An unusual case of orbital involvement and pan-ocular inflammation in Acute Systemic Lupus Erythematosus.

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Abstract:

Systemic lupus erythematosus (SLE) is an autoimmune disease in which organs and cells undergo damage mediated by tissue-binding autoantibodies and immune complexes. At the onset, SLE may involve only one organ (additional manifestations may occur later) or multiple organs. The ocular manifestations are varied and involve the lids, cornea, sclera and or orbit. There may be associated secondary Sjogren's syndrome, conjunctival haemorrhages, retinal vascular disease with retinal haemorrhages and neuro-ophthalmic lesions. Pan-ophthalmic involvement is rare and sight threatening and requires urgent ophthalmic intervention. We hereby present an unusual case of pan-ophthalmic inflammation in case of acute systemic lupus.

Keywords: Ophthalmic manifestations of SLE, Orbital myositis, Panuveitis, Scleral perforation, Systemic lupus erythematosus.

Introduction:

Systemic lupus erythematosus (SLE) is a chronic multisystem immunologically mediated disease of unknown etiology. Production of a number of pathogenic autoantibodies and immune complexes and an inability to suppress or clear them, are the underlying abnormalities in SLE. SLE is rare in India with a prevalence of 3 in 1,00,000. The median age of onset is 24.5 years and the sex ratio (Female : Male) is 11:1. Majority of the patients (90%) are females.[1]

Clinical features of SLE include constitutional symptoms of fever, malaise, arthralgias, myalgias, headache, and loss of appetite and weight. Systemic involvement includes skin manifestations in the form of typical malar butterfly rash, discoid lesions, a nonspecific erythematous maculopapular rash and cutaneous vasculitic lesions; renal glomerular deposits; neurologic manifestations that may involve any part of the peripheral or central nervous system; vascular and hematologic abnormalities including Raynaud's phenomenon, anaemia, and the presence of antiphospholipid antibodies; cardiopulmonary involvement including pleuritis, pleural effusion, pericarditis, myocarditis, vegetative endocarditis, and coronary artery disease; gastrointestinal disease having non-specific signs and symptoms; varied ocular involvement. Often, at the time of presentation, only one organ system may be involved.[2]

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The clinical course of SLE is highly

variable in terms of rate of progression and severity. Most of the patients have periods of remission interrupted by exacerbations rather than a relentless and progressive disease. There is no known cure for SLE, and complete remission is rare. Twenty to thirty percent of patients have mild disease that can be controlled with nonsteroidal anti-inflammatory drugs alone. Life-threatening and severely disabling disease requires high-dose systemic corticosteroids that are gradually tapered.[2] Plasmapheresis, intravenous immunoglobulin and other antimetabolites have been found helpful.

Ocular manifestations affecting the eye or the visual system are seen in 25 - 33% of patients with SLE and may precede or follow the systemic illness. SLE may affect almost any part of the eye and visual pathway including the periorbita, ocular adnexa, eye, and optic nerve. The most common association is keratoconjunctivitis sicca.[3]

SLE may cause ocular disease by mechanisms including immune complex deposition and antibody related mechanisms, vasculitis and thrombosis. Immune complex depositions have 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.[4]

External ophthalmic manifestations include keratoconjunctivitis sicca, conjunctivitis, superficial punctate and interstitial keratitis, episcleritis and scleritis, discoid lupus of the eyelids. Sight threatening manifestations include retinal haemorrhages, branch retinal vein occlusion, central retinal vein occlusion, central retinal artery occlusion, retinal vasculitis, proliferative retinopathy, ischemic choroidopathy, retinal pigment epithelial changes and focal retina detachments, choroidal vasculitis, uveitis, optic neuritis, ischemic optic neuropathy. Orbital manifestations include orbital pseudotumor and orbital myositis[2]. Ocular manifestations have been described in drug-induced lupus caused by hydralazine and procanamide that include retinal vasculitis and occlusive disease.[4]

We hereby present a case of orbital involvement and panocular inflammation in a patient of acute SLE. Panocular inflammation is a very rare manifestation of SLE. Very few cases have been reported till now. No such case has been reported in India.

