Birdshot Retinochoroidopathy

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Background

Birdshot retinochoroidopathy (BSRC) is an uncommon, bilateral inflammatory disease affecting the choroid layer of the eye (between the outer sclera and inner retina). It belongs to a group of disorders known as the idiopathic multifocal white dot syndromes, which themselves are one of the causes of chronic posterior uveitis. It was first reliably identified back in 1949 and variously called ‘candle wax spot retinochoroidopathy’ and ‘rice grain retinochoroidopathy’, which gives an idea of what is seen on examining these patients (see 'Presentation', below). It only gained formal recognition as it is currently known in the early 1980s. Its cause is not entirely clear but it is thought to be related to an inherited immune dysfunction, as there is a strong association with HLA-A29 positivity (about 90% of patients) and, to a lesser extent, HLA-B12 positivity. It is the strongest HLA association with any known disease. There are progressive abnormalities which develop in the small choroidal vessels, resulting in leakages and, eventually, atrophic spots. There is an associated low-grade uveitis which can eventually lead to permanent loss of vision.

Epidemiology

This is rare condition (there are no precise figures but in a multicentre study covering 14 eye clinics over six years, only 102 cases were identified), most commonly affecting Caucasian patients. It tends to occur later in life than many other types of uveitis (average age 50 years, ranging from 40-70 years) and possibly slightly more often in females.

Presentation

History

Patients typically present with a (usually) painless, gradual, bilateral blurring of vision disproportionate to their recorded visual acuity. It is frequently associated with nyctalopia (trouble seeing at night), floaters and disturbance of colour vision. Other symptoms reported have included glare, photopsia (flashes of light), photophobia, shimmering vision, decreased peripheral vision, difficulty in adapting to different levels of lighting and difficulty with seeing objects against a background of another object (poor contrast sensitivity).

Examination

- Function - in the initial stages, visual acuity reduction may be relatively mild (of the order of 6/9-6/12). This gets progressively worse throughout the disease. There is also often some degree of visual field loss.
- Anterior segment of the eye - the eye usually looks quiet. There is no redness and, if you have access to a slit lamp, there are no corneal changes or cells in the anterior chamber (a sign of anterior uveitis).
- **Posterior segment of the eye** - this is where the abnormalities lie. If you are able to focus on the vitreous (jelly filling the greater part of the globe, lying behind the lens, through to the retina), look for cells - seen as little pale specks - floating about. There are distinctive lesions on the retina: these are well-defined (initially) pale yellow/white spots scattered around the retina, usually much smaller than the disc size. As the current (and previous) name suggests, they look somewhat like an explosion of spots scattered over the retina. Over the years, the lesions become ill-defined and confluent. There are several patterns of distribution described - these will be noted by the ophthalmologist, as they have a bearing on the prognosis. There may be associated macular oedema and intraretinal haemorrhages.

**Differential diagnosis**

Other types of idiopathic multifocal white dot syndromes:

- Acute multifocal posterior placoid pigment epitheliopathy (AMPPPE).
- Serpiginous choroidopathy.
- **Multifocal choroiditis with panuveitis**.
- Multiple evanescent white dot syndrome.
- Acute retinal pigment epitheliitis.

Other causes of posterior uveitis including:

- Various causes of choroiditis (such as toxoplasmosis).
- Sarcoidosis.
- Syphilis.
- Ocular histoplasmosis.
- Postoperative endophthalmitis.
- Problems relating to immunocompromise (e.g., cytomegalovirus (CMV) retinitis, candidal infection, herpetic retinitis and so on).
- Retinal vasculitic diseases.
- Other less common causes.

**Investigations**

Initial diagnosis is usually suspected on examination in the eye clinic - this is confirmed with HLA testing and vascular studies including fluorescein angiography (imaging of retinal, choroidal and optic disc vasculature) and indocyanine green angiography (better evaluation of choroidal vasculature in particular). Ultra-high resolution optical coherence tomography (OCT) is very helpful if available. Electroretinogram (measures electrical responses by the various cells of the retina) should also be carried out and responses are diminished in birdshot retinochoroidopathy (BSRC). If there are real uncertainties over the diagnosis, a biopsy may be performed.

**Associated diseases**

The patient is usually otherwise healthy. There have been studies suggesting links to glaucoma and rhegmatogenous retinal detachment but these tend to be weak and findings are probably spurious. Furthermore, vascular disease, sarcoidosis, psoriasis, autoimmune sensorineural hearing loss and vitiligo have been reported in these patients but, again, a clear association is not currently evident.

**Staging**

There are several described distribution patterns of the lesions relating to how widely scattered they are, whether they are symmetrically scattered and whether the macula is involved or not. There is no birdshot retinochoroidopathy (BSRC) staging system as such but it is described as being acute or chronic - in the latter, lesions tend to be confluent (giving the appearance of larger spots on retinal examination).

**Management**
If birdshot retinochoroidopathy (BSRC) - or indeed any of the differentials outlined above - is suspected in the primary care setting, a referral to the local ophthalmology department is mandatory. The urgency depends somewhat on the symptoms but it is best to discuss it with the team within a few days.

There are no definite treatment strategies. Some patients with minimal symptoms and good vision may not require treatment.

Others may benefit from one or more corticosteroids (periocular ± systemic), ciclosporin or cytotoxic agents - particularly where there is macular oedema. These agents may also help to reverse some of the visual field loss. Daclizumab has been found to help stabilise the vision and decrease intraocular inflammation. It is not currently available in Europe (removed for commercial, not safety, reasons).

Some patients develop neovascular membranes which may be amenable to laser treatment, photodynamic therapy or surgery. Treatment will be guided by the severity of the disease, the location of the lesions and the presence of any complicating factors.

Complications

- Macular oedema is the most common complication. Treatment is with topical steroids ± topical non-steroidal anti-inflammatory drugs.
- Subretinal choroidal neovascularisation (this increases risks of vessel leaks leading to oedema and bleeds). Treatment is with laser photoocoagulation.

Prognosis

The natural course of the disease is one of episodic fluctuation and remission of symptoms. Ultimately, about 20% of patients have a self-limiting course with preservation of vision. However, most follow a chronic course over many years, resulting in impaired visual acuity or related problems (such as nyctalopia or colour impairment).

Prevention

Birdshot retinochoroidopathy (BSRC) cannot be prevented. Once the diagnosis is made, preventative measures revolve around early identification and treatment of vision-threatening complications. All patients complaining of floaters will be followed up, even if central visual acuity is stable.

Further reading & references

2. Clinical Ophthalmology A Systematic Approach
3. Samson MC et al; Retinopathy Birdshot, Medscape, Sep 2008

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