Original Research Paper

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Dermatology

OCULAR MANIFESTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS – A CLINICAL STUDY

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ABSTRACT **Aim:** To determine the various ocular manifestations of SLE for early diagnosis, prompt initiation of therapy and better prognosis of SLE cases. **Materials And Methods:** A total of 50 diagnosed patients of SLE attending RIO and department of Dermatology, GMCH were selected for the study. Detailed history and a thorough ocular examination was performed. Laboratory investigations including R/E blood, ESR, RBS, R/E urine, 24 hours urinary protein, RFT and ANA were done. **Results And Observations:** Incidence of ocular manifestations of SLE was 70%. The mean age of presentation was 32.2 years. The female: male was 11.5:1. The median duration of illness was 1.75 years. The commonest systemic involvement was hematological(90%), followed by cutaneous(64%), musculoskeletal(60%), renal(42%), serosal(4%), neurological(2%). 72% had SLEDAI score ≤10 and 28% had >10. Anterior segment involvement(58%) is more than posterior segment(12%). Common anterior segment manifestations were Keratoconjunctivitis sicca, PSC, eyelid lesions, ulcerative blepheritis, subconjunctival haemorrhage, periorbital oedema, PUK, episcleritis. Posterior segment manifestations were lupus retinopathy, hypertensive retinopathy, CSCR, CRVO, optic atrophy and pigmentary maculopathy. 56% were on steroid therapy, 36% were on steroid+HCQS and 8% were on steroid+cyclophosphamide. **Conclusion:** SLE is a multisystem disease with varied manifestations and diagnosis depends upon careful evaluation of all the systems including the eye. The ophthalmologist may play an important role in the care of the SLE patients, since ocular inflammatory lesions may precede potentially serious extraocular disease and may guide towards the diagnosis.

KEYWORDS : SLE, Autoimmune, Keratoconjunctivitis

INTRODUCTION:

Systemic lupus erythematosus(SLE) is a chronic, autoimmune, connective tissue disorder affecting multiple organ systems often with a relapsing and remitting clinical course. The basic pathology in SLE is due to production of a number of pathogenic autoantibodies and immune complexes and to an inability of the immune system to suppress and clear them. SLE is characterized by the hyperactivity of the immune system and prominent autoantibody production, wherein organs and cells undergo damage, mediated by tissue binding autoantibodies and immune complexes[1]. The disease can manifest in a variety of forms, degrees and manifestation, ranging from mild cutaneous and joint involvement to lethal cardiac, renal and cerebral involvement, as reported by **Goodfield *et al*.,(1998)**[2]. It presents commonly in young and middle aged women who comprise up to **90%** of those affected by the disease. The estimated incidence of SLE ranges from 1.8 to 20 per 1,00,000 populations per year. In India, the incidence is 3 in 1,00,000 populations per year[3]. SLE has a strong female preponderance, approximately 9:1.

Although eye itself is regarded as an "immune privileged organ", SLE can affect every ocular structure. If untreated, it may lead to severe visual loss and even blindness. The ocular manifestations affecting the eye or visual system are seen in 25-33% of cases[4]. The ocular manifestations may precede or follow the onset of systemic illness. Ocular manifestations of SLE can be an indicator of potentially serious systemic disease activity. They are usually reflection of systemic disease. Thus, the presence of ocular lupus should alert the clinician to the likely presence of disease activity elsewhere. The eye has been rightly considered to be the **"barometer for SLE"**[5]. The ocular features of SLE can range from lesions of eyelid, keratoconjunctivitis sicca, episcleritis, scleritis to sight threatening complications like retinal vascular diseases and neuro-ophthalmic complications. Lupus choroidopathy or lupus optic neuropathy may lead to vision loss. Also, there can be features related to complications of SLE like neuropathy, nephropathy, gastrointestinal complications etc. Also, the drugs used in the treatment of SLE can lead to ocular complications viz. steroid can cause cataract, glaucoma and various ocular infections.

AIM:

To determine the various ocular manifestations of SLE for early diagnosis and prompt initiation of therapy and better prognosis of SLE cases.

MATERIALS AND METHODS:

This study was conducted in a Tertiary Care Centre for a period of one year between June 2020 and May 2021. A total of 50 diagnosed cases of SLE were selected for this study. Informed and written consents were obtained from each of the patients.

Institutional ethical committee clearance was obtained before starting the study.

Inclusion Criteria:

- Age: diagnosed cases of SLE between 20-60 years.
- Sex: both male and female patients.
- Both indoor and OPD patients attending RIO, GMCH and Department of Dermatology.

