Systemic lupus erythematosus

DESCRIPTION

Systemic lupus erythematosus (SLE) is a rheumatic connective tissue autoimmune disease. It is a chronic, multisystem disease, with relapsing and remitting phases. Damage to various organs occurs in SLE when circulating immune complexes incite local inflammatory responses. The precise cause remains unknown, but well-documented genetic predispositions have been identified (eg HLA-DR2 and HLA-DR3). Up to one-third of patients with SLE may have the lupus anticoagulant, an antiphospholipid antibody. About half of those patients have the antiphospholipid antibody syndrome, a condition associated with vaso-occlusive disease and thrombotic disorders. The syndrome may be a primary disease or may be secondary to SLE.

SYSTEMIC SIGNS

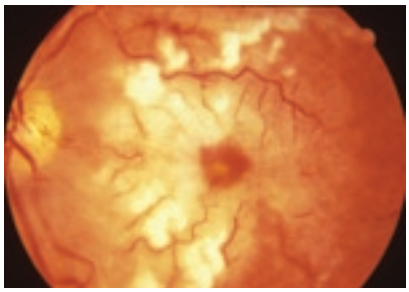
- Cutaneous disease occurs in most patients; perhaps the most distinctive manifestation is the malar or 'butterfly' rash over the nose and cheeks
- Kidney failure and infections are the leading causes of death in SLE patients
- Other areas of involvement in SLE include the skin and mucous membranes, joints (arthritis), serosal surfaces, central nervous and haematological systems.

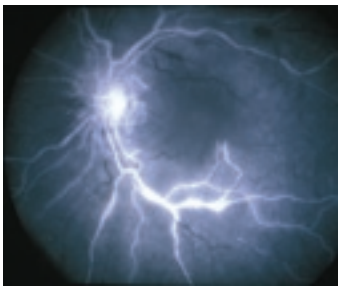
SYMPTOMS

Systemic symptoms during exacerbations may include malaise, fever, arthralgias and loss of appetite. The most common ocular symptom is burning or itching with keratoconjunctivitis sicca. SLE can cause severe loss of vision due to posterior segment complications.

OCULAR SIGNS

- Anterior segment complications may include (a) periocular skin changes, (b) keratoconjunctivitis sicca, (c) scleritis and (d) a spectrum of corneal damage ranging from marginal thinning to ulceration and neovascularisation
- Retinal vascular lesions: Vascular changes in SLE may result from microangiopathy, possibly due to immune complexes interacting with the vessel walls, or may relate to systemic hypertension. The vasculitis, particularly with antiphospholipid antibodies, may lead to confluent areas of ischaemia. Cotton wool spots and retinal haemorrhages are the most common signs. Progression to proliferative retinopathy is rare. Damage to choroidal vessels often results in multifocal serous retinal and retinal pigment epithelium detachments
- Neurological effects: vision may be affected by retrobulbar optic neuritis, anterior ischaemic optic neuropathy, cranial nerve palsies and cerebral disease.





Fluorescein angiogram of the same patient. Complete obliteration of the capillary bed and paucity of flow in the smaller arterioles, which appear like ghost vessels next to vessels filled with fluorescein

SLE with phopholipid antibodies leading to confluent areas of axoplasmic stasis with areas of increased nerve fibre swelling (bright white zones), where there is a meeting point between antegrade and retrograde axoplasmic flow

PREVALENCE

Rare (less than 1 per 10,000 per year), but more common in some populations. Up to 90 per cent of patients are female.

SIGNIFICANCE

Ocular complications tend to occur in patients with active systemic disease.

DIFFERENTIAL DIAGNOSIS

Hypertensive retinopathy, Central retinal vein occlusion, Behçet's disease, Sarcoidosis, Syphilis, Lyme disease, Cytomegalovirus retinitis, HIV retinopathy.

SEE ALSO

Branch retinal artery occlusion, Retinal detachment, Toxic retinopathy – Chloroquine, Posterior scleritis.

MANAGEMENT

Urgent

Antiphospholipid antibody syndrome may evolve rapidly, in a matter of days, requiring urgent treatment.

Blood tests

Over 95 per cent of patients test positive for antinuclear antibodies. Various additional antibody subtypes correlate with disease manifestations, eg Antiphospholipid antibodies.

Additional investigations

Fluorescein angiography documents posterior segment inflammation and response to treatment, and informs decision-making when laser photocoagulation is being considered.

Systemic treatment

Being a rheumatic disease, the mainstay of medical therapy in SLE is systemic. Initial treatments include oral corticosteroids and non-steroidal anti-inflammatory medications. Disease control may require additional immunosuppressive agents (eg cyclophosphamide, azathioprine and hydroxychloroquine). Thrombotic complications are treated with long-term anticoagulant therapy.

Review

Possible adverse ocular effects of systemic medications include cataract and glaucoma (corticosteroids) and macular toxicity (hydroxychloroquine), requiring at least annual review.

Ocular treatments

- Keratoconjunctivitis sicca is treated initially with tear supplements; additional treatments include punctual occlusion and pulse dose topical corticosteroids or cyclosporine
- Laser photocoagulation may be appropriate in cases of retinal neovascularisation
- Vitreous haemorrhage and retinal detachment are treated with vitrectomy and scleral buckling.

The full series of these articles is available in the book *Posterior Eye Disease and Glaucoma A-Z* by Bruce AS, O'Day J, McKay D and Swann P. £39.99. For further information click on the Bookstore at **opticianonline.net**

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