Rare Ocular Manifestations of Systemic Lupus Erythematosus – Two Case Reports

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Abstract

In systemic lupus erythematosus (SLE), ocular involvement is common (prevalence up to 30%) but extremely varied. The most common manifestation is keratoconjunctivitis sicca (in about 25%). Though posterior segment disease is not uncommon, choroidal involvement and optic neuropathy are rare. Visual loss from neuro-ophthalmic involvement is often due to lupus optic neuropathy. Less than five percent of patients only present with sole ocular involvement at diagnosis. We report two patients who presented with decreased visual acuity and rare posterior eye changes as presenting features of SLE in the absence of other clinical manifestations and later developed lupus nephritis.

Introduction

Ocular involvement in SLE can be part of active lupus or antiphospholipid antibody syndrome or due to drugs used in the treatment of SLE. Anterior segment findings include keratoconjunctivitis, scleritis, keratitis and iridocyclitis. Posterior segment abnormalities range from relatively common, asymptomatic microvascular changes, including cotton-wool spots and intraretinal haemorrhages, to rare vision-threatening retinal arterial or venous occlusions.¹ Optic neuritis and papilloedema have also been described. However literature on SLE patients with central serous retinal detachment has been scant.²

Patient 1

A 25 yr old lady had sudden diminution of vision, preceded by a brief febrile illness. Ophthalmic evaluation done elsewhere revealed bilateral disc oedema and exudative retinal detachment (Figure 1a). Fluorescein angiogram showed multiple areas of hyper fluorescence appearing in early phase, increase in intensity and pooling in late phase suggestive of multiple retinal detachment. After 2 months, she developed proteinuria, anaemia, hypertension and antinuclear antibodies (ANA) was positive 2+ [1 in 320]. Renal biopsy showed diffuse mild mesangial proliferation with focal tubular atrophy (Figure 1b) and full house immunofluorescence consistent with Class II Lupus nephritis [ISN – RPS].

Patient 2

A 21 yr old girl had chronic headache, slowly progressive painless loss of vision and easy fatigability for 12 months. There was no history of oedema legs, haematuria or frothy urine. Investigations done after 1.5 years for evaluation of her ocular symptoms revealed nephrotic range proteinuria, serum creatinine -3.5 mg/dl, haemoglobin-11gm/dl, total count - 5400/cu.mm, ESR - 20 mm in 1hr, Direct Coomb’s test –negative, peripheral smear-hypochromic, microcytic RBC’s. ANA was strongly positive (4+,1 in 640 dilution),anti ds DNA - positive, both C3 (< 30 mg/dl) and C4 (< 6 mg/dl) levels were decreased and tested negative for antiphospholipid antibodies. Her fundus showed primary optic atrophy in left eye (Figure 2a) and temporal pallor of optic nerve head in right eye. Her renal histology was chronic sclerosing lupus nephritis (Figure 2b). She attained renal remission after six doses of intravenous pulse cyclophosphamide with serum creatinine of 1.2 mg/dl at last followup.
Discussion

Retinal disease affects around ten percent of SLE patients and are invariably bilateral. Choroidal disease is less common than retinopathy. Angiography of the fundus with fluorescein and indocyanine green often demonstrate leakage into uni/multifocal retinal detachments and choroidal ischaemia. The proposed pathogenic mechanisms are dysfunction of the retinal pigment epithelium with development of serous retinal detachment due to anti-retinal pigment epithelium antibodies and immune complex deposition in the choroid with mononuclear infiltrates resulting in choroidal vascular changes. Other complications include choroidal effusions (which have been reported to cause secondary angle closure), choroidal infarction and choroidal neovascular membrane. Lupus choroidopathy is associated frequently with lupus nephritis and central nervous system vasculitis. Shimura et al reported the effectiveness of pulse methylprednisolone and cyclophosphamide in treating bilateral exudative retinal detachment. If not successful, laser photocoagulation can also be tried.

Prevalence of optic nerve involvement in SLE is one percent. Optic neuropathy associated with SLE can manifest as acute retrobulbar optic neuritis, papillitis, anterior ischaemic optic neuropathy, posterior ischaemic optic neuropathy, or slow, progressive visual loss. It may manifest as a thrombotic, vaso-occlusive event with focal axonal necrosis, or as a general immunological inflammation, such as vasculitis. It is frequently associated with antiphospholipid antibodies. Optic neuropathy has strong predictive value for future cerebral lupus. Before optic atrophy develops, SLE-associated optic neuritis may respond to steroid treatment. Visual prognosis following optic neuropathy is generally poor. Renal biopsy provided a clue for the diagnosis in the absence of clinical findings other than ocular involvement in these patients. Both of our patients presented late to us, despite steroids and cytotoxics their visual acuity did not improve. So it is prudent to evaluate for systemic lupus erythematosus in patients especially women of child bearing age who present with ocular symptoms suggestive of anterior/posterior eye segment involvement.

Fig. 1a: Fundus showing exudative retinal detachment

Fig. 1b: Renal biopsy showing class II lupus nephritis

Fig. 2a: Fundus showing primary optic atrophy

Fig. 2b: Renal biopsy showing chronic sclerosing lesions
Conclusion

Ocular manifestations in SLE may be the presenting feature of disease. It can be sight threatening and is an indicator of active systemic disease even in the absence of other clinical features. Choroidal, retinal and optic nerve involvement in SLE require systemic immunosuppression. Early recognition, prompt assessment and coordinated treatment strategies are key to reduce ocular morbidity in SLE patients.

References


