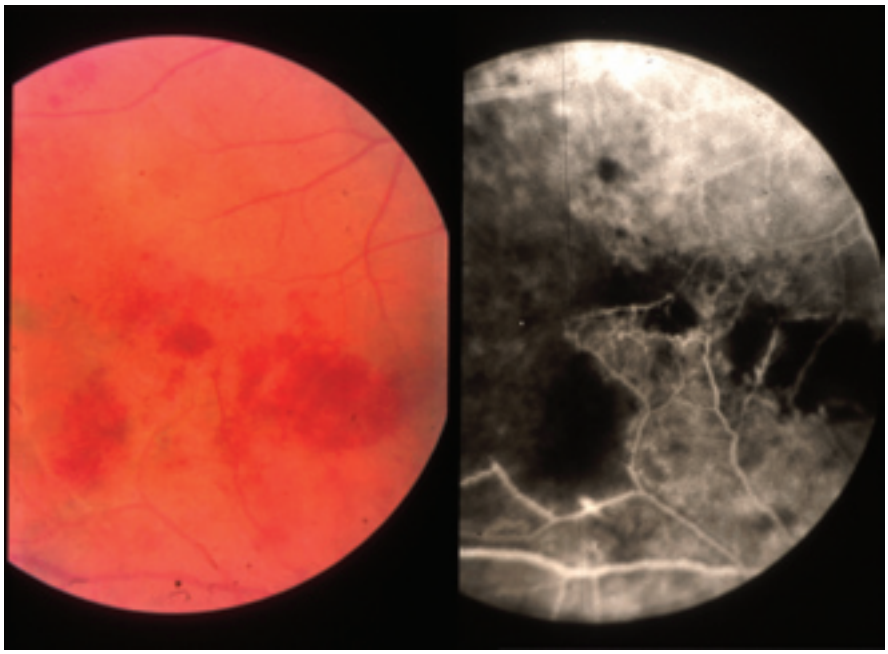
Behçet's disease – syndrome



Left: Fundus photograph of Behçet's disease showing retinal hemorrhages.
Right: Fluorescein angiogram showing confluent retinal ischaemia (hypofluorescence) temporal to the haemorrhages

DESCRIPTION

Behçet's disease is a chronic multisystem inflammatory syndrome that may include anterior uveitis, posterior uveitis and retinal vasculitis. The disease has a characteristic diagnostic triad of iritis, oral and genital ulcers. Often the uveitis is of most concern in patients with the disease, as it is usually bilateral and, with recurrent severe episodes, it is potentially blinding. However, Behçet's disease may affect many body systems, including the pulmonary, gastrointestinal and central nervous systems, making the condition potentially life threatening.

Although the onset of the condition may be at any age, the most common onset is at age 20-30 years. The severity of the inflammatory episodes tends to decrease in the later years, although there is a cumulative effect on morbidity owing to the systemic disease. The aetiology of Behçet's disease is not known, but infectious, immunological and genetic factors may all contribute.

SYMPTOMS

If uveitis is present in Behçet's disease, then ocular symptoms may include blurring of vision, photophobia and ocular pain, usually bilaterally.

SIGNS

In mild cases of Behçet's disease, the oral or genital ulcers, or other skin lesions, may be the only findings; however, occasionally ocular disease may be the initial finding. When present, the uveitis may be severe and recurrent, with hypopyon, iridocyclitis, posterior synechiae, papilloedema, vitritis and cystoid macula oedema. An occlusive vasculitis may also occur, with retinal haemorrhages, oedema and vascular sheathing.

PREVALENCE

The prevalence of Behçet's disease varies between countries. It is uncommon to rare (1 in 1,000–10,000) in Turkey, Iraq, Iran, Korea and Japan, but very rare (less than 1 in 100,000) in countries such as the US, UK and Germany. The variation relates to both genetic and environmental factors.

SIGNIFICANCE

Behçet's disease is potentially sight and even life threatening, requiring prompt investigation and treatment.

DIFFERENTIAL DIAGNOSIS

Other systemic causes of uveitis, including sarcoidosis, syphilis and tuberculosis. Other ocular causes of anterior chamber reactions include rhegmatogenous retinal detachment, posterior segment tumour and intraocular foreign body.

MANAGEMENT

Urgent

Uveitis associated with Behçet's disease is vision threatening and requires urgent investigation and treatment. Referral to a specialist in inflammatory disease may be also be required for management of the systemic conditions.

Laboratory tests

Behçet's disease is positive for erythrocyte sedimentation rate, antinuclear antibodies, C-reactive protein and serum haplotyping, although these results may not be diagnostic.

Additional investigations and office procedures

Behçetine skin test, in which the skin is pricked with a sterile needle; the formation of a pustule within a few minutes is a positive result. Fluorescein angiography will show extensive early vascular leakage.

Topical medication

Uveitis may be managed with steroids (for example, prednisolone acetate 1 per cent *q2h* to *q6h*, depending on severity). More severe disease may require sub-Tenon steroid injection.

Oral medication

Oral steroid medication may be required for management of the systemic disease or more severe ocular disease. Other drugs may also be needed, such chlorambucil, cyclophosphamide or ciclosporin.

Review

The uveitis tends to have an episodic course, and regular review is needed.

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 & Swann P (Elsevier Butterworth-Heinemann, August 2006, £39.99). For further information, including ordering, please click on the Bookstore link at www.opticianonline.net.