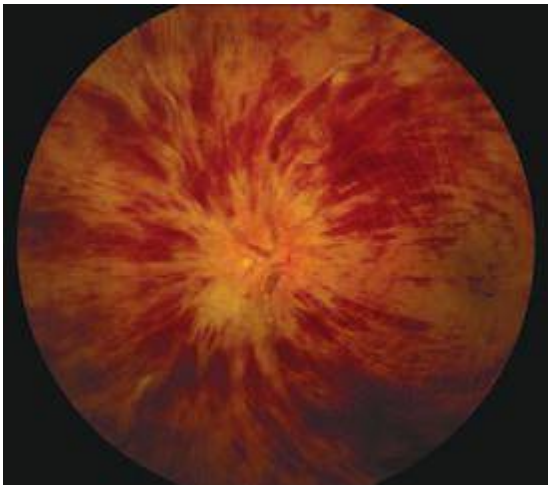

FIGURE 39-8 Central retinal vein occlusion can produce massive retinal hemorrhage ("blood and thunder"), ischemia, and vision loss.

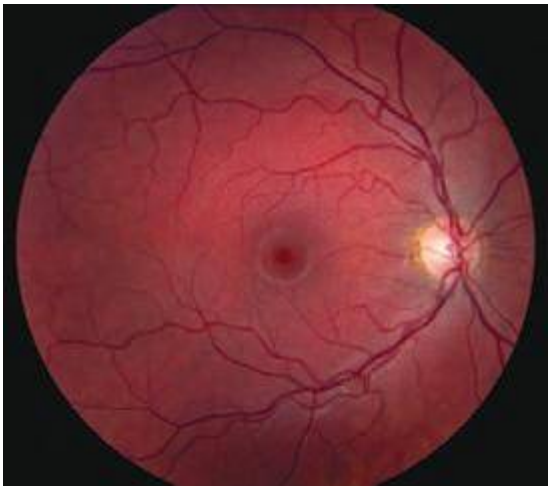

FIGURE 39-10 Retrobulbar optic neuritis is characterized by a normal fundus examination initially, hence the rubric "the doctor sees nothing, and the patient sees nothing." Optic atrophy develops after severe or repeated attacks.

are prominent risk factors. Polycythemia, thrombocythemia, or other factors leading to an underlying hypercoagulable state should be corrected; aspirin treatment may be beneficial.

Anterior Ischemic Optic Neuropathy (AION) This is caused by insufficient blood flow through the posterior ciliary arteries that supply the optic disc. It produces painless monocular visual loss that is sudden in onset, followed sometimes by stuttering progression. The optic disc appears swollen and surrounded by nerve fiber layer splinter hemorrhages (Fig. 39-9). AION is divided into two forms: arteritic and nonarteritic. The nonarteritic form is most common. No specific cause can be identified, although diabetes and hypertension are common risk factors. A crowded disc architecture and small optic cup predispose to the development of nonarteritic AION. No treatment is available. About 5% of patients, especially those age >60, develop the arteritic form of AION in conjunction with giant-cell (temporal) arteritis (Chap. 385). It is urgent to recognize arteritic AION so that high doses of glucocorticoids can be instituted immediately to prevent blindness in the second eye. Symptoms of polymyalgia rheumatica may be present; the sedimentation rate and C-reactive protein level are usually elevated. In a patient with visual loss from suspected arteritic AION, temporal artery biopsy is mandatory to confirm the diagnosis. Glucocorticoids should be started immediately, without waiting for the biopsy to be completed. The diagnosis of arteritic AION is difficult to sustain in the face of a negative temporal artery biopsy, but such cases do occur rarely. It is important to biopsy an arterial segment of at least 3 cm and to examine a sufficient number of tissue sections prepared from the specimen.

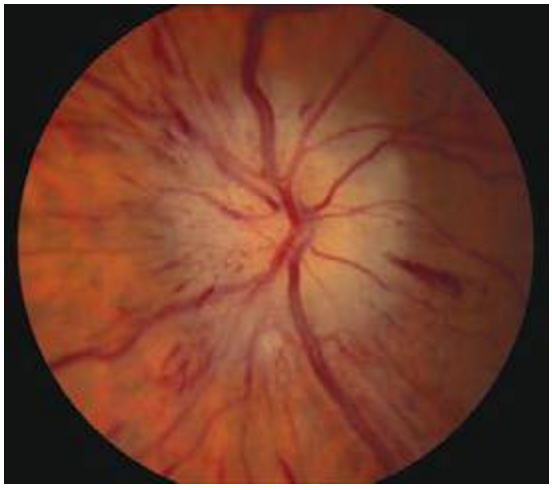

FIGURE 39-9 Anterior ischemic optic neuropathy from temporal arteritis in a 67-year-old woman with acute disc swelling, splinter hemorrhages, visual loss, and an erythrocyte sedimentation rate of 70 mm/h.

Posterior Ischemic Optic Neuropathy This is an uncommon cause of acute visual loss, induced by the combination of severe anemia and hypotension. Cases have been reported after major blood loss during surgery (especially in patients undergoing cardiac or lumbar spine operations), exsanguinating trauma, gastrointestinal bleeding, and renal dialysis. The fundus usually appears normal, although optic disc swelling develops if the process extends anteriorly far enough to reach the globe. Vision can be salvaged in some patients by prompt blood transfusion and reversal of hypotension.

Optic Neuritis This is a common inflammatory disease of the optic nerve. In the Optic Neuritis Treatment Trial (ONTT), the mean age of patients was 32 years, 77% were female, 92% had ocular pain (especially with eye movements), and 35% had optic disc swelling. In most patients, the demyelinating event was retrobulbar and the ocular fundus appeared normal on initial examination (Fig. 39-10), although optic disc pallor slowly developed over subsequent months.

Virtually all patients experience a gradual recovery of vision after a single episode of optic neuritis, even without treatment. This rule is so reliable that failure of vision to improve after a first attack of optic neuritis casts doubt on the original diagnosis. Treatment with high-dose IV methylprednisolone (250 mg every 6 h for 3 days) followed by oral prednisone (1 mg/kg per day for 11 days) makes no difference in ultimate acuity 6 months after the attack, but the recovery of visual function occurs more rapidly. Therefore, when visual loss is severe (worse than 20/100), IV followed by PO glucocorticoids are often recommended.

For some patients, optic neuritis remains an isolated event. However, the ONTT showed that the 15-year cumulative probability of developing clinically definite multiple sclerosis after optic neuritis is 50%. A brain magnetic resonance (MR) scan is advisable in every patient with a first attack of optic neuritis. If two or more plaques are present on initial imaging, treatment should be considered to prevent the development of additional demyelinating lesions (Chap. 458).

LEBER'S HEREDITARY OPTIC NEUROPATHY

This disease usually affects young men, causing gradual, painless, severe central visual loss in one eye, followed weeks to years later by the same process in the other eye. Acutely, the optic disc appears mildly plethoric with surface capillary telangiectasias but no vascular leakage on fluorescein angiography. Eventually optic atrophy ensues. Leber's optic neuropathy is caused by a point mutation at codon 11778 in the mitochondrial gene encoding nicotinamide adenine dinucleotide