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.)

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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 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.
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. 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; 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


Anterior Ischemic Optic Neuropathy Following the Use of a Nasal Decongestant

George D. Fivgas, MD, and Nancy J. Newman, MD

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