Necrotising herpetic retinopathies

Acute retinal necrosis and progressive outer retinal necrosis
Varicella zoster virus, chicken pox virus

DESCRIPTION

The necrotising herpetic retinopathies constitute a spectrum of conditions involving acute, rapidly progressive retinal necrosis, often resulting in retinal detachment. Current opinion is that most cases are caused by reactivation of latent herpes virus infections – predominantly herpes simplex virus, varicella zoster virus and cytomegalovirus. Rare cases have been associated with acute viral meningoencephalitis. Intracellular viral replication is thought to induce vasculitis, vascular occlusion and cell death.

SYMPTOMS

Early cases confined to the peripheral retina are often asymptomatic. With disease progression, patients may experience floaters, decreased peripheral vision, blurred vision and ocular pain. The patient may recall previous episodes of chicken pox, shingles or cold sores. Other relevant considerations include general health, immunosuppressive medications and risk factors for HIV infection.

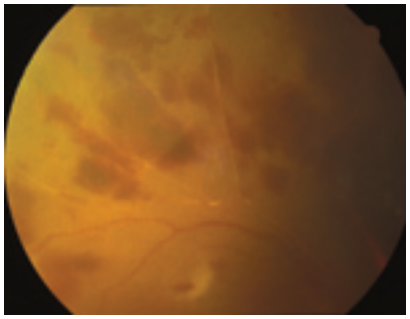
SIGNS AND CLASSIFICATION

1) Acute retinal necrosis (ARN) denotes the presence of the following features:

- Patches of peripheral retinal oedema and necrosis which spread circumferentially and posteriorly
- Inflammation and occlusion of retinal and choroidal vessels, with pale sheathing of retinal vessels
- Vitritis, where inflammatory cells and debris in the vitreous may obscure the fundus
- Rhegmatogenous retinal detachment, in up to 75 per cent of untreated cases.

ARN becomes bilateral in at least one-third of patients, usually within several weeks. Without treatment, the inflammatory phase typically lasts six to 12 weeks. The necrotic retina develops holes or breaks which predispose to retinal detachment.

2) Progressive outer retinal necrosis (PORN) describes necrotising herpetic retinopathy with multifocal, poorly-demarcated pale retinal lesions. The lesions rapidly progress to confluent, full-thickness areas of retinal necrosis. In contrast to ARN, there is minimal evidence of intraocular inflammation, rapid progression to total retinal detachment and an extremely poor visual prognosis. PORN generally occurs in severely immunocompromised patients (often with HIV/AIDS) unable to sustain an effective immune response to viral proliferation. Involvement of the fellow eye with bilateral loss of light perception often develops within weeks.



Retinal periphery: Acute retinal necrosis with pale necrotic retina, sheathing of retinal vessels and scattered haemorrhages

PREVALENCE

Very rare. ARN usually occurs in otherwise healthy patients, and less commonly in association with varying degrees of immunosuppression.

SIGNIFICANCE

Examination of the peripheral retina is essential in all cases of unexplained acute loss of vision. Prompt treatment of ARN reduces the incidence of involvement of the fellow eye, and minimises loss of vision.

DIFFERENTIAL DIAGNOSIS

Cytomegalovirus retinitis, Syphilis, Toxoplasmosis, Behcet's disease, Intraocular lymphoma.

SEE ALSO

Retinal detachment – Classification, Retinal break or tear, AIDS retinopathy.

MANAGEMENT

Modern treatments – including antiviral therapy, laser photocoagulation and retinal detachment surgery – have improved outcomes in ARN. Up to half of patients retain visual acuity of at least 20/40 (6/12) following prompt diagnosis and treatment.

Investigations

ARN is a clinical diagnosis. Additional investigations to assist management may include intravenous fluorescein angiography, visual fields, fundus photographs and B-scan ultrasonography. Serological tests may be indicated for HIV, syphilis and toxoplasmosis. Cerebral imaging and lumbar puncture are required when central nervous system infection is suspected. Viral studies may be performed for epidemiological reasons.

Medications

Patients with ARN are admitted to hospital for intravenous antiviral therapy. This usually halts the progression of the retinitis, reduces the risk of involvement of the fellow eye and is followed by several months of oral antiviral therapy. Optimal treatment of PORN is unknown – visual loss often progresses or recurs despite antiviral therapy. Other treatments that may be considered include topical and/or systemic corticosteroids, topical cycloplegic and analgesic therapies.

Laser and incisional surgery

Laser photocoagulation posterior to areas of active retinitis discourages posterior extension of retinal detachment. Since it does not prevent extension of retinitis, repeat treatment is often necessary. Retinal detachment is often difficult to treat: additional procedures may be required, including scleral buckling, vitrectomy and tamponade with gas or silicone oil.

Review

Once stabilisation of ARN is achieved, three-monthly review is recommended for the first year, followed by yearly examinations. Although rare, the fellow eye may become involved at any time, even decades after the initial episode.

The full series of these articles will be 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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