

INTRODUCTION

The arterial circulation to the eye involves many branches. Any one of these branches may have impaired flow that results from an obstruction. An embolus can occlude the vessel or lead to thrombus formation. Vasculitis may also lead to an occlusion or thrombus formation. In general, an obstruction of either an ophthalmic or retinal arteriole requires a careful evaluation for systemic diseases. Numerous studies have reported an association between symptomatic retinal artery occlusion (RAO) and stroke.⁴⁻¹³ Since the retina and brain share an embryologic origin, the retina is considered part of the central nervous system and is actually brain tissue. Treatment of a retinal ischemic event should therefore be approached the same way as a brain infarction. In 2013, the American Heart Association/American Stroke Association updated their definition of ischemic stroke to include retinal cell death attributable to ischemia in their definition of central nervous system infarction. Central nervous system infarction is brain, spinal cord, or retinal cell death attributable to ischemia, based on (1) pathological, imaging, or other objective evidence of cerebral, spinal cord, or retinal focal ischemic injury in a defined vascular distribution; or (2) clinical evidence of cerebral, spinal cord, or retinal focal ischemic injury based on symptoms persisting 24 hours or more or until death.¹⁴

DISEASE DEFINITION

An ophthalmic artery occlusion (OAO) is a partial or complete obstruction of the ophthalmic artery (branch of the internal carotid artery) and may lead to severe ischemia of the affected globe and associated ocular tissues. A central retinal artery occlusion (CRAO) is a partial or complete obstruction of the central retinal artery, after it branches off from the ophthalmic artery. A branch retinal artery occlusion (BRAO) is a partial or complete obstruction of any of the branch tributaries of the central retinal artery.¹⁵ Arteritic retinal arterial occlusion is caused by giant cell arteritis (GCA), whereas most nonarteritic RAOs are embolic in origin.

A CRAO is a rare condition that has an annual incidence of approximately 1.33 per 100,000 in an Olmsted County, Minnesota population¹⁶ or 1.8 per 100,000 person-years in an entire Korean population over age 65.¹⁷ The incidence likely increases with age,^{17, 18} and the mean age at presentation in the United States is the early 60s.¹⁸ Central retinal artery occlusions have also rarely been documented in children.¹⁹ A CRAO commonly leads to retinal ischemia and subsequent retinal cell death.

CLINICAL FINDINGS CHARACTERISTIC OF RETINAL AND OPHTHALMIC ARTERY OCCLUSION

Patients presenting with a CRAO typically describe a sudden, painless decrease in the visual acuity and field of vision in one eye that occurs over a period of seconds.¹⁸ In 1% to 2% of cases, occurrence is bilateral, which should raise the suspicion of a vasculitic process, such as GCA.²⁰ The presenting visual acuity may vary, but most patients have severe vision loss unless they have cilioretinal sparing, and the patient may or may not have readily visible fundus abnormalities. A deep retinal whitening (i.e., from paracentral acute middle maculopathy [PAMM]) with an inner nuclear layer of hyper-reflectivity on optical coherence tomography (OCT) may be noted initially before progression to a more complete CRAO.²¹⁻²⁴ The classic appearance of retinal whitening and a cherry red spot may be absent or very subtle in the early stages, depending largely on the timing of the examination relative to the onset of obstruction. The classic appearance of retinal whitening with the associated cherry red spot on the foveal center typically presents within several hours of onset. (See Figure 1.) Retinal whitening is caused by opacification of the swollen, ischemic parafoveal nerve fiber layers. The cherry red spot is seen because the foveal center, free of nerve fiber layers, transmits the normal coloration of the underlying, unaffected choroid. Hence, the normally perfused tissues appear red against the ischemic background of the nerve fiber layer.