Choroidal disorders

The choroid: what is it?

The choroid is the pigmented, highly vascular layer of the globe, lying between the sclera (on the outside) and the retina (on the inside). It is one of the three components of the uveal tract and is shaped a little like the body of a rounded wine glass.

The optic nerve emerges at its base and the other two components of the uveal tract sit anteriorly (the ciliary body lies around the rim and the iris stretches over the opening). It is made up of three layers, each of which can be affected by disease processes. There is the external vessel layer, the capillary layer and the internal sheet-like Bruch's membrane.

The main function of the choroid is to nourish the outer layers of the retina but it is also thought to:

- Regulate retinal heat.
- Assist in the control of intraocular pressure.
- Provide pigment absorption of excess light, so avoiding reflection.

The choroid can be subject to inflammatory disorders (often in conjunction with the retina - see the separate Chorioretinal Inflammation article), tumour formation, and a number of other disorders outlined below.

Hypertensive choroidopathy

Systemic hypertension affects ocular vasculature at various levels. Within the choroid, certain anatomical and histological characteristics make it particularly susceptible to the effects of severe systemic hypertension (chronic or acute).

Choroidal detachment and haemorrhage

Nature

- The choroid detaches from the underlying sclera with an associated accumulation of serum-like fluid or blood.
- Causes of choroidal detachment include a low intraocular pressure associated with eye surgery, blunt trauma, inflammatory diseases (eg, scleritis), intraocular tumours, uveal effusion syndrome, and treatment with acetazolamide.[1]
- More unusually, a choroidal detachment can occur on doing the Valsalva manoeuvre or spontaneously, particularly in older patients (>65 years) on anticoagulants.

Presentation

- Photopsia and floaters. Where there is a large detachment, there may be a visual field defect.
- Pain is not generally a feature unless it is a postoperative haemorrhagic detachment, in which case there is sudden, excruciating pain with loss of vision (these are almost pathognomonic symptoms).

Investigations

It is diagnosed on examination of the fundus. Occasionally, this needs to be confirmed with ultrasonography.

Treatment

- As soon as the diagnosis is made, topical steroids, cycloplegics and mydriatics are given.[2] Intraocular pressure-lowering drugs are also given.
- Surgery may be required depending on the nature of the detachment.[3]
- Operative choroidal haemorrhage tends to occur during surgery and is managed there and then in theatre.

Prognosis

- This depends on the cause and extent of the detachment. Damage can occur anteriorly (corneal endothelial damage, anterior synechiae, lens opacities) and posteriorly (maculopathy).
- Chronic detachment can also lead to globe phthisis (where it wastes away).
- Haemorrhagic choroidal detachment is associated with greater morbidity and may cause loss of useful vision.

Choroidal rupture

Nature

- A choroidal rupture occurs when there is a tear in the choroid, Bruch's membrane, and the retinal pigment epithelium.
- Indirect blunt trauma that occurs at the posterior pole away from the site of impact is the most common mechanism.[4] Direct blunt trauma is a less common mechanism.
- Acute preretinal, retinal, or subretinal haemorrhaging follows a rupture.

Presentation

Patients may be asymptomatic but they may equally complain of decreased vision or of a paracentral scotoma.

Investigations

- The diagnosis is made on examination but there may be a need for ultrasound or CT/MRI investigations if additional injury is suspected.
- There may be a place for fluorescein angiography where the patient presents years later with complications such as the formation of a neovascular membrane.

Treatment

- Most patients do well with conservative management.[4]

- They will be monitored for about five years to assess for the formation of a neovascular membrane ± associated delayed bleeding. Should this occur, laser treatment may be needed.

Prognosis

- Patients with uncomplicated choroidal ruptures have a good chance of full recovery.
- May cause choroidal neovascularisation, which can lead to haemorrhagic or serous macular detachment. This most often occurs during the first year after the injury but has been reported up to five years after injury.
- Another complication is the formation of an epiretinal membrane. This membrane can be removed surgically.

