factors and must be ruled out. Essential thrombocytosis should be included in the differential diagnosis of an avascular retina in otherwise healthy persons.

REFERENCES

1. Abe S, Shiono T. Retinochoroidal circulatory disturbances and blood component abnormalities. Nippon Ganka Gakkai Zasshi 1995;99:255–261.
2. Galetta SL, Plock GL, Kushner MJ, Wyszynski RE, Brucker AJ. Ocular thrombosis associated with antiphospholipid antibodies. Ann Ophthalmol 1991;23:207–212.
3. van Nouhuys CE. Signs, complications, and platelet aggregation in familial exudative vitreoretinopathy. Am J Ophthalmol 1991;111:34–41.
4. Goldberg MF, Custis PH. Retinal and other manifestation of incontinentia pigmenti. Ophthalmology 1993;100: 1645–1654.
5. Greven CM, Wall AB. Peripheral retinal neovascularization and retinal vascular occlusion associated with activated protein C resistance. Am J Ophthalmol 1997;124: 687–689.

Bilateral Ocular Ischemic Syndrome Secondary to Giant Cell Arteritis Progressing Despite Corticosteroid Treatment

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PURPOSE: To report the development of a bilateral ocular ischemic syndrome despite corticosteroid treatment in a patient with giant cell arteritis.

METHOD: Case report.

RESULTS: Despite receiving high-dose intravenous methylprednisolone and oral prednisone for biopsy-proven giant cell arteritis that presented as a severe anterior ischemic optic neuropathy in the right eye, a patient developed progressive ocular ischemia in that eye as well as an ocular ischemic syndrome in the fellow eye.

CONCLUSIONS: Some patients with giant cell arteritis, possibly patients with other underlying systemic vasculopathies, are refractory to what should be adequate treatment with systemic corticosteroids and may develop a bilateral ocular ischemic syndrome. (Am J Ophthalmol 1999;127:102–104. © 1999 by Elsevier Science Inc. All rights reserved.)

Accepted for publication July 8, 1998.

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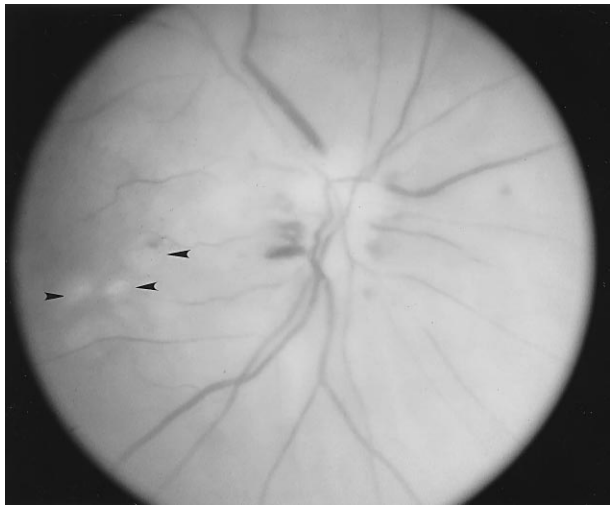

FIGURE 1. Right ocular fundus 2 days after onset of visual loss. Note pallid swelling of the optic disk, which is surrounded by several flame-shaped hemorrhages. Also note cotton-wool spots in posterior pole (arrowheads). The photograph is somewhat blurred because of mild corneal edema.

GIANT CELL ARTERITIS IS A SYSTEMIC VASCULITIS AFfecting large and medium-sized arteries and can produce a variety of ocular manifestations, including anterior and posterior ischemic optic neuropathy, central retinal artery occlusion, and the ocular ischemic syndrome.[1–3] We report a patient who developed a progressive bilateral ocular ischemic syndrome despite early treatment with high-dose corticosteroids.

A 76-year-old woman with a history of age-related macular degeneration, migraine headaches, pernicious anemia, hypothyroidism, osteoporosis, and hypertension developed jaw claudication, double vision, and increasingly severe headaches. Two weeks later, she experienced sudden complete loss of vision in the right eye.

