

OCULAR MANIFESTATIONS OF SYSTEMIC DISEASE

Congenital

Neurofibromatosis

- Melanotic hamartoma of the iris (Lisch nodules)
- The iris, eyelid, orbit and optic nerve may be involved
- Lisch nodules may be used in screening - 95% of NF 1 patients have the nodules by age 6 years

Vascular

Hypertension

- Flame-shaped haemorrhages of the retina
- Cotton-wool patches
- Copper and silver wiring of the arteries (long standing hypertension)
- Lipid exudates
- Optic disc swelling (malignant hypertension)

Other causes of optic disc swelling – sarcoidosis, syphilis, tumour, pseudo disc oedema from optic nerve head drusen

Embolic

- Amaurosis fugax – central retinal artery occlusion usually from dislodged cholesterol plaques from the carotid bifurcation (Hollenhorst plaque)
- Cardiac sources – AF, atrial myxoma, valve prolapse

Central retinal vein occlusion (CRVO)

Found in 50% of patients with open angle glaucoma and/or systemic hypertension

- Retinal haemorrhages
- Cotton wool spots
- Macular oedema
- Blindness
- Retinal and iris neovascularisation

Autoimmune disorders

Eye signs may be the only manifestation

Sjogren's syndrome

- Dry eyes (keratoconjunctivitis sicca) and dry mouth
- Ankylosing spondylitis
- Eye signs in 25% of cases

- Photophobia, redness and reduced visual acuity

Rheumatoid arthritis

- Scleritis, episcleritis, keratoconjunctivitis sicca and peripheral corneal ulceration

Juvenile rheumatoid arthritis (JRA)

- Pauciarticular form, rh negative, ANA positive)
- Triad of iritis, cataract and band keratopathy (whitish calcium deposit in the cornea)

SLE

- Dry eye
- Scleritis,
- Peripheral corneal ulceration
- Cotton wool spots
- Retinal haemorrhages and oedema
- Hard exudates
- Microaneurysms
- Arterial narrowing
- Venous engorgement and vascular tortuosities
- Lupus vasculitis

Giant cell (temporal) arteritis

- A form of systemic vasculitis
- Ischaemic optic neuropathy
- Unilateral visual loss
- Pale, swollen optic nerve head
- Central retinal artery occlusion-initial presentation in 10% of cases
- Third nerve palsy with papillary sparing due to occlusion of the blood supply if the nerve (ptosis and outward deviation of the eye)

Reference

1. *Ocular manifestations of systemic disease. American Academy of Ophthalmology. 1989, 1996*
2. *Retinal and choroidal manifestations of systemic disease. Copyright 1991. William and Wilkins Baltimore, USA.*