Aberrant regeneration in idiopathic oculomotor nerve palsy

Case report

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Aberrant regeneration following injuries to the oculomotor nerve is considered rare except in cases of posterior communicating artery aneurysms and trauma. A case of idiopathic third nerve palsy with pupillary involvement occurred in an otherwise healthy 38-year-old man. Follow-up examination 32 months later showed evidence of oculomotor function with aberrant regeneration.

KEY WORDS: idiopathic mononeuritis, oculomotor palsy, cranial nerve palsy, aberrant regeneration

A berrant regeneration following injuries to the oculomotor nerve is considered rare except in cases of posterior communicating artery aneurysms and trauma. Aberrant regeneration may cause lid elevation with adduction of the eye, pseudo-Graefe phenomenon or elevation of the lid with attempted downward gaze, and lack of vertical movement with retraction or adduction of the eye on attempted vertical gaze. In addition, any extraocular movement except abduction may result in synkinetic pupillary constriction.

We document the development of aberrant regeneration of the oculomotor nerve in a patient in whom no etiology was found, despite an extensive search.

Case Report

This 38-year-old man had the sudden onset of generalized malaise and excruciating right periorbital pain. Two days later he developed diplopia, and noted ptosis of the right eyelid. Over the ensuing 10 days, there was progressive dilatation of the right pupil and development of a complete right external and internal ophthalmoplegia. There was no history of trauma, diabetes, hypertension, fever, or antecedent illness.

On examination, he appeared healthy, but was apprehensive and complained of a dull headache. He was afebrile, blood pressure was 128/80 mm Hg, and pulse was regular at 88 per minute. The general physical examination was normal. Neurological examination showed complete ptosis of the right eyelid, a fully dilated, unreactive pupil, exotropia, and complete paralysis of the superior, inferior, and medial rectus muscles on that side. The right fourth and sixth cranial nerves were intact. Corneal reflexes were symmetrical. Funduscopic examination was unremarkable, and the remainder of the neurological examination was normal.

The patient's course over the next 10 days suggested the presence of an aneurysm or tumor. Skull and sinus x-ray films were normal. Sedimentation rate was 10 mm/hr. Hemogram, routine blood chemistries, and urinalysis were normal. Right carotid and verteobasilar angiography was normal. Cerebral and orbital computerized tomography scans with contrast enhancement were also normal. On spinal tap, opening pressure was normal. The cerebrospinal fluid (CSF) was acellular with mild protein elevation of 58 mg/dl. Repeat CSF examination 3 weeks later showed a normal CSF protein of 44 mg/100 ml, and normal immunoglobulins. Testing for antinuclear antibodies was negative. Three-hour glucose tolerance test was within normal limits. The CSF and throat and rectal swabs were negative for virus isolation.

There was no further deficit. Three months later, the patient began to experience signs of oculomotor...
Aberrant regeneration of third nerve regeneration, manifested by twitches of the eyelid and a periorbital tingling sensation. Function to the levator palpebrae returned within 6 months. Examination at that time showed a dilated, unreactive pupil, partial voluntary elevation, and depression of the right eye and medial rectus function.

Follow-up examination 32 months after the onset of ophthalmoplegia showed a fixed partial third nerve palsy on the right with evidence of aberrant regeneration. The voluntary elevation of the right eye noted 6 months after admission was no longer present (Fig. 1A). Medial rectus function was present. The eyelids were symmetrical in the primary position (Fig. 1B), but the right eyelid fell on right lateral gaze (Fig. 1C), and there was paradoxical elevation of the right eyelid and pupillary constriction on left lateral gaze (Fig. 1D) and on downward gaze (Fig. 1E).

Discussion

Aberrant regeneration of the oculomotor nerve in this case is manifested by synkinetic elevation of the right eyelid and pupillary constriction with medial rectus action on left lateral gaze. The loss of superior and inferior rectus motility, after its initial recovery, is further evidence of aberrant regeneration.

Gowers first described aberrant regeneration of the oculomotor nerve 100 years ago. Bielschowsky was among the first to postulate misdirection of the regenerating nerve fibers to explain the phenomenon. Bender and his co-workers confirmed this in chimpanzees. They found that aberrant regeneration was permanent and could be abolished by section of the oculomotor nerve. They likened aberrant regeneration to the well-known phenomenon of facial synkinesis following Bell’s palsy. Holland showed with electromyography that residual limitation of motion was due not to failure of reinnervation but to simultaneous reinnervation of antagonistic muscles in oculomotor nerve injury.

Isolated oculomotor nerve palsy is commonly thought to be due to either trauma, diabetes mellitus, or aneurysm of the posterior communicating artery. Rucker’s series, however, showed that fully 25% of all oculomotor nerve palsies had no apparent cause. Although the etiology of idiopathic oculomotor nerve palsy is unknown, the transient elevation of CSF protein in this case suggests a possible similarity to the mechanism of seventh nerve injury in Bell’s palsy. Clinical and electrodiagnostic studies suggest that idiopathic seventh nerve palsies may represent a cranial polyneuropathy akin to Guillain-Barré syndrome. Documentation of isolated oculomotor nerve palsies following infectious mononucleosis further suggests the possibility of postinfectious demyelination as a mechanism of injury.

While aberrant regeneration is common in cases of third nerve palsies due to aneurysm and trauma, it is not thought to occur in diabetes, ophthalmoplegic migraine, or in most tumors. Rare causes of aberrant regeneration include inflammatory processes, such as syphilis, cavernous sinus thrombosis, cavernous sinus meningioma, and cholesteatoma of the temporal lobe. Bilateral and congenital aberrant regeneration have both been reported. Walsh, in his Montgomery lecture to the Irish Ophthalmological Society in 1957, presented

Fig. 1. Aberrant regeneration of the right oculomotor nerve following idiopathic mononeuritis. A: Failure to elevate the eye on upward gaze. B: Symmetrical eyelids with pupillary asymmetry in the primary position. C: Ptosis on right lateral gaze. D: Lid elevation and pupillary constriction on left lateral gaze. E: Paradoxical lid elevation on attempted downward gaze.
three cases of aberrant regeneration in which no cause could be found. None of those cases, however, had pupillary involvement.

Aberrant regeneration following spontaneous oculomotor nerve palsy need not always be ominous. On the other hand, idiopathic oculomotor nerve palsy with or without pupillary sparing cannot be predicted to resolve without aberrant regeneration, and may persist with disabling diplopia.

References

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