Exclusion Criteria:

- The patients with other autoimmune diseases like scleroderma, rheumatoid arthritis, Sj□gren's syndrome.
- The patients with other retinal vascular diseases like diabetic retinopathy and hypertensive retinopathy without SLE.

A detailed history including age, sex, initial manifestations, duration of illness, present complaints and any ocular complaints in particular, details regarding laboratory investigations done, treatment taken and similar history of illness in the family were taken and a thorough systemic examination was done. Ocular examination was done very meticulously in every patient assessing Visual acuity, IOP, ocular motility, pupillary reflexes, Schirmer's test, diffuse light and slit lamp examination of the anterior segment including tear film breakup time

and tear film meniscus, fundus evaluation by direct, indirect, slit lamp biomicroscopy using +90D lens after full mydriasis and OCT imaging were performed. Laboratory investigations including R/E blood, ESR, RBS, R/E urine, 24 hours urinary protein, RFT and ANA were done.

The diagnosis of SLE was made based on revised 11 diagnostic criteria proposed by American Rheumatism Association, as described by Tan *et al*, 1982[6].

The statistical data has been calculated using IBM SPSS Latest version of 2022.

RESULTS AND OBSERVATION:

Table 1: Incidence Of The Ocular Manifestations In The Total Number Of SLE Patients Examined During The Study Period:

CATEGORY	NO. OF PATIENTS
Total no. of patients examined during the study period	50
Total no. of patients with ocular manifestations	35

Thus, the incidence of ocular manifestations of SLE in the present study was approximately 70% of the total number of SLE cases examined during the study period.

Table 2: Age Distribution:

AGE GROUP (IN YEARS)	NO. OF PATIENTS	PERCENTAGE
20-30	28	56%
31-40	14	28%
41-50	6	12%
51-60	2	4%

The mean age of presentation is 32.2 years and median age of presentation is 30 years. Majority of the patients were in the age group of 20-30 years.

Table 3: Sex Distribution:

SLE PATIENTS	NUMBER	PERCENTAGE
Female	46	92%
Male	04	8%

The female: male ratio is 11.5:1.

Table 4: Distribution Of Patients With Different Duration Of Illness:

Duration Of Illness	No. Of Patients	Percentage
< 1 year	18	36%
1-5 years	28	56%
6-10 years	2	4%
>10 years	2	4%

Most of the patients (56%) were in the range of 1-5 years. Within the subgroup, 12 patients had the disease duration of 1year, 3 patients of 1.5 years, 6 patients of 2 years, 4 patients of 3 years, 2 patients of 4 years and 1 patient had disease duration of 5 years. The mean duration of illness was 1.89 years.

Table 5: Number Of Systems Involved In Various Patients In The Present Study:

No. Of Systems Involved	No. Of Patients	Percentage
1	2	4%
2	18	36%
3	27	54%
4	2	4%
5	1	2%

Table 6: Number Of Patients With Different Systemic Involvement:

System	No. Of Patients	Percentage
Cutaneous	32	64%
Musculoskeletal	30	60%
Haematological	45	90%
Renal	21	42%
Neurological	1	2%
Serosal	2	4%

Maximum number of patient had haematological involvement. 45 patients, out of 50 (90%) had either anaemia or leucopenia (<4000) or thrombocytopenia (<100000).

The next commonest involvement was cutaneous, followed by musculoskeletal. 32 patients (64%) had cutaneous involvement in the form of malar rash, discoid rash, photosensitivity or oral ulcers. Musculoskeletal involvement was seen in 60% of cases.

Renal involvement, i.e. lupus nephritis, in its various stages, was seen in 21 patients out of 50 patients (42%). One patient had neurological involvement in the form of seizure disorder. 2 patients had serosal involvement in the form of pleural effusion.

Assessment Of Disease Activity:

The disease activity of all the SLE cases was assessed at presentation in relation to the Systemic Lupus Erythematosus Disease Activity Index (SLEDAI). Each of the patients was assigned a score depending on the number of organs involved. Each organ involvement carried a fixed score. Score ≤10=mild to moderate disease. Score≤10=greater disease activity. In the present study, SLEDAI score ranged from 3 to 28. Majority of the patients had score ≤ 10 (72%). Rest of the patients had score>10 (ranging from 11 to 28). The mean score was 9.62.

