Letters

Optic Neuropathy Secondary to Spontaneous Intracranial Hypotension (SIH) as Related to Experimental Primate Model

We read with great interest the recent article by Yang et al.1 regarding their primate model investigating optic neuropathy resulting from reduced cerebrospinal fluid pressure. The authors conducted a thorough case-control study looking at the effects of intracranial hypotension from insertion of a lumbar-peritoneal shunt in four rhesus monkeys, adding five control monkeys with lumbar-peritoneal shunt inserted but immediately occluded. This is an innovative study, giving some vital information about the structural damage to the optic nerve that can ensue from decreased intracranial pressures.

This study is of particular interest to us given a clinical case we recently encountered. In brief, a 74-year-old man developed slowly progressive vision loss over 9 to 12 months. He had a history of nonpostural headache. On examination, the visual acuities (VA) were 20/125, right eye and 20/20, left eye. The remainder of his examination was significant for a 2+ right relative afferent pupillary defect, right eye dyschromatopsia, and obvious optic atrophy in the right eye with 3+ pallor. Visual field testing showed a temporal defect respecting the vertical meridian in the right eye, and superior and inferior constriction in the left eye (Figs. 1A, 1B). Magnetic resonance imaging (MRI) revealed diffuse dural thickening and enhancement in the vicinity of the optic canals bilaterally (Fig. 2A). A lumbar puncture demonstrated an opening pressure of 80 mm H2O and an elevated cerebrospinal fluid (CSF) protein of 88 mg/dL. Cytology was negative.

Five days after the lumbar puncture, the patient noted a slowly progressive decline in vision, now also involving the left eye. The visual acuities were 9/200 in the right eye and 20/30 in the left eye. Visual field testing showed worsening (Figs. 1C, 1D). The patient then underwent a lumbar epidural blood patch procedure. Subsequently, the visual loss ceased and the visual fields remained stable in the right eye, and gradually improved in the left eye, over the next 6 months (Figs. 1E, 1F). Repeat neuroimaging demonstrated significantly improved meningeal thickening and decreased enhancement in the region of the optic canals (Fig. 2B).

Our presumption is that the visual loss was a consequence of intracranial hypotension, supported by the fact that the progressive loss ceased on the right and function was restored on the left once the blood patch was done. Our MRI of this patient showed dural thickening with noticeable overall morphologic improvement after the patch was placed. This also supports spontaneous intracranial hypotension (SIH) as a probable etiology.

Spontaneous intracranial hypotension is an uncommon disorder that is often missed or misdiagnosed due to its variable clinical manifestations, especially in the absence of postural headache.2–8 A case series by Horton and Fishman9 described two patients with visual field loss in the setting of SIH. After blood patch, the perimetry normalized in both subjects. The authors postulated that the visual field defects were likely related to compression or vascular congestion of the intracranial portions of the optic nerve.9

In SIH, the CSF opening pressure is typically low, but a significant minority of the patients with a documented active CSF leak and typical clinical and imaging manifestations of the
disorder have CSF opening pressures that are within normal limits. The opening pressure of 80 mm H₂O in our patient is compatible with the diagnosis of spontaneous CSF leak. Yang et al. found that two of their hypotensive monkeys experienced damage to only one optic nerve over the course of several months, which is similar to our patient with who experienced visual loss in only one eye for several months. Only after the lumbar puncture, which likely caused further reduction in intracranial pressure, did the visual loss accelerate in both eyes.

The authors have conducted a very interesting study on the structural abnormalities that are seen with the induced intracranial hypotension. Our patient showed structural damage in addition to functional decline. Optical coherence tomography imaging demonstrated thinning of the retinal nerve fiber layer temporally in the right eye (mean 82 μm) and borderline nasally in the left eye (mean 91 μm). Three months later, it had worsened in the right (mean 64 μm) and left (mean 85 μm) eyes. This appears to be fairly consistent with the proportional reduction in RNFL measured in the monkey subjects.

Yang et al. may want to consider occluding the shunt to see to what extent the damage that was induced is structurally reversible. In our case, the blood patch halted the damage and induced some functional improvement.

As the authors pointed out, there are many reasons why monkeys and humans may react differently. But based on many primate similarities, it is reasonable to assume these effects may be anticipated in predisposed individuals among our patient populations with intracranial hypotension. It is useful for clinicians to be alerted to this possible etiology of vision loss.

Sylvia L. Groth
Michael S. Lee
Alexander M. McKinney
Bahram Mokri

1Department of Ophthalmology, University of North Carolina, Chapel Hill, Chapel Hill, North Carolina, United States; 2Department of Ophthalmology, University of Minnesota Medical School, Minneapolis, Minnesota, United States; 3Department of Radiology, University of Minnesota Medical School, Minneapolis, Minnesota, United States; and 4Department of Neurology, Mayo Clinic, Rochester, Minnesota, United States.

E-mail: mikelee@umn.edu

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