Choroidal folds

Nature

These are parallel grooves involving Bruch's membrane. When looking at them on fundoscopy, they give the impression of a sheet of cling film that is being pulled taught centrally around the disc. There are a number of causes, including orbital disease (such as retrobulbar tumours), choroidal tumours, ocular hypotony (following significant surgery) and a number of other miscellaneous causes (idiopathic, chronic papilloedema and posterior scleritis).[5]

Presentation

Many patients are asymptomatic but they may complain of metamorphopsia or impaired vision over time.

Investigations

Fluorescein angiogram.

Treatment

This depends on the underlying cause but asymptomatic folds do not require treatment.

Prognosis

This depends on the underlying cause.

Choroidal dystrophies[6]

Choroideremia

This is a very rare condition that is inherited in an X-linked recessive manner and only affects males. Patients present in the first decade of life with an inability to see in dim conditions (nyctalopia) and visual field loss. A number of abnormalities can be seen both on examination of the fundus and on functional testing of the retina. Vision is usually usefully retained until about the sixth decade of life, after which there is severe visual loss. No treatment is currently available.

Gyrate atrophy

This is another inherited disorder (autosomal recessive) characterised by axial myopia (short-sightedness caused by a long axis between the front and the back of the eye) and nyctalopia. There will be chorioretinal atrophy present and elevated ornithine levels in the plasma, urine, cerebrospinal fluid and aqueous humour (due to the lack of the ornithine-degrading enzyme, ketoacid aminotransferase). The extraocular features are absent or subtle. Two subgroups of patients are identified based on their response to treatment with vitamin B6 (pyridoxine): those responsive to treatment have a more slowly progressing course of the disease.

Central areolar choroidal dystrophy

An autosomal dominant inherited condition presenting in the fourth to fifth decade of life, with poor central vision and nyctalopia. Both eyes tend to be affected and the prognosis is poor with severe visual loss by the sixth to seventh decade of life.

Diffuse choroidal atrophy

Another autosomal dominant inherited condition similar to central areolar choroidal dystrophy. Patients tend to present a decade earlier and early macular involvement rapidly impairs vision.

Helicoidal peripapillary chorioretinal degeneration

An autosomal dominant condition presenting in childhood, characterised by strips of choroidal atrophy that variably affect the individual: there may be early age visual loss or elderly mild visual impairment.

Pigmented paravenous retinochoroidal atrophy

A rare condition usually found in young men, with no clear inheritance pattern, and a good prognosis due to rarity of macular involvement.

Choroidopathy: the idiopathic multifocal white dot syndromes[7]

This is a heterogeneous group of conditions characterised by chorioretinal inflammation.[8] Some of these conditions are associated with systemic infection and they all broadly have similar treatment options: immunosuppressive therapy, laser photocoagulation, topical or systemic steroid therapy, photodynamic therapy and, most recently, antivascular endothelial growth factor agents.

Acute posterior multifocal placoid pigment epitheliopathy (APMPPE)

- **Nature** - this is an uncommon, idiopathic, bilateral, self-limiting condition.[9] Often associated with HLA-B7 and HLA-DR2.
- **Presentation** - third to fifth decade in life, after a flu-like illness in about one third of the cases: progressive subacute impairment in one eye, followed by the other a few days later.
- **Investigations** - fluorescein angiography.
- **Treatment** - none.
- **Prognosis** - generally very good, although in those patients who have recurrent episodes, a residual paracentral scotoma may remain.

Serpiginous choroidopathy

- **Nature** - uncommon idiopathic bilateral progressive disease with a prolonged, fluctuating course over many years. Relapses occur after several months of remission.
- **Presentation** - fourth to sixth decade in life. There is unilateral blurring of vision and image distortion (metamorphopsia) with the fellow eye being affected some time later (this time period varies).
- **Investigations** - fluorescein angiography.
- **Treatment** - monotherapy: ciclosporin or triple therapy: systemic steroids, azathioprine and ciclosporin; stem cell treatment.[10]

- **Prognosis** - generally poor as recurrent episodes cause progressive atrophy and 50% of patients have foveal involvement, so markedly affecting vision.