Case Report:

A 38 years old male presented with chief complain of decrease in vision in right eye (RE) for 15 days. This was sudden, progressive and painful in nature. It was associated with diffuse orbital swelling and inability to move right eye inward and upward. There was no history of trauma, foreign body or any ocular surgery in the RE. He had a history of jaundice and generalised body weakness for last 2 months and was diagnosed as an acute systemic lupus erythematosus. Intravenous methyl predisolone 1 gram stat for 5 days followed by tablet methotrexate 10 milligram once a week was given for the treatment of SLE. There was a history of whole blood transfusion twice in last 10 days for treatment of microcytic hypochromic anaemia which is one of the clinical manifestations of SLE.

On clinical examination, his visual acuity in RE was appreciation of hand movements and light perception but defective perception of rays. On gross examination of RE he had diffuse orbital swelling.

On detailed slit lamp evaluation, RE showed ciliary congestion along with chemosis, clear cornea, nasal sclera having scleral abscess of 6 x 9 mm along with perforation of 2 x 2 mm within the scleral abscess. Anterior chamber was normal in depth with hyphema and reaction +4, pupil was semidilated with posterior synechiae and the lens details could not be appreciated due to hyphema and severe anterior chamber reaction. There was restriction of superior and medial ocular movements. The fundal glow was altered. Digitally, the intra ocular pressure was very low.

Image 1 : RE showing lid oedema, ciliary congestion, chemosis and nasal scleral abscess

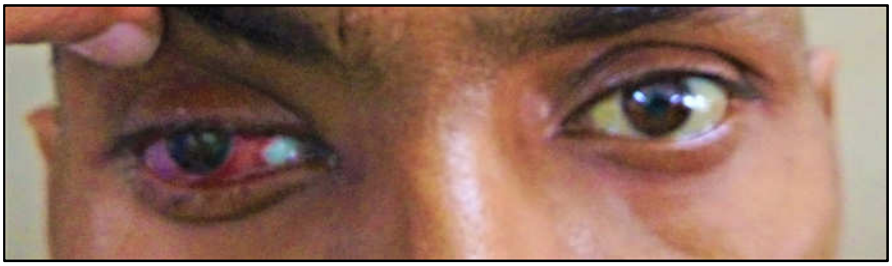


Image 2: RE showing Hyphema & Nasal scleral abscess

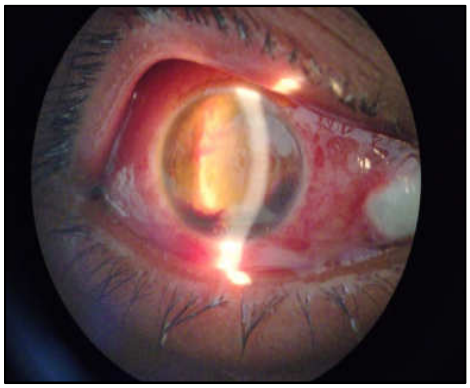


Image 3: Fundus of LE showing Roth spots, Intraretinal haemorrhage & Macular oedema.

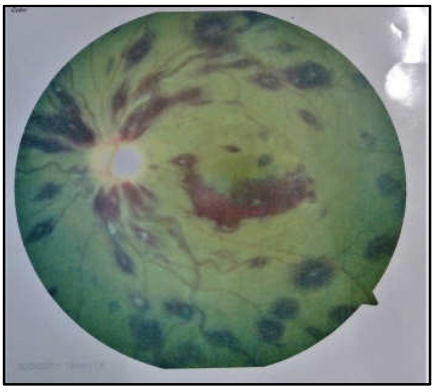


Image 4: B-scan ultrasonography of RE showing Exudative retinal detachment, Choroidal thickening, Suprachoroidal effusion

