Chapter 10

Chronic Ocular Ischemia and Carotid Vascular Disease

John E. Carter

Although most ocular manifestations of extracranial cerebrovascular disease have a strokelike temporal profile, some patients have ocular abnormalities that are most compatible with a chronic state of ocular ischemia and have evidence of severe, usually bilateral, extracranial cerebrovascular occlusive disease. Similar ocular findings may be seen in other conditions that produce chronic retinal ischemia or altered retinal blood flow, but in some situations it is difficult to dismiss the associated cerebrovascular occlusive disease as an incidental finding.

Although they represent two ends of a spectrum of pathology secondary to chronic ischemia, the ocular findings in this setting can be separated into a hemorrhagic retinopathy, which usually is referred to as venous stasis retinopathy, and a more diffuse ocular condition termed ischemic oculopathy (1,2). In the neurology literature, the term venous stasis retinopathy usually is reserved for this specific setting. In the ophthalmology literature, however, some confusion exists because the term venous stasis retinopathy has been used to describe other conditions with a similar retinopathy. No generally acceptable term specific to the findings associated with carotid vascular disease has been suggested. Discussions of patients with cerebrovascular disease in the ophthalmology literature generally use the term “venous stasis retinopathy of carotid occlusive disease”. For the purposes of this review venous stasis retinopathy refers to the retinal changes described in association with carotid atherosclerotic occlusive disease.

Clinical Findings and Natural History of Venous Stasis Retinopathy and Ischemic Oculopathy

Venous stasis retinopathy, although not referred to as such, was recognized as a manifestation of aortic arch syndromes (3–5) for some years before 1963 when Hedges (6) and Kearns and Hollenhorst (7) described its association with carotid occlusive disease. The earliest changes in venous
stasis retinopathy consist of microaneurysms and small dot-and-blout hemorrhages. These findings are most prominent beginning at the midperiphery of the fundus. More severe ischemia produces dilatation and darkening of the retinal veins, often with marked irregularity of the caliber of the major retinal veins. Hemorrhages in the nerve fiber layer may occur. Swelling of the optic disk is unusual but may be present to mild degree, as is sometimes seen with severe diabetic retinopathy. Fig. 10–1 illustrates these changes. If the ischemia persists, neovascularization of the retina and optic disk may develop (Fig. 10–2). More severe ischemia may produce pallor or a gray cast to the macula that is due to retinal edema. Although visual impairment and retinal edema always should coexist, the remainder of the signs of venous stasis retinopathy may be present in patients who have normal visual function. Venous stasis retinopathy may be seen in patients who recently have had a stroke (6,7), who have ongoing transient monocular or cerebral ischemic attacks (7–9), who have had known cerebrovascular disease in the past but have no recent symptoms (7), or who have symptoms limited to mild visual loss (9,10). Occasionally, the finding is incidental (11).

In ischemic oculopathy (1,2) additional changes take place in the anterior segment of the eye if the ischemic state continues. This condition also has been termed ischemic ophthalmitis, ischemic ophthalmitis, and ischemic ocular inflammation. These findings also were recognized earlier in aortic arch syndromes (4, 5). Smith (12) first called attention to the association of neovascular glaucoma and occlusion of the ICA. The spectrum of changes seen in ischemic oculopathy subsequently were described in detail by Hoefnagels (13) and Knox (14). The abnormalities include episcleral vascular congestion, a cloudy cornea, cells and flare in the anterior chamber, and neovascularization of the iris (Fig. 10–3). The pupil often is middilated and reacts sluggishly or not at all to either direct or consensual stimulation. Intraocular pressure usually is elevated but may be normal or even decreased. In most cases venous stasis retinopathy continues to be present, but the retinal arteries and veins may become markedly attenuated, with visibly sluggish blood flow through the veins. Optic atrophy may develop.

Patients with venous stasis retinopathy or ischemic oculopathy invariably have a marked reduction of the central retinal artery pressure on ophthalmodynamometry. Often pressure is so low that touching the eye produces pulsation in the central retinal artery so that pressure cannot be measured. In some cases diastolic pressure in the central retinal artery is so low that spontaneous pulsations are present (7,13,15–17).

Ischemic pain is a feature in some patients with venous stasis retinopathy and ischemic oculopathy. It is characterized as a constant aching over the orbit, upper face, and temple and may be worse when the patient is in an upright position (16,18). This pain is not related to glaucoma, which also may be a source of pain in patients with ischemic oculopathy.