VISION SERVICES

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Retinal Artery Occlusions

There are two sources of blood supply to the retina:

- The outer retinal pigment epithelium (RPE), the photoreceptors and a few of the overlying tissue layers are supplied by the choroid, itself supplied by the various ciliary arteries (branches of the ophthalmic artery - itself a branch of the internal carotid artery).
- The inner neural retina is supplied by the central retinal artery - directly derived from the ophthalmic artery. After emerging from the centre of the optic nerve, it divides into two equal superior and inferior branches nasally (medially) and temporally (laterally) which supply all the inner layers of the neural retina.

The clinical significance of the two blood supplies is that, depending on the problem and at what level it occurs, different layers of the retina can be affected or spared. Thus, an embolus occurring proximally in the ophthalmic artery will have a devastating effect, as all layers of the entire retina lose their blood supply, whereas a small embolus lodged in a distal end branch of the retinal artery will only affect the inner neural retina of that part of the retina, sparing the photoreceptors and limiting any symptoms.

This article is concerned with the relatively common problem of retinal artery occlusion. This may occur centrally, as the retinal artery emerges from the optic nerve and hasn't yet branched out - central retinal artery occlusion (CRAO)

Pathophysiology of retinal artery occlusions

There are a number of ways in which the retinal artery can become blocked. The most common one is by an embolus. Alternatively, there may a sudden narrowing of the vessel (eg, haemorrhage into an atheromatous plaque) or inflammation. The associated diseases include:

- Atherosclerosis-related thrombus - this accounts for about 80% of CRAOs, with hypertension accounting for about 60% of those cases. Diabetes exists in 25% of cases.
- Embolism:
  - Carotid: this may be a cholesterol, fibrinoplatelet or calcific embolus.
  - Cardiac: this may be calcific, vegetations from the cardiac valves (endocarditis) or a mural thrombus (eg, in atrial fibrillation).

    - Aortic disease (including dissection) may be another embolic source.
- Inflammatory - such as giant cell arteritis, polyarteritis nodosa, systemic lupus erythematosus (SLE), Wegener's granulomatosis,Takayasu's arteritis, pancreatitis.
- Thrombophilic disorders such as antiphospholipid antibody syndrome, protein S/C deficiencies, leukaemias or lymphomas.
- Infectious origin - toxoplasmosis, mucormycosis, syphilis.
- Pharmacological causes - oral contraceptive pill, cocaine.
- Ophthalmic causes - severely raised intraocular pressure,[2] trauma or optic nerve drusen.
- Rarely, retinal migraine (vasospasm)[2] - this might be seen in young individuals where all other causes have been excluded.

Damage becomes increasingly lethal to the retina over time. The optimal treatment window during which something can be done is somewhat controversial but believed to be about 90-100 minutes (some say up to 105 minutes)[4] although some form of treatment will usually be attempted within 24-48 hours of onset.

Central retinal artery occlusion

Description

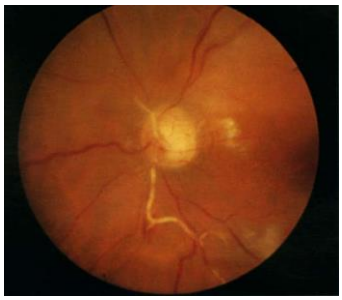
CRAO: the central retinal artery occludes before it branches out as it emerges from the optic nerve, so resulting in almost complete hypoxia of the inner retina.

Incidence

- It has an estimated incidence of 0.85/100,000/year.[1]
- It is one of the more common causes of blindness in the elderly. The majority of patients are aged >60 years. Younger patients usually have valvular heart disease.
- Men are slightly more affected than women.

Presentation

- Sudden (over a few seconds), unilateral painless visual loss.
- In 94% of cases, vision is usually reduced to counting fingers (worse suggests that the ophthalmic artery may also be affected).
- There may be a history of amaurosis fugax (amaurosis fugax precedes loss of vision in up to 10% of patients).
- In 1-2% of patients, the loss is bilateral (although the degree is not necessarily the same in each eye).[3]



Examination reveals an afferent pupillary defect, a pale retina with attenuation of the vessels. There may be segmentation of the blood column in the arteries ('cattle-trucking') and the centre of the macula (supplied by the intact underlying choroid) stands out as a cherry-red spot.

Systemic examination should include carotid auscultation for bruits, heart sounds for murmurs, radial pulse for atrial fibrillation and blood pressure.

Investigations

In the acute setting, diagnosis is usually clinical and investigations are aimed at ruling out underlying diseases (see associations in 'Pathophysiology of retinal artery occlusions' above). The most important cause to rule out is giant cell arteritis because, with appropriate and timely intervention, the visual loss is reversible and the fellow eye will be protected. Occasionally, there may be doubt about the diagnosis if the presentation or findings are atypical. Fluorescein angiography and optical coherence tomography may be helpful.