Birdshot retinochoroidopathy[11]

- **Nature** - rare, autoimmune bilateral progressive and permanent disease.
- **Presentation** - most commonly in the fifth decade of life. Early symptoms are floaters and blurred vision. Patients may experience visual blurring that is disproportionate to their visual acuity; acuity may be relatively well preserved. Later symptoms include nyctalopia (night blindness), loss of contrast sensitivity, and loss of depth perception.
- **Investigations** - fluorescein angiography, indocyanine green angiography, and electroretinography. In patients with a typical clinical appearance of birdshot chorioretinopathy, testing for HLA-A29, syphilis serology, serum ACE, interferon gamma release assay (IGRA), chest X-rays may help support the diagnosis or exclude other causes.
- **Treatment** - steroid-sparing immunomodulatory therapy, such as methotrexate, mycophenolate mofetil, ciclosporin and intravenous immunoglobulin. Systemic corticosteroids may be used as an initial or rescue therapy to bridge to other immunomodulatory therapies.
- **Prognosis** - about 20% of patients experience a self-limiting course with preserved vision. However, in others, it can lead to significant visual impairment, although central visual acuity may remain preserved until late in the disease. Good data on long-term outcomes with immunomodulatory treatment are lacking.

Punctate inner choroidopathy (PIC)

- **Nature** - uncommon, idiopathic bilateral disease usually affecting myopic women.[12]
- **Presentation** - fourth to fifth decade of life. Blurring of central vision ± photopsia (seeing flashing lights).
- **Investigations** - fluorescein angiogram.

- **Treatment** - in certain cases, patients may be amenable to laser photocoagulation, systemic steroids or surgery (where neovascular membranes are excised).
- **Prognosis** - reasonable if the lesions do not involve the macula.

Multifocal choroiditis with panuveitis (MCP)

- **Nature** - uncommon, recurrent disease which is usually bilateral and associated with a panuveitis. Its aetiology is not clearly identified but the Epstein-Barr virus may have a role.
- **Presentation** - fourth decade (female:male = 3:1) with blurring of vision, floaters and photopsia.
- **Investigations** - fluorescein angiogram; optical coherence tomography.[13]
- **Treatment** - systemic steroids have been shown to be effective in at least 50% of cases. Immunosuppressive agents are also used in order to reduce the amount of systemic steroids required long-term. Some patients may also benefit from laser photocoagulation.
- **Prognosis** - this condition recurs over months and frequent foveal involvement gives rise to a relatively poor prognosis.

Multiple evanescent white dot syndrome (MEWDS)

- **Nature** - uncommon, idiopathic and self-limiting condition. It is usually unilateral.
- **Presentation** - third to fifth decade, more often in females than in males (4:1). The patient complains of decreased vision and photopsia, most often in the temporal visual field.
- **Investigations** - fluorescein angiogram and/or optical coherence tomography.[14]
- **Treatment** - none.
- **Prognosis** - full recovery after a few weeks.

Acute zonal occult outer retinopathy (AZOOR)[15]

- **Nature** - possibly part of a spectrum of diseases comprising - among others - PIC, MCP and MEWDS. It is uncommon, may be unilateral or bilateral and typically affects young/middle-aged women after a flu-like illness.
- **Presentation** - acute scotoma (worse in bright light) and photopsia. There may be an associated vitritis.
- **Investigations** - electrodiagnostic tests are invariably abnormal.
- **Treatment** - many use immunosuppression but its benefit hasn't been fully proven yet.
- **Prognosis** - although one third of patients may develop recurrence and have a poorer visual outcome, the majority of patients have one episode with good visual recovery.

Further reading

- PIC Society

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