Dorsal Midbrain Syndrome With Bilateral Superior Oblique Palsy Following Brainstem Hemorrhage

Lesions affecting the dorsal midbrain can result in a constellation of ocular findings such as vertical gaze disturbances, convergence retraction nystagmus, light-near dissociation of pupils, and eyelid retraction.1,2 Although bilateral superior oblique palsy can occur after a stroke, its occurrence secondary to nontraumatic brainstem hemorrhage is extremely rare.3 We report a combination of dorsal midbrain syndrome and bilateral superior oblique palsy following brainstem hemorrhage.

Report of a Case. A 43-year-old man noted sudden onset of binocular vertical and torsional diplopia subsequent to a stroke 2 years prior to presentation. He complained of oscillopsia more pronounced in upgaze along with an anomalous chin-down position since the stroke. He was receiving anticoagulant therapy for coagulopathy at the time of the stroke and underwent a right frontal ventriculoperitoneal shunt for acute hydrocephalus secondary to the intracranial bleed.

Uncorrected visual acuity was 20/25 OU. Pupils were 3 mm in both eyes with a light-near dissociation. A 20° chin-down position was noted in primary gaze. Motility examination showed 30 prism diopter (Δ) esotropia with 4 Δ right hypertropia in primary position, 20 Δ esotropia with 7 Δ right hypertropia in upgaze, and 35 Δ esotropia with 4 Δ right hypertropia in downgaze. In right gaze, there was 20 Δ esotropia with 9 Δ left hypertropia, and in left gaze, there was 20 Δ esotropia with 10 Δ right hypertropia. Right head tilt revealed 20 Δ esotropia with 12 Δ left hypertropia, and left head tilt showed 25 Δ esotropia with 4 Δ left hypertropia. There was reduced depression in adduction and reduced elevation in both abduction and adduction in both eyes. Double Maddox rod test showed 30° excyclotorsion OS in primary gaze increasing to 42° in downgaze. An eyelid retraction and convergence retraction nystagmus was noted in attempted upgaze with hypometric vertical saccades both in upgaze and downgaze (Figure 1).

Sagittal noncontrast magnetic resonance imaging (Figure 2A) performed 3 months prior to ocular examination revealed prior brainstem hemorrhage extending

Figure 2. Magnetic resonance imaging findings of hypertensive brainstem encephalopathy. A, Axial T2-weighted image on initial examination shows abnormal signal and enlargement of the pons consistent with edema. B, Axial T2-weighted image 8 days later, after blood pressure was controlled, shows improvement of abnormal signal and decreased enlargement of the pons.
Figure 1. Preoperative photographs of the subject in 9 diagnostic positions of gazes demonstrating undererelevation in both adduction and abduction and underdepression in adduction in both eyes. A V pattern esotropia along with an alternating hypertropia in side gazes is also seen.

Figure 2. Magnetic resonance imaging (MRI) of the head. A, Sagittal T1 MRI scan without contrast. It shows evidence of prior brainstem hemorrhage (arrowheads) extending vertically from the posterior pontomesencephalic junction into the cerebral peduncle with hemosiderin staining of the superior cerebellar peduncle. B, Axial T1 MRI scan with contrast. Following administration of contrast, there was enhancement along the margins of the hemorrhagic cleft (arrow).
vertically from the posterior pontomesencephalic junction into the cerebral peduncle with hemosiderin staining of the superior cerebellar peduncle. Axial contrast magnetic resonance imaging (Figure 2B) showed enhancement along margins of the hemorrhagic cleft, which was felt to be an underlying occult vascular malformation/sequelae from prior hemorrhage. The patient underwent bilateral superior oblique tucking with bilateral medial rectus recession, which improved torsion in primary gaze, although moderate esotropia and excyclotorsion persisted in downgaze.

Comment. Dorsal midbrain syndrome can be due to a number of conditions such as pineal region neoplasms, obstructive hydrocephalus, arteriovenous malformations, multiple sclerosis, mesencephalic hemorrhages, or dorsal midbrain infection. Spontaneous nontraumatic, nonhypertensive, midbrain hemorrhage is an uncommon cause of dorsal midbrain syndrome and may be due to an underlying occult vascular malformation.

Bilateral superior oblique palsies are usually congenital or the consequence of a closed head trauma; those secondary to a nontraumatic brainstem hemorrhage are extremely rare. Although an isolated case of dorsal midbrain syndrome and bilateral superior oblique palsy has been reported before, the association of the two following a nontraumatic, nonhypertensive brainstem hemorrhage is extremely rare.

