrome keratoconjunctivitis, episcleritis, scleritis, uveitis, steroid induced glaucoma, steroid induced cataract, retinopathy and optic neuropathy.

2. Patients & Methods

This is a hospital based Retrospective observational case series analysis of patients. 101 patients with CTD attending GGH, Guntur, between March 2014-FEB 2015 are included in the study. Data regarding age, gender, antibody profile, corticosteroid therapy or chloroquine therapy, ocular symptoms were recorded from the patients files. All patients underwent a complete ophthalmic examination included visual acuity, slit lamp examination, measurement of intraocular pressure, fundus examination, and dry eye syndrome, diagnosis using Schirmer test.

3. Discussion

	Total cases	females	Males
R.A	76	57	19
S.L.E	25	23	2

R. A - Results

Dry eye	40%
Uveitis	2%
Episcleritis	5%
Scleritis	3%

S.L.E - Results

Dry eye	20%
Scleral changes	15%
Steroid induced glaucoma	4%
Steroid induced cataract	7%
Retinopathy	5%
Optic Neuritis	1%

Rheumatoid Arthritis (RA) is the most common rheumatic disorder affecting approximately 1% of adults. RA is classically an additive Symmetric, deforming, peripheral polyarthritis characterized by synovial membrane inflammation. All joints may be involved but this disorder affects primarily the small joints of the hands and feet. Approximately 80% of patients with RA are positive for a rheumatoid factor, which is an autoantibody directed against immunoglobulin G (IgG). Human leukocyte antigen DR4 (HLA-DR4) is found in 70% of white seropositive patients. A more recent test involves identification of anticyclic Citrullinated peptide (antiCCP) antibodies (1).

Dry eye is the most common ophthalmic manifestation of RA; with a reported prevalence of 15-25%. Lacrimal and salivary gland secretion are significantly reduced in patients with RA. While it is well known that dry eye is frequently associated with RA, the correlation between dry eye severity and the activity of RA is unclear. Sjögren's syndrome commonly accompanies RA. The full triad of Sjögren's syndrome consists of KCS (dry eyes), xerostomia (dry mouth), and a connective-tissue disorder, usually RA. Episcleritis and scleritis are classically described in patients with RA. Episcleritis is sudden in onset, and the patient complains of discomfort rather than pain. Attacks last 1 to 2 weeks and are self-limiting but recur at intervals of 1 to 3 months. The prevalence of episcleritis in RA is reported to be about 0.17%, and in patients who present with episcleritis, about 5.6% have

RA.⁽³⁾. The prevalence of scleritis in RA is reported to be 0.67% to 6.3%, Initial therapy includes oral indomethacin or other nonsteroidal anti-inflammatory drugs (NSAIDs). Patients who do not respond to these medications should be treated with topical steroids or systemic immunosuppressive medications.

Other, less common ocular manifestations of RA include uveitis 2-4%(5), episcleral nodules, (5)Posterior subcapsular cataracts in patients of rheumatoid arthritis who were on long term oral steroids has been well documented(6).

S.L.E-The first use of the term “lupus,” which is Latin, for wolf, is attributed to the 12th-century physician **Rogierus** to describe the classic malar rash that resembles the pattern of fur on a wolf's face. Systemic lupus erythematosus (SLE) is a chronic systemic autoimmune disease (1).Seen in women of child bearing age. Revised criteria for diagnosis – at least 4 out of 11 criteria described (8).ocular manifestations are seen in 1/3 of patients. SLE may cause ocular disease by mechanisms including immune complex deposition and other antibody related mechanisms, vasculitis and thrombosis.(6) Immune complex deposition has been identified in blood vessels of the conjunctiva, retina, choroid, sclera, ciliary body, in the basement membranes of the ciliary body and cornea, in the peripheral nerves of the ciliary body and conjunctiva apart from throughout the body[6]. Antibody dependent cytotoxicity may cause retinal cell death and demyelination of the optic nerve. Cutaneous manifestations of eyelids (DLE), orbital disease, kerato conjunctivitis sicca (20% prevalence), PUK, scleral inflammatory disease, uveitis, neuroophthalmic lesions, retinal vasculopathy(6). SLE – Lupus retinopathy-prevalence is 10% (6). Cotton wool spots with/without intra-retinal haemorrhages, Severe retinal vascular occlusive disease (7). Posterior scleritis can occur in patients with SLE and may sometimes be the presenting sign of systemic disease. . Choroidopathy in SLE may present as multiple, serous detachments of the retinal pigmented epithelium and the neurosensory retina. Visual loss can occur if the detachment affects the macula.(7) In some cases, the subretinal fluid can progress to large, bullous exudative retinal detachments. Treatment of underlying active disease often results in resolution of the choroidopathy and associated subretinal fluid. Suspected pathogenesis may be due to a microangiopathy caused by a mononuclear inflammatory infiltrate of the choroid, immunoglobulin and complement deposition in the choroidal blood vessels and damage to the overlying RPE.(6)(7) Optic neuropathy prevalence in SLE is 0.7%.Reported ocular findings may include optic neuritis, ischemic optic neuropathy, retrobulbar optic neuropathy and optic atrophy, and optic nerve disease can affect one or both eyes.(6).

Managing posterior segment disease typically involves the use of systemic immunosuppression with corticosteroids in the acute setting, followed by non-corticosteroid immunosuppressive agents. For severe posterior segment involvement, intravenous corticosteroid pulse therapy may be needed acutely.(5) The agents used in the treatment of

SLE can themselves cause significant ophthalmic morbidity. Corticosteroids are commonly used in SLE and may cause cataract formation usually with topical compared to systemic.(6) Hydroxychloroquine can cause reversible visually insignificant changes in the cornea (vortex keratopathy) and, more importantly, an irreversible sight-threatening maculopathy.(6)

4. Conclusion

Eye manifestations in CTD may be sight-threatening, can be an indicator of active systemic disease or at certain times present as an initial symptom. Early recognition by the rheumatologist, prompt assessment by the ophthalmologist and coordinated treatment strategies are key in reducing the ocular morbidity associated with this disease.



Figure 1: Malar rash



Figure 2: Scleral thinning



Figure 3: Chronic Uveitis in C.T.D

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