Table 7: SLEDAI Score Distribution:

SLEDAI SCORE	NO. OF PATIENTS	PERCENTAGE
≤ 10	36	72%
>10	14	28%

Table 8: BCVA:

Best Corrected Visual Acuity	No. Of Patients	Percentage
6/6 - 6/12	44	88%
6/18 - 6/60	4	8%
< 6/60	2	4%

Out of the 44 patients in the first group (6/6-6/12), 6 patients had BCVA 6/12 in one or both the eyes.

Table 9: Segmental Involvement Of Eye:

SEGMENT INVOLVED	NO. OF PATIENTS	PERCENTAGE
Only Anterior segment	22	58%
Only Posterior segment	6	12%
Both segments	7	14%

Table 10: Various Anterior Segment Manifestations In The Study Group:

Anterior Segment Manifestation	No. Of Patients	Percentage	No. Of Eyes
Keratoconjunctivitis sicca	18	36%	34
Posterior subcapsular cataract	10	20%	20
Eyelid lesions	4	8%	8
Ulcerative blepharitis	3	6%	6
Subconjunctival haemorrhage	2	4%	2
Periorbital oedema	1	2%	2
Peripheral ulcerative keratitis	2	4%	2
Episcleritis	1	2%	1

Table 11: Various Posterior Segment Involvements:

Posterior Segment Manifestations	No. Of Patients (% Of Patients)	No. Of Eyes
Lupus retinopathy	5(10%)	10
Hypertensive retinopathy	3(6%)	6
CSCR	2(4%)	2
Optic atrophy	1(2%)	2
CRVO	1(2%)	2
Pigmentary maculopathy	1(2%)	2

Table 12: Different Treatment Received By The Patients:

Treatment	No. Of Patients	Percentage
Steroid	28	56%
Steroid+HCQS	18	36%
Steroid+Cyclophosphamide	4	8%

Therapy And Its Complications:

Out of 50 patients, 11 patients had ocular manifestations arising from the drugs used in the treatment of SLE. Out of 50 patients on steroid, 10 patients developed **bilateral posterior subcapsular cataract**. Out of 18 patients on steroid + HCQS, 1 had **bilateral pigmentary**

maculopathy. However, none of the complications were related with the duration of therapy.

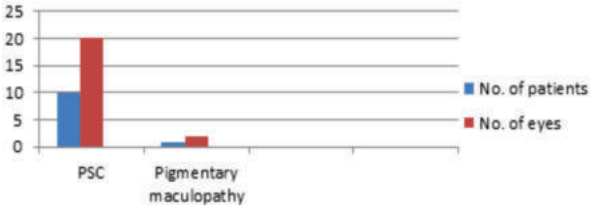

Figure 13: Complications Of Therapy In The Study Group:

DISCUSSION:

In the present study involving 50 diagnosed cases, 35 patients had ocular involvement which amounts to 70%. **Ushiyama *et al*.,(2000)**[4] in their study reported that ocular complications are seen in around 25-33% cases of SLE.

In the present study, the mean age of presentation was 32.2 years. **Yap *et al*.,(1998)**[7] reported the mean age of the study population as 32.9 years in their study. Thus, this present study was comparable with the earlier studies.

In the present study, out of 50 cases, 46 patients were female and 4 were male. Thus, the F:M ratio was 11.5:1. **Kumar *et al*.,(2002)**[3] in their study observed the F:M ratio to be 11:1. Thus, in the present study the sex ratio was comparable to the earlier studies.

In the present study, the mean duration of illness was 1.89 years. **M P K Soo et al.,(2000)**[8] in their study found that the mean duration of illness to be 3.5 years. Earlier study involved mainly the patients who were asymptomatic. However, the present study involved most of the admitted cases with more severe involvement. This may be the only reason for shorter duration of illness in the present study as compared to the earlier study.

In the present study, systemic involvement was seen in all the cases, ranging from 1-5 systems. Majority of the patients (60%) had three or more systems involvement while 4% and 36% patients had one and two systems involvements respectively. **Soo *et al*.,(2000)**[8] reported in their study that one or two system involvement in 4% and 35% patients respectively while 60% had three or more system involvement. Thus, the present study was comparable with this study. In the present study, the disease activity was assessed using the **SLE disease activity index (SLEDAI)**. Out of 50 patients, 14 had SLEDAI score >10 which indicated increased disease activity. Rest of the patients had score ≤10, which indicated mild to moderate disease. The mean SLEDAI score was 9.62. **Narayanan et al.,(2010)**[9] in their study of 50 patients, found the mean score of 10.88. Thus, the present study is comparable to the earlier study.

