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# ORBITAL PSEUDOTUMOUR IN SLE- A RARE CASE REPORT

Ophthalmology

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## ABSTRACT

A 40 year old female came to OPD with proptosis of R/E since 5 days, with marked conjunctival chemosis, hyperemia, restricted Extraocular muscle movement(EOM), gradual diminution of vision and watering since 5 days.Swelling gradually regressed, involved the other eye within 5 days,with restricted EOM. V/A was PL in R/E and HMCF in L/E. CT scan orbits: diffuse thickening of right orbit involving sclera, suggesting orbital pseudotumour with scleritis. ANA IFA+(1:80) dilution, ANA BLOT-antids DNA-+/- ,antiribP0-+/-, antiss A1R060-+, ANA hep 2 + TREATMENT: Case responded well to oral prednisolone : 1 mg/kgbw tapered over 90 days,oral azathioprine 50 mg for 60 days, along with moxifloxacin eye drops and ointment.

## KEYWORDS

pseudotumour,scleritis,prednisolone,azathioprine

## I. INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic, autoimmune, connective tissue disorder affecting multiple organ systems often with a relapsing and remitting clinical course1. Ocular manifestations-occurring in up to one third of patients, can be associated with significant morbidity and also a marker for overall systemic disease activity.

SLE can affect the periorbita, ocular adnexa, eye, and optic nerve. The most common association is keratoconjunctivitis sicca, while the most visually devastating sequelae occur secondary to optic nerve involvement and retinal vasoocclusion.

Orbital pseudotumor is a non-specific, non-neoplastic disease of the orbit without any obvious cause. It is the second most common orbital disease. In the acute form of the disease, patients present with unilateral eye pain, lid swelling, proptosis, and occasionally vision loss2. In the chronic form, patients present with diplopia.

Orbital pseudotumour is a nonspecific benign ,noninfective, inflammatory process of the orbit characterised by a polymorphous lymphoid infiltrate with varying degrees of fibrosis.Some common ocular manifestations of SLE include keratoconjunctivitis sicca, , orbital inflammation, retinal vasculitis, retinal vaso-occlusive disease, iritis, scleritis, optic neuropathy.Neuro-ophthalmic manifestations in SLE has a prevalence of 3.6% in adults. Out of which involvement of orbital tissues is rarest.

Orbital involvement is rare in SLE. Most orbital problems develop bilaterally. Orbital inflammation including myositis and panniculitis has been reported3,4. Orbital inflammatory pseudotumor is a rare complication of systemic lupus erythematosus.

## CASE REPORT

A 40 year old woman came to OPD with the complaints of painless swelling of the right upper and lower eyelids , associated with watering and redness since 17 days, insidious in onset and gradually progressive in nature. It was associated with visible whiteness of the cornea since 24 days. There was a past history of hydroxychloroquine intake (200 mg) for 1 month prior to admission. There was no history of trauma, headache, recurrent episodes, vomiting, fever, weight loss ,previous history of malignancy that rules out cavernous sinus thrombosis, thyroid ophthalmopathy, orbital cellulitis, neoplastic mass. On examination, there was diffuse swelling of the right upper and lower eyelids ,that was tender ,associated with conjunctival congestion and chemosis ,excessive lacrimation and mucopurulent discharge associated with slight protrusion of the eyeball(could not be measured).The right cornea had a central leucomatous opacity ,round in shape, regular margins, 6mm in diameter. There was restricted mobility in all gazes, with diplopia. The left eye had a similar leucomatous corneal opacity since 24 days, round in shape, regular margins covering almost the entire cornea.

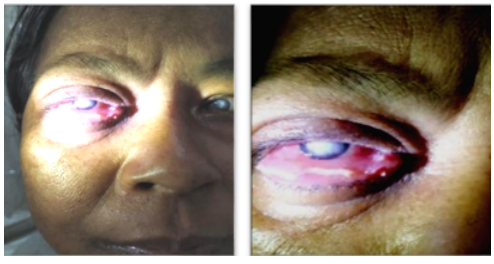

**Fig 1: R/E findings of the patient**

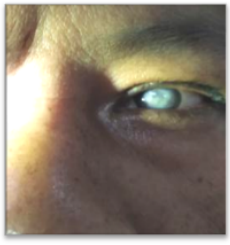

**Fig 2: L/E findings of the patient**

**Fig 3: Extraocular muscle movements**

Based on the above findings, it was suspected to be orbital cellulitis and intravenous antibiotics(ceftriaxone) were started. Due to no improvement in two days, oral steroids were added (prednisolone acetate 1 mg/kgbw).

### Investigations

Investigations were carried out to rule out other inflammatory etiologies. RE blood –normal,Thyroid function tests-normal, .Renal function tests-normal, LFT-normal, CRP-0.68,ESR-60 mm AEFH.CT orbit shows - diffuse thickening of right orbit with scleral involvement

,suggesting orbital pseudotumour with scleritis.