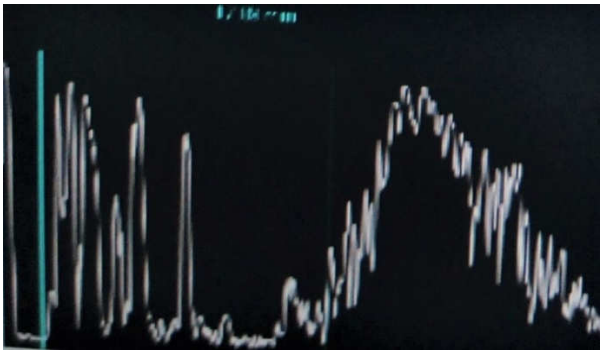
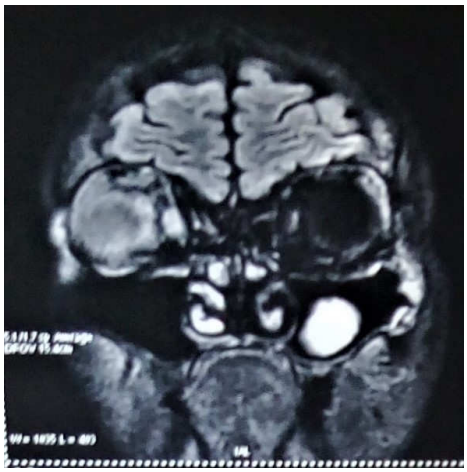


Image 5: MRI brain with orbit



[RE = Right Eye, LE = Left Eye]

In left eye, the best corrected visual acuity (BCVA) was 6/24 on Snellen's chart and

anterior segment was within normal limits.

No fundus details were visible in RE because of severe anterior chamber reaction and vitritis. In left eye, multiple roth spots were seen on the retina with multiple intraretinal haemorrhages and macular oedema.

B-Scan 10 MHz frequency ultrasonography of RE showed exudative retinal detachment, choroidal thickening and supracoroidal effusion.

MRI brain with orbit showed hyperintensity with inflammatory changes involving right eyeball with involvement of retro-orbital fat, superior and medial recti muscles and orbital proptosis. No definite evidence of involvement of optic nerve, orbital apex or cavernous sinus region was seen. Left orbit, eyeball and optic nerve appeared normal.

Following other investigations were carried out :

- Hemogram and smear study showed haemoglobin 7.3 gm/dL, total RBC count of 2.31 million/ml with mild microcytic hypochromic RBCs with moderate polychromasia, total white blood cells count of 9900 cells/ml with relative neutrophilia and platelet count of 1,43,000.
- ESR was 150 mm/hr
- HIV-Non-reactive
- HBsAg- Non-reactive
- Bilirubin (direct and indirect) levels were high.
    - Total bilirubin 2.46 mg/dL.
    - Direct 1.05 mg/dL
    - Indirect 1.41 mg/dL
- SGPT levels were raised - 70.19 units/litre.
- Anti Ds-DNA (anti double stranded DNA) and ANA (anti nuclear antibody) were positive.

Treatment was started in RE in form of topical fortified cefazoline 5% eyedrops every 1 hourly, fortified amikacin 2% eyedrops every 1 hourly, carboxy methyl cellulose(0.5%) eyedrops 6 times and atropine 1% eyedrops 3 times a day. In LE nepafenac eyedrops three times a day was given for macular oedema.

Systemic intravenous cefoperazone with sulbactum 1.5 gm 12 hourly, intravenous amikacin 500 mg 12 hourly, intravenous metronidazole 400 mg 8 hourly were given. Tablet vitamin B-complex, vitamin C and Folic acid were given twice a day. Tablet serratiopeptidase 15 mg with nimesulide 100 mg was given three times a day.

As per rheumatologist’s opinion, tablet methotrexate 10 mg once a week was continued.

Patient was followed up on a daily basis. There was gradual improvement in symptoms. Proptosis, lid edema, conjunctival chemosis, anterior chamber reaction decreased. The scleral abscess did not respond to treatment. In left eye, the Roth spots and intraretinal haemorrhages decreased in size, macular oedema showed no changes.

