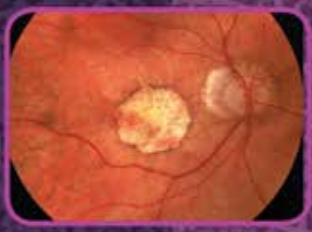
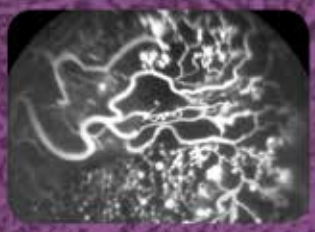


HANDBOOK OF RETINAL DISEASE

A CASE-BASED APPROACH

SECOND EDITION

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Contents

Section 9: Neoplasms and focal lesions

Section 10: Trauma

Section 11: Surgical complications

4 Arterial occlusive disease

How to approach a patient with vision loss and an acute occlusion of a retinal artery

Identify the primary pathologic clinical finding(s)

On examination, the patient will demonstrate arterial occlusion with an area of retinal ischemia/whitening, and if acute, there may be the presence of a cherry-spot in the center of the macula.

Formulate a differential diagnosis

Most likely	Less likely	Least likely
• Central or branch retinal arterial obstruction	• Commotio retinae, infectious (herpetic) retinitis, ocular ischemic syndrome (OIS)	• Tay–Sachy's disease, Niemann–Pick's disease

Query patient history

- How long has the patient been experiencing symptoms?
- Any previous episodes of amaurosis fugax? Pain?
- Any history of trauma?
- Any previous history of stroke, atrial fibrillation, or carotid disease?
- Is the patient immunocompromised?

Decide on ancillary diagnostic imaging

The diagnosis of an arterial obstruction is clinically apparent acutely when there is diffuse retinal whitening secondary to ischemia in the territory of a retinal artery accompanied by abrupt, painless vision loss. Fluorescein angiography can be helpful for confirmation or in nonacute cases.

Ancillary diagnostic imaging interpretation

Color fundus photographs

Color fundus photographs may demonstrate the presence of a cherry-red spot within the macula as well as retinal whitening and ischemia (**Figure 4.1**).

Optical coherence tomography

Retinal thinning will be visible on optical coherence tomography in chronic cases with loss of the inner retina in areas of previous ischemia. Acutely, there may be diffuse retinal swelling in the region of ischemia (**Figure 4.2**). However, there can also be isolated retinal whitening at the level of the inner nuclear layer. This subset of retinal ischemia is termed paracentral acute middle maculopathy (PAMM) based on ischemia of the inner retinal plexus (**Figure 4.3**).

Fluorescein angiography

There is evidence of delayed arterial filling, often with a leading edge of dye in the region of the obstructed arteriole (**Figure 4.4**). Fluorescein angiography may also highlight emboli.

Figure 4.1 Color mosaic photo of central retinal artery occlusion in the left eye. There is the classic cherry-red spot centrally with retinal whitening diffusely involving the macula. There is also a splinter hemorrhage in the peripapillary region.

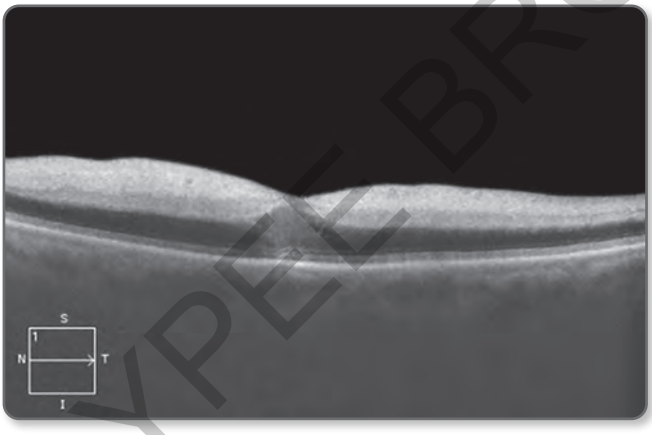

Figure 4.2 Optical coherence tomography in new onset central retinal artery occlusion. Note the thickening of the inner retina consistent with swelling from acute infarction.