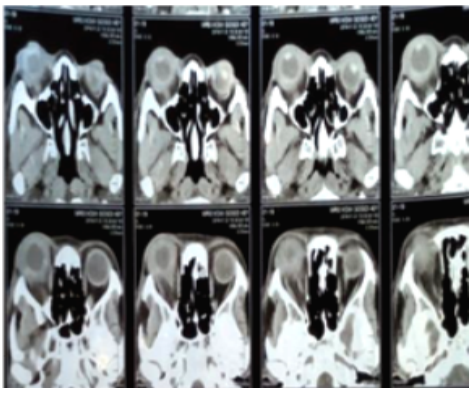

**Fig 4: CT scan of the orbit**

**Treatment**

On administration of steroids , symptoms gradually improved, proptosis resolved, swelling and congestion decreased, but involved the left eye within 5 days. Rapid response to steroids confirmed the diagnosis of pseudotumour .The patient had diffuse swelling of the left upper lid with complete ptosis, restricted mobility,and central leucomatous corneal opacity.The patient was referred to rheumatology OPD for opinion regarding further management. Laboratory analysis showed ANA Hep 2 positive, ANA IFA positive (1:80 dilution),ANA BLOT : ds DNA+/-,Rib P0+/-,ssAIR060-+ that confirmed the diagnosis of SLE. 50 mg once daily dose of azathioprine was added along with oral steroids that helped in resolution of the symptoms of left eye.

**Fig 5: Involvement of L/E within 5 days**

**Follow up**

No recurrence at 1 month and 6 month. Improved BCVA at 6 months was R/E-6/36, –, L/E-6/36,N-.

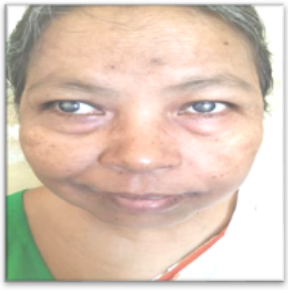

**Fig 6: Follow up at 6 months**

## II. DISCUSSION

Idiopathic orbital inflammatory syndrome(IOIS) as described ,first in 1905 by Birsch Hirshfield,is inflammation of orbital structures not associated with any definite etiology.

IOIS can be further classified on the basis on location of inflammation into myositis(rectus muscles),dacryoadenitis(lacrimal gland), perisclerits (sclera), papillitis (optic nerve), and diffuse (orbital fat) subtypes[5].

IOIS can be associated with many systemic rheumatic diseases like rheumatoid arthritis,Wegeners granulomatosis,and SLE. Patients present with the complaints of pain,proptosis,diplopia(due to infiltration of extraocular muscles)eyelid swelling,impaired visual acuity.The differential diagnosis includes orbital cellulitis,cavernous sinus thrombosis,Graves disease,lymphoma,Wegeners granulomatosis. Initial laboratory assessments are done like thyroid function tests,renal function tests,hepatic function tests,white blood cell count,acute phase reactants,serum complement levels. Autoantibody testing to screen the presence of any underlying rheumatic disease should be performed like ANA, ANCA,rheumatoid factor.

The pathogenesis includes deposition of immune complexes.in the basement membrane of small blood vessel endothelial cells[6].It is associated with diffuse inflammatory infiltration of affected orbital structures. The source of autoantigens that trigger the formation of the aforementioned antibodies is thought to arise from apoptotic cells.

The diagnosis is confirmed by CT scan that demonstrates irregular diffuse or focal thickenings of enlarged orbital structures with heterogenous enhancement.

In our case ,there was associated posterior scleritis. Presenting symptoms include pain,blurred vision,limited eye movements,and proptosis. Treatment options for SLE range from NSAIDS, corticosteroids, antimalarials, immunomodulatory agents. Significant ocular involvement –orbital inflammation, scleritis, retinal vasculitis, choroiditis, and optic neuritis warrants systemic therapy. Corticosteroids are the mainstay of acute treatment for treating ocular SLE1. For mild disease, NSAIDS, antimalarials are chosen for treatment. A treatment protocol has been described starting with high dose oral corticosteroids, followed by pulse parenteral doses if the condition is still unresponsive. The next step is radiotherapy(10-30 Gy),followed by cytotoxic agents like cyclophoshamide, methotrexate, cyclosporineA, azathioprine,mycophenolate mofetil.

Only a few cases of Idiopathic Orbital Inflammatory Syndrome have been reported in patients of SLE. Our case is peculiar in that our case is a female in contrast that orbital pseudotumour is more common in males. Out of all ocular manifestations of SLE,keratoconjunctivitis sicca is commonest,while orbital involvement is rarest.Out of which optic neuritis is commonest(3.6%)[8],in contrast to which our case presents with scleritis and myositis. This case is worth reporting because the involvement of orbital structures in SLE and, specifically inflammatory pseudotumor is very rare and therefore may go unnoticed .Ocular manifestations can be associated with significant morbidity and also a marker for overall systemic disease activity..A considered assessment of both systemic and ophthalmic findings is necessary in determining proper therapy and duration of the treatment.Close communication between consultant ophthalmologist and treating rheumatologist is critical for effective management of such a case.

## II. CONCLUSION

Proptosis in SLE patients represents a diagnostic dilemma, as well as therapeutic challenge.Orbital findings in SLE are a poor prognostic risk factor in both ophthalmic and systemic morbidity.Therefore such a case must be dealt very meticulously and urgently from the beginning.

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