Original Research Article

Ocular manifestations in systemic lupus erythematosus

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Abstract

Aim: To report the significance of ocular manifestations in Systemic lupus erythematosus **Materials and methods:** Seven diagnosed cases of Systemic lupus erythematosus, which included both newly diagnosed cases and cases already on treatment, were referred from the treating physician for ophthalmological evaluation. Comprehensive ophthalmic examination was done using BCVA, intraocular pressure measurement, Slit lamp examination of the anterior segment, fundus examination using Slit lamp biomicroscopy and indirect ophthalmoscopy, and Schirmers testing was done. In selected patients, certain special investigations like visual fields 24-2, 10-2 testing, SD-OCT was done. However ERG, couldnot be done due to lack of resource availability. Patients with history of Diabetes mellitus, Hypertension and other ophthalmic diseases like age related cataract were excluded. **Results:** Out of the seven cases of SLE examined, xerosis was the most commonly encountered manifestation, seen among 6 patients. Episcleritis was seen in one patient, Anterior uveitis seen in 2 patients, SLE associated retinopathy was seen in 3 patients. Treatment induced side effects like glaucoma was seen in 1 patient, posterior subcapsular opacities in 2 patients and Bull's eye maculopathy suggestive of HCQ toxicity seen in 1 patient. While dryness, episcleritis and retinopathy were managed conservatively, Anterior uveitis was managed with topical steroids. Patients with HCQ associated toxicity were advised to stop the HCQ therapy or to go for an alternative regimen, those with posterior subcapsular opacities were advised cataract surgery, while glaucomatous patients were started on anti glaucoma therapy. **Conclusion:** Early diagnosis, prompt referral, coordinated treatment and long term follow-up through multi disciplinary approach involving Ophthalmologists, Rheumatologists and Dermatologist play a key role in reducing ocular and systemic morbidity associated with Systemic lupus erythematosus.
Key Words: Hydroxychloroquine (HCQ), systemic lupus erythematosus (SLE), Uveitis.

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INTRODUCTION

Systemic Lupus Erythematosus (SLE) is an autoimmune disease with multiple organ system involvement. ocular manifestations of SLE was first described in 1929. In 1933 Semon and Wolff, described the histopathological features of choroiditis and subretinal exudation.[1,2] The prevalence of SLE is 20–150 cases per 100 000. Male to female ratio is 1:9. [1,3] According to the American College Of Rheumatology, SLE is diagnosed based on the presence of 4 out of 11 criteria either in the present or past. malar rash, discoid rash, photosensitivity, oral ulcers, non-erosive arthritis, serositis, renal disorder, neurological disorder (seizures or psychosis), haematological disorder (anaemia, leucopenia, thrombocytopenia), immunological disorder (anti-DNA antibody, anti-Sm antibody and false positive Venereal Disease Research Laboratory testing) and presence of antinuclear antibodies.[4] Ocular manifestations, although not included in the diagnostic criteria, are seen in one third of patients of SLE. Once affected, ocular lesions in SLE cause significant morbidity. These can also be a marker for overall systemic activity[4]. In addition, medications used to treat SLE can lead to vision threatening complications like cataract, glaucoma, and

chloroquine maculopathy. SLE can involve every ocular structure leading to significant visual loss or blindness if left untreated.

Pathophysiology of ocular disease in SLE: The pathogenesis of lesions is multifactorial and complex. Auto antibodies, immune complexes which trigger inflammation by activation of compliment system cause multiorgan damage.

- Immune complex deposition and other antibody related mechanisms cause vasculitis, and thrombosis[5]. Immune complexes have been identified in blood vessels of the conjunctiva, retina, choroid, sclera, ciliary body, and in the basement membranes of the ciliary body, cornea, in the peripheral nerves of the ciliary body and conjunctiva[1].
- Antibody dependent cytotoxicity leads to retinal cell death and demyelination of the optic nerve. Secondary Sjogren's syndrome is a result of antibody dependent cytotoxicity in the lacrimal gland with consequent dry eyes (kerato conjunctivitis sicca) due to inadequate tear production.
- Corticosteroids used in the treatment of SLE cause failure of osmotic regulatory systems due to Na, K-AT Pase inactivation, leading to increased membrane leakiness or increased osmolality. This gives rise to localised accumulation of water and fluctuations in the refractive index, leading to scatter of light. Free radicals produced cause membrane and protein damage leading to in solubilisation of lens proteins.