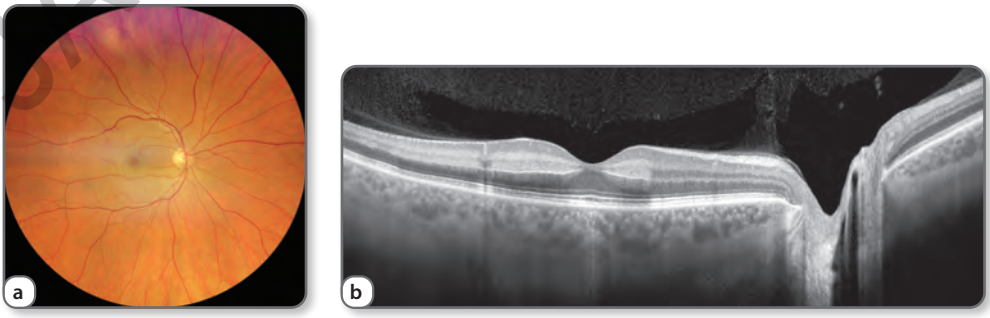

Figure 4.3 Color photograph (a) of the right eye showing deep central retinal whitening and correlating optical coherence tomography (b) showing inner retinal whitening consistent with paracentral acute middle maculopathy.

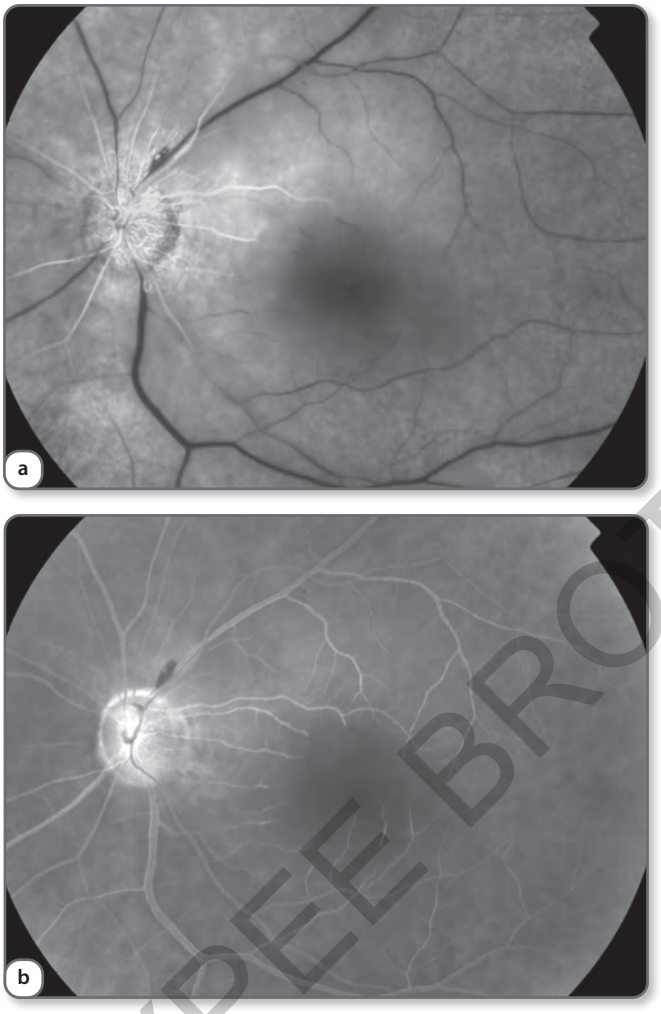

Figure 4.4 Fluorescein angiogram of a central retinal artery occlusion in the left eye at 24 seconds (a) and 300 seconds (b). Note the leading edge of dye in the arteriole, which never completely fills.

Final diagnosis: retinal arteriole occlusion

Epidemiology/etiology

Central retinal artery obstruction (CRAO) is rare, with the average patient age of 60 years. Men are affected twice as commonly as women. A central retinal arterial obstruction is most commonly due to thrombus formation at the region of the lamina cribrosa, the cause of which is most often related to atherosclerosis. Emboli are possible causes, but less common. Additional risk factors include hypercoagulable states, trauma, low-flow states from more proximal disease, optic disc drusen, active ocular infection, congenital abnormalities of the central retinal artery, and migraine. The presence of detectable emboli is visible in only 25% of patients.

