Bilateral aberrant regeneration of the third cranial nerve following trauma

Case report

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This 6-year-old boy, after recovery from prolonged coma following head trauma, exhibited the rare phenomenon of bilateral oculomotor nerve misdirection manifested by ipsilateral eyelid elevation on adduction of either eye.

KEY WORDS □ third nerve aberrant regeneration □ head trauma □ bilateral third nerve injury

Misdirection of regenerating oculomotor nerve fibers is a well-known phenomenon, particularly likely to occur after third cranial nerve damage by carotid aneurysms or trauma. The peculiar eye movements characteristic of aberrant regeneration of the third nerve were described by Gowers in 1879; the currently accepted explanation of misdirection of regenerating peripheral nerve fibers was suggested by Bielschowsky in 1935, and experimentally reproduced by Bender and Fulton in 1938. It has become increasingly apparent that minor degrees of misdirection are common; however, the bilateral aberrant regeneration we are reporting is rare and the associated synkinetic eye movements may be diagnostically perplexing.

Case Report

A previously healthy 6-year-old boy was kicked in the left side of his forehead by a horse and was promptly admitted to a local hospital.

Examination. The patient was comatose, with fixed dilated pupils and decerebrate posturing in response to painful stimuli. A nearly circumferential, horizontal skull fracture was present on x-ray films. Cerebral swelling was diagnosed but an emergency bifrontal decompression craniectomy revealed no confirmatory evidence of swelling.

Course. The patient's condition had changed little 2 weeks later. Painful stimuli evoked decerebrate posturing, pupils were fixed at 8 mm, and there was no eye movement spontaneously or on oculocephalic maneuvers. Caloric testing was not performed. One month after the accident he began to grimace and show purposeful withdrawal from painful stimuli; by 2 months he was able to open his eyes halfway, to say a few words, and walk a few steps with assistance. Four months after the accident, he was oriented, emotionally labile, and walking with an unsteady gait. He had bilateral ptosis, which was greater on the right, fixed 7-mm pupils, and marked exotropia. Although he exhibited good lateral eye movements, up-
ward and medial movements were more limited in the right than in the left eye. Two years after the accident, extraocular muscle surgery on the right eye to decrease the exotropia and right hypertropia resulted in modest cosmetic improvement.

Follow-up Studies. When seen in ophthalmoneurological consultation 2½ years after trauma, the patient was well coordinated and able to participate in sports. There was an impressive decrease in mental function characterized by hyperactivity, a short attention span, poor memory, and slowness in school work. The ophthalmological findings were significant. The visual acuity was 20/20 in the right eye and 20/30 in the left, and color vision was normal in each eye when tested with Hardy-Rand-Rittler plates. Visual fields were full and the funduscopic examination was normal except for mild bilateral optic disc pallor. Exotropia, right hypertropia, and a slight right head tilt were present, and on preferred fixation with the left eye, the right palpebral fissure was smaller than the left (Fig. 1). The pupils were each 7 mm in diameter, fixed to light, and did not constrict with eye movement. The horizontal range of eye movements was full; elevation was mildly limited bilaterally, and downward gaze could not be elicited. No globe movement occurred on forced lid closure. Gaze to either side was accompanied by lid elevation on the side of the adducting eye and lid droop on the abducting side (Fig. 1). Upward gaze produced moderate upward-beating nystagmus with marked synchronous left upper eyelid jerks. Attempted downward gaze produced unsustained left eyelid nystagmus. A moderate upward nystagmus response accompanied by left lid nystagmus could be elicited on optokinetic stimulation, but no downward beats were evoked; horizontal optokinetic nystagmus was normal aside from minimal right medial rectus slowing. Neither convergence nor miosis occurred on near fixation with either eye.

Discussion

Misdirection has been observed following third cranial nerve injury from aneurysms, trauma (including surgical trauma), syphilis and other meningovascular inflammation, congenital causes, and cavernous sinus thrombosis.8,9,10 By far the most common association is with carotid aneurysm, with various studies disclosing an 84%, 5, 38%, 7 and 50% 4 incidence of aberrant regeneration following third nerve palsy due to aneurysm. Bilateral misdirection is rare. One case following a subacute traumatic subdural hematoma with probable secondary tentorial herniation has been described briefly.6 Knowledge of another case following trauma is mentioned by Walsh and Hoyt.11 The specific pattern of regeneration is unpredictable in a given instance, but certain phenomena are frequently observed; these include synkinetic lid elevation and pupillary constriction when innervation of other third nerve muscles is attempted, vertical eye movement limitation (upward usually greater than downward), absent vertical optokinetic response, and medial eye deviation on attempted vertical eye movement.

In the present case, ipsilateral lid elevation on adduction of either eye indicated that some fibers normally innervating the medial recti had regenerated aberrantly to supply the levator muscles. Vertical nystagmus indicated residual brain stem dysfunction, but the syn-
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chronous left eyelid nystagmus probably represented misdirection of superior rectus fibers to the levator muscle. The inability to depress the globes, in the presence of nearly normal elevation, would be unusual in misdirection, and may represent a central palsy of downward gaze.

Differentiation between direct third nerve damage and oculomotor dysfunction from secondary tentorial herniation is a common and frequently difficult task in the patient with head trauma. Diagnostic confusion with the resulting unnecessary surgical measures in the present case might have been obviated by the observation of immediate pupillary and globe paralysis; however, the rarity of bilateral traumatic oculomotor nerve injury in patients with reasonable survival potential made this a difficult diagnosis.

References

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