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Financial Disclosure: None reported.

In recent years, intravitreal triamcinolone acetone (IVT) injection has emerged as an important therapeutic modality and has been evaluated as treatment of a large range of macular pathologic abnormalities, including cystoid macular edema, choroidal neovascularization, and diabetic macular edema. Even though the functional and anatomical improvement observed is promising, in particular when treating macular edema, the therapeutic efficacy is somewhat shadowed by the limited therapeutic options available for those maculopathies, and considering that in patients with pre-existing glaucoma the peripheral vision is also under serious threat, we believe that the concept of treating glaucomatous eyes with IVT is worth reconsidering. Herein, we report the clinical course of 11 patients with glaucoma who subsequently developed macular disease and were injected with 4 mg of triamcinolone acetonide.

Nine patients had undergone filtering surgery between 2 weeks and 12 years before their first IVT injection, and all blebs and shunts were assessed to be functional at the time of triamcinolone treatment. Follow-up period in this study ranged from 3 to 13 months; visual acuity (VA) changes reported were calculated using the baseline VA and the VA recorded at the last visit. The maximum increase of IOP after administering IVT to eyes with surgically controlled glaucoma was 6 mm Hg; the mean difference between pre-IVT and maximum post-IVT IOP was 1 mm Hg. Two patients required additional medication to achieve control of their IOP (patients 2 and 6 in the Table); 3 others were already receiving topical medications, which were continued following IVT. All but 1 subject showed at least short-term improvement in their visual function as well as anatomical regression of macular lesion. The clinical characteristics of the patients, their responses to IVT, and the measures used to achieve IOP control are summarized in the Table; in addition, the clinical course of 2 patients treated with triamcinolone before and after filtering surgery is described.

Report of Cases. Case 1. An otherwise healthy 96-year-old man (patient 1) with a history of primary open-angle glaucoma controlled with travoprost and dorzolamide complained of recently deteriorating vision in his left eye; his right eye had reportedly had very poor vision for 60 years. Visual acuity was 20/400 OD and 20/60 OS. The diagnosis was central retinal vein occlusion in the left eye; its IOP at the time was 16 mm Hg. Optical coherence tomography revealed cystoid macular edema and a central macular thickness of 376 µm. Three weeks later, his vision deteriorated to 20/200 and macular thickness increased to 548 µm. The decision was made to treat him with IVT. Four weeks later, his vision had improved to 20/80, his macular thickness was 234 µm, and his IOP was elevated at 24 mm Hg. During the following 3 months, his VA remained stable, but his IOP was above normal despite medical treatment, reaching a maximum of 34 mm Hg. Four months after IVT, the cystoid macular edema recurred and the patient’s VA deteriorated to 20/150. Since adequate control of IOP had not been achieved, a second IVT was deemed unacceptable despite favor-
extensive, the location of the drainage device should be explored directly if the restriction needs to be released.

In summary, strabismus after implantation of a glaucoma drainage device is an uncommon but serious complication. Removal of the fibrous capsule surrounding the implant is essential to release restrictions identified on forced-duction testing. However, appropriate rectus extracocular muscle recession with the use of adjustable sutures is required to achieve ocular alignment.

Submitted for Publication: April 19, 2007; final revision received July 26, 2007; accepted August 5, 2007.

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Financial Disclosure: None reported.

Funding/Support: Dr Rosenbaum is a recipient of a Research to Prevent Blindness Physician Scientist Merit Award.

Previous Presentation: This study was presented at the Annual Meeting of the American Association for Pediatric Ophthalmology and Strabismus; April 14, 2007; Seattle, Washington.

REFERENCES


Correction

Error in “Report of a Case” Section. In the Case Report titled “Dorsal Midbrain Syndrome With Bilateral Superior Oblique Palsy Following Brainstem Hemorrhage,” published in the December 2006 issue of the Archives (2006;124(12):1786-1788), on page 1786, third column, line 9, the incorrect published statement reads “Right head tilt revealed 20 Δ esotropia with 12 Δ left hypertropia, and left head tilt showed 25 Δ esotropia with 4 Δ left hypertropia.” The statement should have said that on left head tilt the patient showed 25 Δ esotropia with 4 Δ right hypertropia.