In the present study, out of 50 patients, 44 patients had best corrected visual acuity (BCVA) 6/12 or better in either of the eyes. 4 patients had BCVA between 6/18 and 6/60 and 2 patients had vision worse than 6/60. Among the 44 patients, 6 patients had BCVA of 6/12. All these patients had bilateral posterior subcapsular cataract. **Yap *et al*.,(1998)**[7] in their study found BCVA less than 6/12 in 7 patients out of 70 patients (10%). Out of 7 patients, two each had cataract, optic atrophy and retinal vaso-occlusive disease. Thus, the present study is comparable with the earlier study. Out of 35 patients with ocular manifestations, 22 patients had anterior segment involvement and 6 had posterior segment involvements and 7 patients showed both systems involvements.

In the present study, the commonest anterior segment manifestation was **keratoconjunctivitis sicca,** present in 36% of patients (18 in no.) as recorded by Schirmer's strip with a recording of less than 10 mm. **Jensen *et al*.,(1999)**[10] in their study reported the incidence of KCS in SLE to be up to 35%. The next common anterior segment manifestation was **posterior subcapsular cataract (PSC)**, seen in 10 patients (20%). This was considered to be a complication of the use of corticosteroids in the treatment of SLE. **Yap *et al*.,(1998)**[7] in their study reported the incidence of steroid induced PSC in SLE to be 20%. In the present study, **scaly eyelid lesions and ulcerative blepharitis** was found in 8% and 6% of the cases respectively. **Huey *et al*.,(1983)**[11] in their study found eyelid involvement in SLE patients in 6% of the cases, which is comparable to the present study. In the present study, each **subconjunctival haemorrhage and peripheral ulcerative keratitis (PUK)** are seen in two of the patients. **Foster *et al*.,(2000)**[12] in their study reported peripheral ulcerative keratitis as a rare manifestation and an ominous sign of active vasculitis with higher disease activity. In the present study, out of 50 patients, one patient (2%) presented with **episcleritis. Frith *et al*.,(1990)**[13] in their study reported the occurrence of repeated episodes of episcleritis in patients with SLE and the incidence may rise up to 28%.

In the present study, 13 patients (26%) presented with posterior segment involvement. The commonest finding being **lupus retinopathy (benign form)**, seen in 10% of the patients. All patients had bilateral involvement. **Ushiyama *et al*.,(2000)**[4] in their study of 69 patients reported the incidence of lupus retinopathy to be 10%. The classical findings like cotton wool spots, hard exudates and sub-conjunctival haemorrhages were present in all the 6 patients bilaterally. In this group of patients, vision loss was ranged from 6/6 to 6/36. In the present study, 1 patient presented with **bilateral central retinal vein occlusion**. The vision of the patient was finger counting at two feet in both the eyes. There was also involvement of **central nervous system**. Thus, the present study also supported the association between severe vaso-occlusive retinopathy in SLE and CNS lupus. In the present study, out of 50 cases, 2 patients (4%) presented with **central serous chorioretinopathy**. **Sivaraj *et al*.,(2007)**[14] in their study reported that lupus retinopathy and scleritis are the hallmarks of systemic disease activity and require systemic immunosuppression. In the present study, 1 patient (2%) presented with **bilateral optic atrophy** with gross diminution of vision. In the study group, features of **hypertensive retinopathy** were seen in 3 (6%) patients out of 16 hypertensive patients. Also, in the present study, one patient presented with **pigmentary maculopathy** in both the eyes, which was considered to be a complication of hydroxychloroquine (HCQS) therapy. In the present study, follow up was difficult as most of the patients didn't turn up for check-up. In majority of cases, either they were asymptomatic or they had other comorbidities for which follow up could not be done. For the above mentioned reason, long term monitoring of the ocular involvement in relation to systemic status was not possible.

CONCLUSION:

According to the results obtained from the present study, it can be concluded that systemic lupus erythematosus and its ocular manifestations are not uncommon in this part of the country. Also, some of the ocular manifestations are associated with significant morbidity. In our present study, ocular involvement was found to be 70% of the total SLE cases. Both anterior and posterior segments were involved, anterior being more common than posterior. Thus, we conclude that SLE is a multisystem disease with varied manifestations and diagnosis depends upon careful evaluation of all the systems including the eye. Therefore, ophthalmologist should include SLE in the differential diagnosis of many retinal vascular and neuro-ophthalmic disorders. In addition, all the patients with ocular lupus should be carefully evaluated for systemic involvement to detect potentially treatable and preventable complications of the disease. The ophthalmologist may play an important role in the care of the SLE patients, since ocular inflammatory lesions may precede potentially serious extraocular disease and may guide towards the diagnosis.

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