SLE can involve all the ocular structures. Kerato conjunctivitis sicca is the most common ocular manifestation whereas the retinal lesions are vision threatening. ocular involvement in orbit is in the form of myositis[6], panniculitis. Patients may present with proptosis (painful /painless) ptosis, ocular movement restriction. Eye lids may show discoid lupus like rash. Keratoconjunctivis sicca occurs due to involvement of lacrimal glands. Corneal lesions include superficial punctuate keratitis and rarely a peripheral ulcerative keratitis like picture. Episcleritis or the more severe necrotizing scleritis, iridocyclitis are the other anterior segment manifestations.[7,8] Posterior segment complications include retinopathy which is immune complex mediated. It may manifest in milder form as microangiopathy (cotton wool spots, microaneurysms, hard exudates, dor haemorrhages) or severe vaso occlusion due to ischaemia resulting from occlusion of major central retinal vessels, cilio retinal vessels. vasculitis is another manifestation of SLE in retina[9,10]. Choroidopathy[11], optic neuropathy[12,13,14] are other manifestations.

Treatment: Due to its multi system involvement, multi disciplinary approach is needed. The main goal of treatment is to induce remission, maintain it and prevent relapses. Rheumatologist, nephrologist, dermatologist and ophthalmologist are involved in the treatment of this disease. Drugs used to treat SLE include non-steroidal anti-inflammatory drugs, hydroxychloroquine, systemic corticosteroids, immunosuppressive therapy (azathioprine, methotrexate, mycophenolate mofetil and cyclophosphamide) and biologics (rituximab, epratuzumab, sifalimumab, epratuzumab).15 Hydroxychloroquine is is now recommended long term for all patients with SLE. Patients must be advised to undergo regular check up to detect macular toxicity caused by this drug.16 Local treatment depends on the manifestation; it may vary from using lubricants for dry eye, to topical steroids for uveitis, pan retinal photocoagulation, anti VEGF to treat neovascularisation secondary to retinal vascular occlusion.

MATERIAL AND METHODS

In this study we present a series of cases with SLE and ocular manifestations. Patients who presented from July 2016 to April 2017 at to the ophthalmology OPD, Deccan College of Medical Sciences, Hyderabad, and Telangana, India were diagnosed with SLE underwent a comprehensive ophthalmic examination. Patients symptoms were treated accordingly.

Table 1

Sr. No	Case	Chief complaints	BCVA	POSITIVE FINDINGS	Investigations	Treatment	Follow up
1	50/F	FB sensation	BE 6/9	Episcleral congestion	TBUT 8 sec, Schirmers' 10mm	NSAIDs Lubricants	3 months better
2	51/F	DV (RE),Pain, redness (BE)since 1 month	RE- 6/18 LE-6/9	Ciliary congestion, Kp, Cells++, flare + Normal fundus	TBUT 9 sec Schirmer 11mm	Topical steroids Cycloplegics lubricants	3 months better
3	45/F	DV, pain, redness in BE 1 month	RE-6/18 LE-6/12	Ciliary congestion, KP,cells++,flare++posterior synaechiae at 5 clock		Topical Steroids, cycloplegics	1month better

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4	55/F	Dv, irritation, Glare, haloes(BE) recurrent episodes on topical steroids	BE-6/60	Fundus normal PSC, IOP -30mm Hg,Gonio –open angles garde 4 pigmentation 0.8:1 CDR, inferior notch,	TBUT 8 sec Schirmer 12 mm HVF 24-2 superior arcuate scotoma TBUT 7Sec Schirmer 4mm	Antiglaucoma medications Stop topical steroids Start lubricants	Stable at 6 months
5	60/F	Decreased central vision, dryness since 1 month,	BE-6/60	SPK Iin both eyes fundus -ring of depigmentation surrounded by ring of hyperpigmentation	FFA- window defects in a bulls' eye pattern OCT- thinning of retina in parafoveal region	discontinue HCQs and started on alternative therapy Lubricants, cyclosporine	Stable at 6 months
6	50/F	decrease of vision RE, dryness both eyes since 1 month	RE CF 3m, LE-6/9	SPK optic atrophy with neovascularisation in the supero temporal quadrant in the right eye, left eye normal	TBUT 8 Sec Schirmer 10 mm	sectoral prp done right eye	Stable at 1 year follow up
7	55/F	decrease of vision in both eyes since one month	BE-6/9	Fundus- cotton wool spots with normal macula	TBUT 8 sec, Schirmers' 10mm	observation, regular follow up to check for progression of retinopathy and early intervention needed if any.	Stable at 6 months