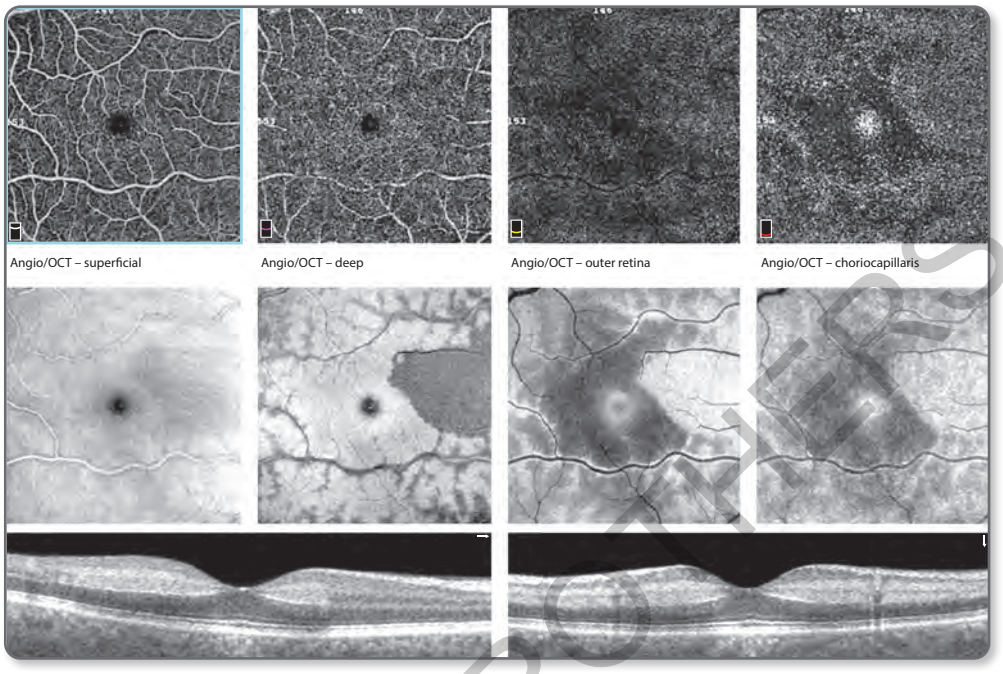

Figure 4.5 Optical coherence tomography angiography (OCTA) showing a fern like pattern highlighting the deep capillary plexus ischemia. CRAO- with OCT on bottom line showing PAMM.

Branch retinal artery obstruction (BRAO) more commonly affects the right eye and is more commonly related to emboli (two-thirds of cases). Typically the emboli are visible at the arterial bifurcation, and originate from anywhere between the heart and ophthalmic artery. Risk factors include hypertension, hypercholesterolemia, diabetes mellitus, smoking, and a positive family history.

Symptoms and clinical findings

Patients with central artery occlusion present with painless, severe vision loss. Visual acuity is typically 20/800 or worse unless a cilioretinal artery is present and not involved. There is typically an afferent pupillary defect. On fundoscopy, there is retinal whitening indicative of acute ischemia. There may be the classic 'cherry-red' spot in the central macula that occurs because retinal ischemia causes blockage of the normal choroidal coloration everywhere except centrally where the nerve fiber layer is thin. Optical coherence tomography angiography (OCTA) can highlight the level at which retinal capillary ischemia occurred by displaying a fern like pattern in areas of capillary dropout (**Figure 4.5**).

Associated features in central artery occlusion include previous episodes of amaurosis fugax, visible embolus in 25%, carotid artery disease in 33%, giant cell arteritis in 5%, neovascularization of the iris in 18%, and arterial collaterals on the optic disc.

Patients with branch retinal artery occlusion may present with sectoral visual field loss typically with preservation of central vision unless the cilioretinal artery is involved.

Treatment/prognosis/follow-up

There is no proven treatment for central retinal artery occlusion. If central retinal artery obstruction is secondary to giant cell arteritis then treatment with high-dose corticosteroids is initiated to prevent contralateral vision loss.

Unfortunately, most central retinal arterial obstructions result in profound, permanent loss of vision. About one-third of patients will experience some improvement in visual acuity. The patient should be worked up medically for causes of acute stroke and often these patients are admitted to the hospital for stroke evaluation. These patients can develop neovascularization of the retina, optic disc, and iris with subsequent neovascular glaucoma. Neovascularization is treated with the application of panretinal photocoagulation.

Further reading

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HANDBOOK OF RETINAL DISEASE
A CASE-BASED APPROACH

The 2nd edition of *Handbook of Retinal Disease: A Case-Based Approach* offers a high-yield case-based model of a wide range of retinal presentations, from patients' signs and symptoms to analyses of diagnostic results and comprehensive outlines of treatment plans.

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- Guides the ophthalmologist through the selection of diagnostic imaging techniques and interpretation of images
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ISBN 978-1-78779-170-1