Management

Although there is no generally agreed treatment regime, there are some steps taken in the very early stages which *may* prevent irreversible loss of vision.

This is therefore an ophthalmological emergency!

Depending on the cause, different approaches may theoretically have different success. It is not always evident what the cause is in the acute setting so the following generic steps are tried:

- If the patient presents within 90-100 minutes of onset of symptoms, you could try firm ocular massage (repeatedly massage the globe over the closed lid for ten seconds with five-second interludes) - the idea is to try to dislodge the obstruction. However, this only works very occasionally and immediate referral is mandatory.
- Lowering of intraocular pressure with an an anterior chamber paracentesis may be attempted (withdrawal of a little fluid from the anterior chamber under local anaesthetic, done in the clinic) and acetazolamide given but, again, the outcome is variable. Other intraocular pressure-lowering drugs include apraclonidine and beta-blockers.
- Other treatment options have been variously tried:

    - Dilation of the artery has been attempted (eg, sublingual isosorbide dinitrate) but these attempts also produce systemic vasodilation with limited positive visual outcome.[4]
    - Oral pentoxifylline, for example, showed improved retinal perfusion but no improvement in vision.
    - Intra-arterial fibrinolysis: local injection of urokinase or recombinant tissue plasminogen activator into the proximal part of the ophthalmic artery is a possibility and a study has shown good results when given within six hours of symptom onset.[7] However, systemic reviews have found insufficient evidence to support the routine use of Intra-arterial thrombolysis to treat CRAO.[8]
    - Another technique which may hold some hope in the future is the technique of enhanced external counterpulsation (EECP). This is a non-invasive technique reported to help relieve angina and improve exercise tolerance. Pneumatic cuffs on the lower extremities are inflated sequentially at the onset of diastole, causing aortic counter pulsation and increased venous return. It has been suggested that this may have a role in other ischaemic disease, including CRAO.[4]
- If giant cell arteritis is strongly suspected, this also needs immediate treatment (intravenous steroids followed by oral steroids), as it is in itself an ophthalmological emergency.
- Long-term management aims are:
    - To identify and address the underlying cause in order to prevent further ischaemic events (eg, investigate and treat hypertension).[9]
    - To reduce risk factors for atherosclerosis, and low-dose aspirin may be beneficial.[4]
- A carotid endartectomy may be necessary depending on the degree of carotid occlusion and local policy.[2]
- Follow-up in the eye department will be necessary to assess for complications, the main one being iris neovascularisation, which itself can cause problems.

Outcome

- Treatment must be started very soon after the onset of symptoms but, even with early treatment, the prognosis is generally very poor, as the inner neural layer becomes atrophic and all useful vision is generally lost.
- Only about a third of patients show any improvement.[1] Spontaneous resolution of the blockage has been reported in 1-8% (some have reported up to 15%) of patients.[4] These patients are monitored in the outpatient clinic for evidence of neovascularisation which can give rise to a different set of problems.
- Patients with retinal emboli have a 3 x increased mortality rate compared to those without.[3] They should be managed in the same way as patients with a personal history of ischaemic heart disease.

Branch retinal artery occlusion

Description

BRAO: one or multiple branches of the central retinal artery may be affected.

Presentation

As for CRAO but only part of the vision is lost: the defect is usually altitudinal or sectoral. The retinal pallor corresponds to the area supplied by the affected artery. Occasionally, an embolus (cholesterol, fibrinoplatelet, calcific) can be seen within the artery and cotton wool spots may eventually occur around that area.

Management

- Refer within 24 hours. However, there is no proven effective management.
- Full and thorough assessment of the cardiovascular system. Carotid endarterectomy or anticoagulation may be indicated.
- Thorough assessment for coagulopathies should also be performed if no embolic source is found.

Outcome

- Generally, the outcome is reasonable with most patients achieving 6/12 or greater vision.[2]
- Occasionally, recanalisation of the artery occurs but this is rare. A quarter of eyes have collateral arteries serving the macular retina so that central vision can be retained to some extent.

Cilioretinal artery occlusion

Ciliary arteries are branches of the ophthalmic arteries that supply the posterior portion of the globe, known as the posterior pole. In up to 30% of the population, there may be an occlusion in a branch of this group of arteries.[1] It may be:

- Isolated - young individuals, associated with vasculitides but has a good prognosis.
- Combined with a central retinal vein occlusion - usually young individuals, and the prognosis is reasonable.
- Associated with arteritic ischaemic optic neuropathy - elderly patients, may be associated with giant cell arteritis, and the prognosis is poor.