RESULTS

Table 2

Ocular lesion	Number of patients
Kerato conjunctivitis sicca (xerosis)	6
Episceritis	1
Anterior uveitis	2
Cataract	1
Glaucoma	1
Retinopathy	3
Maculopathy	1

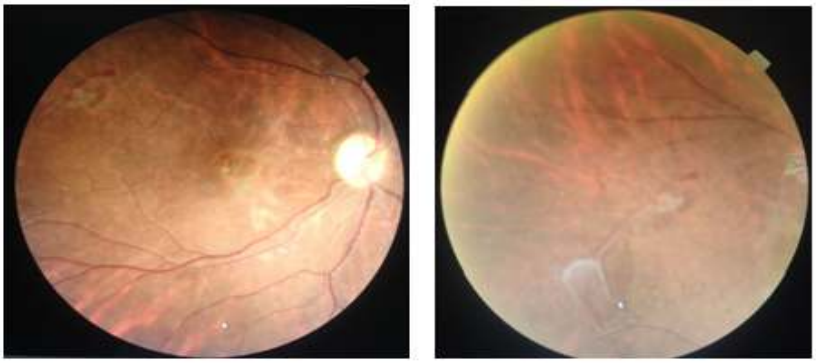


Figure 1: Fundus picture of RE showing active vasculitis and optic atrophy

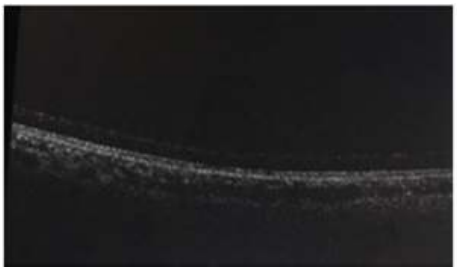


Figure 2: OCT Of RE showing foveal thinning

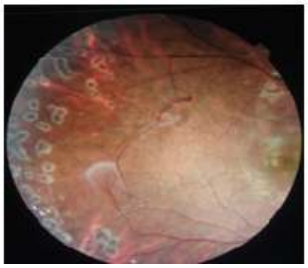
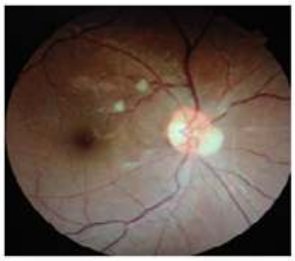


Figure 3: Post sectoral laser Treatment



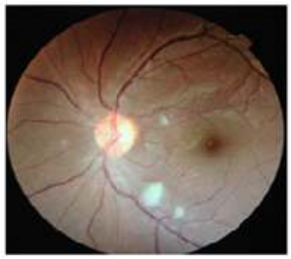


Figure 4: Fundus picture showing both eyes SLE associated retinopathy

Dry eye, episcleritis and retinopathy were managed conservatively; Anterior uveitis was managed with topical steroids. Patients with HCQ associated toxicity were advised to stop the HCQ therapy or to go for an alternative regimen. Patient with Posterior sub capsular opacities were advised cataract surgery, while patient showing glaucomatous changes was started on anti Glaucoma therapy.

CONCLUSION

Early diagnosis, prompt referral, co ordinated treatment and long term follow-up through multi disciplinary approach involving Ophthalmologists, Rheumatologists and Dermatologists play a key role in reducing ocular and systemic morbidity associated with Systemic lupus erythematosus. Since ocular inflammation in SLE can antedate the diagnosis of SLE and cause significant morbidity, early detection, early diagnosis and prompt referral to ophthalmologists can prevent permanent visual loss in some instances.

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