On examination 2 days after the onset of visual loss, visual acuity was no light perception in the right eye. Best-corrected visual acuity was 20/30 in the left eye, and the patient could identify correctly only one of 10 Hardy-Rand-Rittler pseudoisochromatic color plates. There was a right relative afferent pupillary defect. Slit-lamp biomicroscopy disclosed folds of Descemet membrane in the right eye and mild anterior chamber flare in both eyes. Intraocular pressures were RE, 3 mm Hg and LE, 8 mm Hg. Ocular motor examination showed diminished abduction with right eye and an inability to adduct the left eye past the midline. The right optic disk was moderately swollen, several flame-shaped hemorrhages surrounded the disk, and there were cotton-wool spots in the posterior pole (Figure 1). The left optic disk appeared normal. The patient's hematocrit was 36. Westergren erythrocyte sedimentation rate was 67 mm/h.

The patient was hospitalized and treated with intravenous methylprednisolone, 250 mg every 6 hours for 3 days.

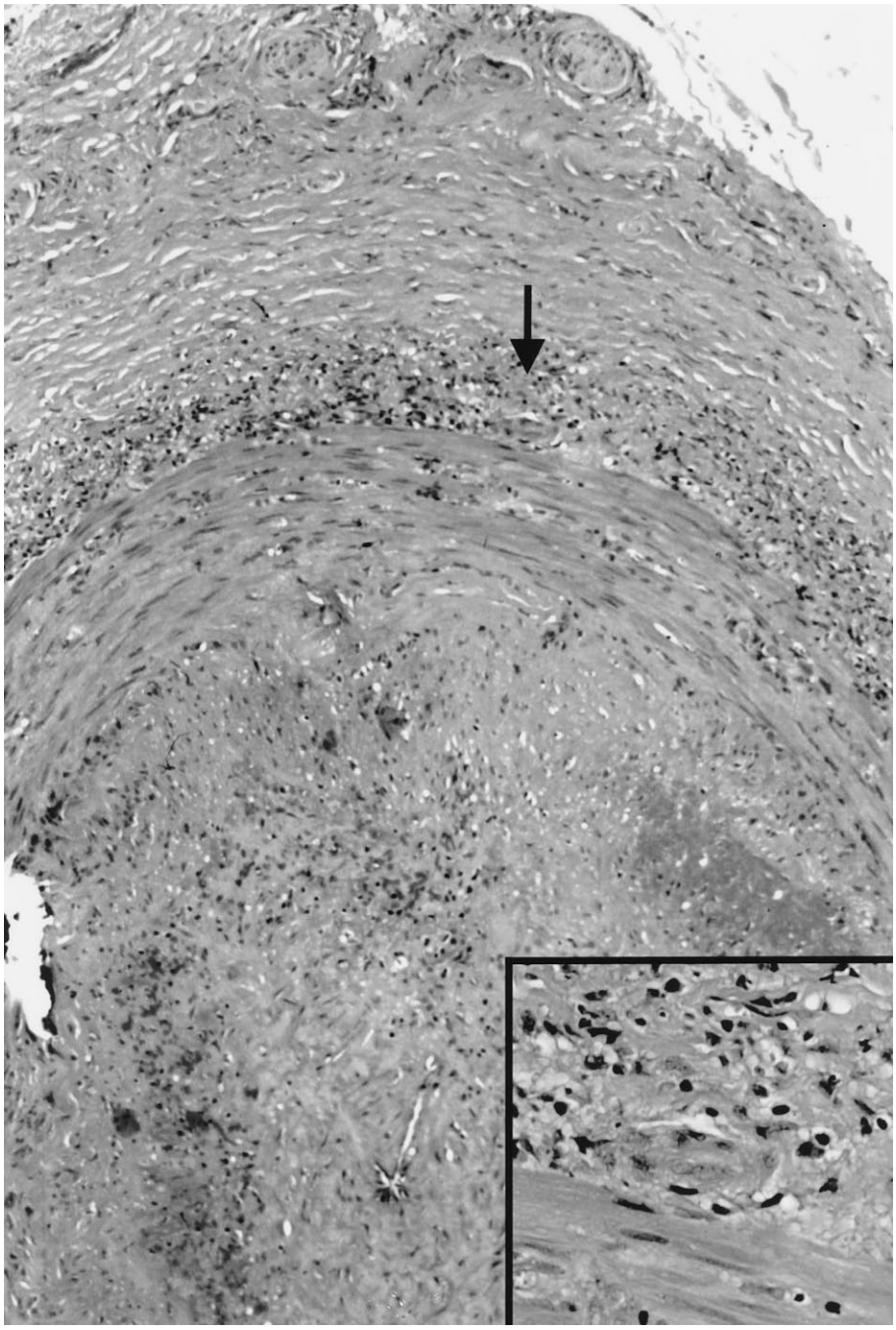

FIGURE 2. Histologic appearance of temporal artery biopsy specimen. There is extensive inflammation throughout the wall of the artery. The arrow points to a giant cell, which is seen at higher power in the inset.

During this time, a temporal artery biopsy confirmed the diagnosis of giant cell arteritis (Figure 2). Carotid Doppler imaging revealed severe stenosis of the right internal carotid artery and mild stenosis of the left internal carotid artery. There was no flow in the right ophthalmic artery, whereas flow was normal in the left ophthalmic artery. The patient's erythrocyte sedimentation rate decreased to 4 mm/h during her hospitalization. She was subsequently discharged on 60 mg/day of oral prednisone.

Seven days following discharge, the patient developed blurred vision in her left eye. According to both the patient and her husband, she had been taking the oral prednisone as directed. Nevertheless, the examination revealed best-corrected visual acuity of RE, no light perception and LE, 20/160. There was mild edema of the left cornea, and intraocular pressures were 6 mm Hg in both eyes. The right optic disk was swollen; the left optic disk appeared normal. The following day, there was mild right

