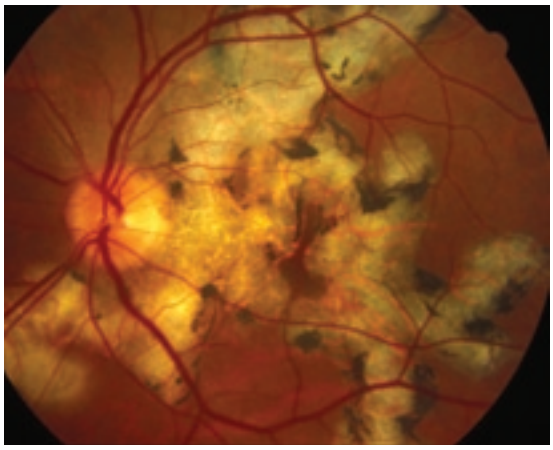
Serpiginous chorioretinitis

Geographic choroiditis, geographic helicoid choroiditis

DESCRIPTION

Serpiginous chorioretinitis is a chronic, bilateral, recurrent and progressive chorioretinitis. It is often classified among the white dot syndromes, which include acute posterior multifocal placoid pigment epitheliopathy, Birdshot chorioretinitis and multiple evanescent white dot syndrome. The age of onset is usually between 30 and 60 years, with a slight male predominance. The aetiology is unknown.

Acute lesions appear as pale subretinal patches which eventually result in areas of chorioretinal scarring and atrophy. Macular involvement and choroidal neovascularisation are potential sight-threatening complications; patients therefore require education and regular, long-term review.



Chorioretinal lesions are seen in differing stages of evolution. Between disc and macula are the oldest chorioretinal scars and the areas along the superior vascular arcade and inferior to the disc are more recent

SYMPTOMS

The usual presentation is painless monocular loss of vision, the severity of which depends on the proximity of lesions to the fovea. Involvement of the fellow eye may be delayed by several weeks, or even years. Recurrences typically occur at intervals of months to years.

SIGNS

Active inflammatory lesions appear as grey-yellow subretinal patches. They often first appear near the optic disc and extend along the vascular arcades in a snakelike (serpiginous) pattern. Lesions are usually contiguous, but may be separate. Mild vitritis and areas of vascular sheathing occasionally accompany acute lesions.

The acute phase usually lasts several weeks, and is followed by progressive, atrophic chorioretinal scarring and RPE changes which often expose the underlying choroidal vessels. Disease recurrences characteristically border these atrophic areas, resulting in a geographic (map-like) pattern of scars with pale, oedematous edges representing active disease. Choroidal neovascularisation is a sight-threatening complication of established serpiginous chorioretinopathy that has been reported to occur in up to 25 per cent of cases.

When the patient presents early (with only acute lesions), fundoscopy may be suggestive of acute multifocal placoid pigment epitheliopathy (APMPPE). Repeat examination is critical in these cases. APMPPE usually affects both eyes relatively symmetrically; lesions generally follow a similar time course and resolve in weeks. Scarring and disease recurrences are rare; and the prognosis for vision is highly favourable compared with serpiginous chorioretinopathy.

PREVALENCE

Rare; slight male predominance; usually begins in young adulthood to middle age.

SIGNIFICANCE

May lead to severe loss of central vision.

DIFFERENTIAL DIAGNOSIS

Acute posterior multifocal placoid pigment epitheliopathy, Toxoplamosis, Angioid streaks.

SEE ALSO

Multiple evanescent white dot syndrome, Birdshot chorioretinopathy, Choroidal neovascularisation.

MANAGEMENT

Ocular tests and imaging investigations

Visual field testing reveals absolute scotomas corresponding to affected areas. Intravenous fluorescein angiography aids management decisions by identifying neovascular membranes and areas of acute inflammation. Active lesions block choroidal fluorescence early and stain late – initially at the edges then spreading centrally.

Medications

In the absence of established evidence-based treatment recommendations, the medical management of serpiginous chorioretinitis is controversial. Systemic corticosteroids are commonly administered during acute episodes, and may be continued at a lower maintenance dose between recurrences. Other immunosuppressive medications may also be employed as steroid-sparing or additional treatments.

Laser and injection treatments

Laser photocoagulation or photodynamic therapy with verteporfin (Visudyne, Novartis) is applied to choroidal neovascular membranes when central vision is threatened. CNV may also be treated with repeated doses of anti-angiogenic drugs: Anecortave acetate (Retaane, Alcon) is an angiostatic cortisene delivered by periocular injection; Pegaptanib sodium (Macugen, Eyetech) and ranibizumab (Lucentis, Genentech and Novartis) are anti- vascular endothelial growth factor (anti-VEGF) drugs delivered by an intravitreal injection.

Prognosis

Most patients retain satisfactory vision in at least one eye.

Review and self-monitoring

During periods of disease inactivity, review is recommended at six-month intervals. Patients are advised to report new visual symptoms promptly, and may be supplied with equipment for home vision monitoring.

Refractive correction or LVAs

Optimal refractive correction and low vision aids are often valuable.

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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