After one week, fortified antibiotics were stopped and substituted with topical moxifloxacin eyedrops six times a day and tobramycin eyedrops six times a day. Carboxy methyl cellulose eyedrops six times a day and atropine eyedrops three times a day were continued. Intravenous antibiotics were stopped and oral Tablet amoxicillin 500 mg+clavulanic acid 125 mg three times a day was started.

After 2nd week, in RE, the inflammation reduced but the scleral abscess was nonresponding. RE showed further hypotony, increased suprachoroidal effusion with decreased axial length on USG B-scan suggestive of pthysical changes. In Left eye, the haemorrhages regressed, macular oedema decreased and BCVA improved to 6/12.

Discussion:

Systemic lupus erythematosus (SLE) is a chronic multisystem autoimmune disease. The eye is frequently involved in SLE. SLE may affect any structure of the eye and adnexa, unlike other autoimmune diseases, which may have a predilection for either anterior or posterior segment of the eye.[5]

Orbital involvement is a rare manifestation of SLE. Orbital disease in patients with SLE may include primary inflammation of the orbital tissue (e.g., orbital pseudotumor or orbital inflammatory syndrome) or inflammation of the extraocular muscles (e.g., orbital myositis). The most common sign of orbital disease is proptosis that may be caused by a space-occupying lesion or inflammatory process. Associated symptoms and signs include pain, blurred vision, diplopia as a result of limitation of eye movement, eyelid swelling and displacement of the globe.[6]

Acute orbital inflammation is a rare manifestation of SLE. Very few cases of orbital inflammatory syndrome in SLE have been reported. Orbital pseudotumor is a nongranulomatous inflammatory disorder that may affect any structure in the orbit. It is defined according to the orbital structures affected, with the presenting symptoms reflecting the degree of inflammation and the location of the inflamed tissue.[6]

Orbital myositis secondary to SLE may be misdiagnosed as bacterial orbital cellulitis. Orbital vasculitis may lead to nonperfusion of the globe and extraocular muscles that cause restriction of extraocular movements. CT scan, MRI or ultrasound of the orbit may reveal enlargement of the extraocular muscles in these cases. Treatment of orbital disease is with systemic immunosuppressant drugs[3]. Several reports of SLE presenting with proptosis due to orbital pseudotumor have been published.[6,7,8]

Scleritis is more painful and potentially vision-threatening. Scleritis in patients with SLE may present as anterior diffuse scleritis or anterior nodular scleritis. Necrotizing scleritis in patients with SLE is rare but may lead to significant scleral thinning and subsequent perforation and vision loss. Scleritis requires systemic therapy including non steroidal anti-inflammatory drugs like flurbiprofen or in severe cases corticosteroids and other immunosuppressive agents.[4]

Uveitis is rare but may occur in patients with SLE. Adjacent scleral inflammation may also lead to mild uveitis. The inflammation of the anterior segment usually improves with the use of systemic immunosuppressive therapy.

Lupus choroidopathy with exudative retinal detachments is rare with less than 40 patients reported in the literature.[3]

Orbital involvement in SLE patients represents a diagnostic dilemma, as well as a therapeutic challenge. In our case although the proptosis, lid and conjunctival oedema improved with immunosuppression ultimately irreversible visual loss occurred and the eye eventually developed pthysis bulbi. Early ophthalmic consultation and intervention at the onset of symptoms could have led to better visual prognosis in this case. Thus close cooperation between ophthalmologist and rheumatologist is essential in the management of these patients, in order to prevent visual handicap and blindness.

Conclusion:

The eye manifestations in SLE are variable. Significant ocular pain or reduction of vision is serious symptom requiring urgent assessment by an ophthalmologist. Careful assessment by the ophthalmologist is mandatory to prevent sight-threatening complications. Close communication between the consultant ophthalmologist and treating rheumatologist is critical in the effective management of these complex clinical situations.

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