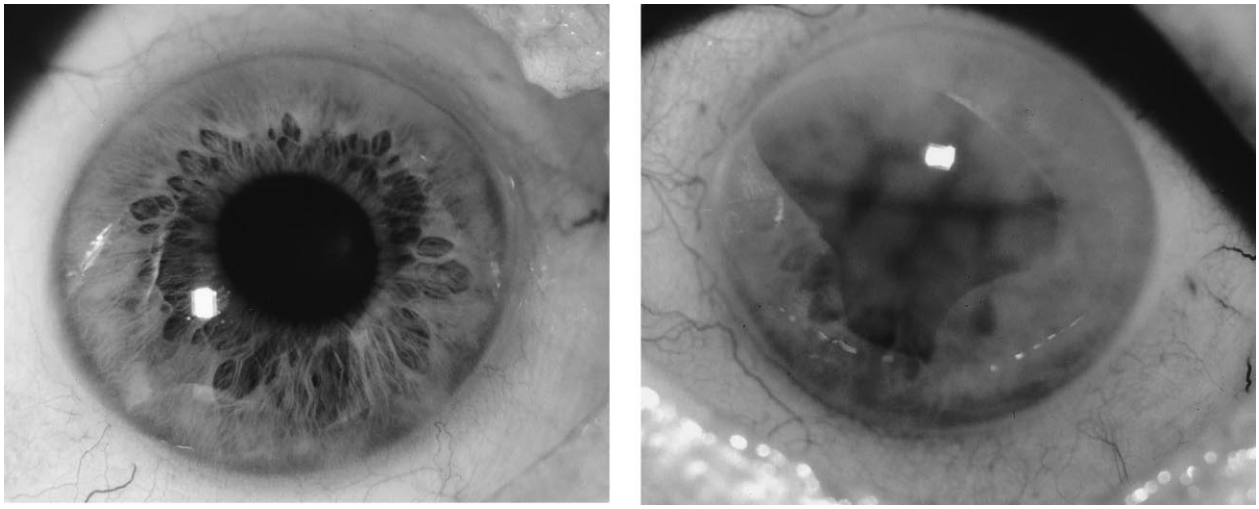

FIGURE 3. External appearance of the eyes after initial treatment with high-dose intravenous corticosteroid therapy and while taking oral prednisone. (Left) The right cornea is relatively clear, but the right pupil is ovoid and slightly displaced. There is mild flare in the anterior chamber. (Right) The left cornea shows severe edema, and the left pupil is markedly distorted. Note that the conjunctival vessels of the left eye are dilated compared with those of the right eye.

anterior chamber flare and cell in the right eye, and the right pupil was distorted (Figure 3). The left cornea showed severe edema that prevented visualization of the ocular fundus. Intraocular pressures were RE, 3 mm Hg and LE, 1 mm Hg. Westergren erythrocyte sedimentation rate was 7 mm/h. Color Doppler sonography showed no blood flow in either ophthalmic artery. Despite retreatment with intravenous methylprednisolone in a dose of 250 mg every 6 hours, as well as heparin, the patient's vision rapidly declined to no light perception in the left eye.

The ocular ischemic syndrome is an uncommon disorder characterized by hypotony, uveitis, corneal edema, and visual loss caused by decreased vascular perfusion of the eye and orbit. This syndrome is occasionally reported in association with giant cell arteritis, but we are aware of only one other report of a patient with a bilateral ocular ischemic syndrome associated with giant cell arteritis.[4] That patient's visual acuity improved to 20/50 in one eye following treatment with systemic corticosteroids. To the best of our knowledge, our case is the first reported example of a progressive bilateral ocular ischemic syndrome in giant cell arteritis resulting in no light perception in both eyes despite high-dose intravenous corticosteroid treatment, and we therefore question the role played by the patient's systemic hypertension and pernicious anemia in the pathogenesis of the process.

Early diagnosis of giant cell arteritis and immediate treatment with intravenous corticosteroids may not only prevent further damage to the affected eye but also prevent visual loss in the opposite eye[5]; however, some patients with giant cell arteritis appear to be refractory to conventional corticosteroid treatment. Such patients, perhaps those with other underlying systemic vasculopathies, may develop a progressive bilateral ocular ischemic syndrome.

REFERENCES

1. Ghanchi FD, Dutton GN. Current concepts in giant cell (temporal) arteritis. Surv Ophthalmol 1997;42:99–123.
2. Galetta SL. Vasculitis. In: Miller NR, Newman NJ, editors. Walsh and Hoyt's clinical neuro-ophthalmology, 5th Edition, Volume 3. Baltimore: Williams & Wilkins, 1998:3755–3782.
3. Hayreh SS, Podhajsky PA, Zimmerman B. Am J Ophthalmol 1998;125:509–520.
4. Hamed LM, Guy JR, Moster ML, Bosley T. Giant cell arteritis in the ocular ischemic syndrome. Am J Ophthalmol 1992;113: 702–705.
5. Liu GT, Glaser JS, Schatz NJ, Smith JL. Visual morbidity in giant cell arteritis: clinical characteristics and prognosis for giant cell arteritis. Ophthalmology 1994;101:1779–1785.

Anterior Ischemic Optic Neuropathy Following the Use of a Nasal Decongestant

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PURPOSE: To report a case of sequential anterior ischemic optic neuropathy temporally related to the sequential use of a decongestant nasal spray.

Accepted for publication July 7, 1998.

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This work was supported in part by an unrestricted grant from Research to Prevent Blindness, Inc, New York, New York (Ophthalmology) and National Institutes of Health core grant P30-EY0 6360 (Ophthalmology).

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