Aberrant Regeneration of the Third Cranial Nerve in a Patient with Severe Head Injury

Ağır Kafa Travmalı Bir Hastada Aberan Üçüncü Sinir Rejenerasyonu

BAŞAR ATALAY, ŞANSAL GEDİK, HAKAN CANER,
SİBEL OTO, MURAD BAVBEK, NUR ALTINÖRS

Başkent University Faculty of Medicine, Department of Neurosurgery (BA, HC, MB, NA)
Başkent University Faculty of Medicine, Department of Ophthalmology (ŞG, SO)

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Abtract: Aberrant regeneration of the third cranial nerve is a well-known condition that can develop after head trauma or intracranial aneurysms. Horizontal gaze-eyelid synkinesis, pseudo-Von Graefe phenomenon, and limited elevation and depression of the eyelid with retraction of the globe are common signs of aberrant regeneration of the third nerve. These features may appear months to years after third nerve injury. Patients who have suffered severe head trauma or have had an intracranial aneurysm near the third cranial nerve should be followed in consultation with ophthalmologists. They should also be told about the possibility of aberrant regeneration of this nerve, and about the features of this condition.

Key Words: Aberrant regeneration, intracranial aneurysm, third nerve palsy, trauma

Özet: Aberan üçüncü sinir rejenerasyonu kafa travmaları ve intrakranial anevrizmalar sonrasında görülebilen iyi tanımlanmış bir durumdur. Yatay bakışta gözkapağı sinkinezisi, psödo Von Graefe Bulgusu, yukarı ve aşağı bakış kısıtlılığı ve gözün içeri retraksiyonu aberan rejenerasyonun sık görülen bulgularıdır. Bu bulgular üçüncü sinir hasarından aylar ya da yıllar sonra ortaya çıkabilir. Üçüncü sinir felci olan ağır kafa travması veya intrakraniyal anevrizma hastaları oftalmologlarla birlikte takip edilmeli ve aberan rejenerasyon bulguları hakkında bilgilendirilmelidir.

Anahtar Kelimeler: Aberan rejenerasyon, intrakraniyal anevrizma, travma, üçüncü sinir felci

INTRODUCTION

The third cranial nerve, or oculomotor nerve, supplies most of the extraocular muscles, including the inferior oblique, the medial, superior and inferior recti, and the levator muscle of the upper eyelid. It is responsible for ocular motility in the horizontal, vertical and torsional planes, and for levator function (1). In addition, the oculomotor nerve carries the parasympathetic fibers to the smooth muscles of the pupillary sphincter and ciliary muscle.

The congenital and acquired forms of oculomotor nerve palsy may be partial (one or more muscles affected) or complete (pupillary function also affected). Ptosis, a fixed and dilated pupil, and a down-and-out resting eye position are the classical manifestations of complete third nerve palsy (5,6,8,11). The common clinical findings of partial third nerve palsy are variable ptosis and paresis of ocular motility in accord with the muscles affected. Aberrant regeneration of the oculomotor nerve was first described by Gowers (4) in 1879. This condition is a well-known eye movement disorder that usually occurs after acute third nerve injury. Trauma and aneurysms are the most common causes of this problem (2,3,7). Here we report a case of aberrant regeneration of the third cranial nerve in a patient who had suffered severe traumatic brain injury.

CASE REPORT

A 17-year-old female presented to our neurosurgery clinic with the complaint of increased frequency of epileptic fits. At the time, she was experiencing generalized tonic-clonic seizures 1-2 times weekly. Before admission she was on phenytoin treatment and the seizures were under control. The patient's medical history included cranial surgery at 2 years of age for treatment of a right frontoparietal intracerebral hematoma caused by severe head trauma. She had started to have epileptic attacks at 7 years of age. At the age of 12 (10 years after the initial surgery), the patient had undergone cranioplasty for reconstruction of frontoparietal bone defects.

Her neurological examination at admission revealed mild left-sided hemiparesis and right total oculomotor nerve palsy. An electroencephalogram showed right parietotemporooccipital paroxysmal epileptic activity. Cranial magnetic resonance imaging showed encephalomalacic changes in the right frontoparietal region, a porencephalic cyst in the right lateral ventricle, and atrophy of the corpus callosum (Figure 1).

Treatment with 5-mg/kg/day phenytoin was prescribed, and this brought the seizures under control.

Ophthalmic Findings:

The patient's visual acuity was 20/400 in the right eye and 20/20 in the left eye. The right eye showed severe ptosis (Figure 2). The vertical fissure height in that eye was 4 mm, and the upper lid excursion was 2 mm. It was not possible to measure the margin-reflex distance, so only one-third of the iris was visible. In primary position, there was 25 PD exotropia and 6 PD hypotropia. All left eye movements appeared normal, but the right eye showed severely restricted adduction,

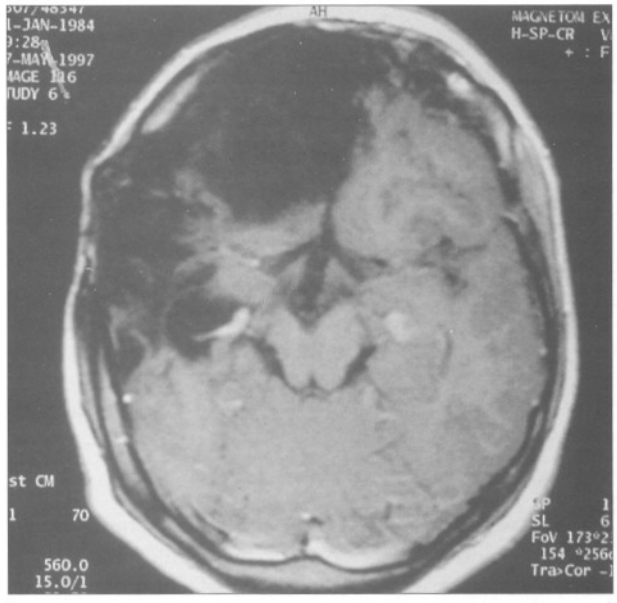

Figure 1. Cranial magnetic resonance imaging showed encephalomalacic changes in the right frontoparietal region, a porencephalic cyst in the right lateral ventricle, and atrophy of the corpus callosum.

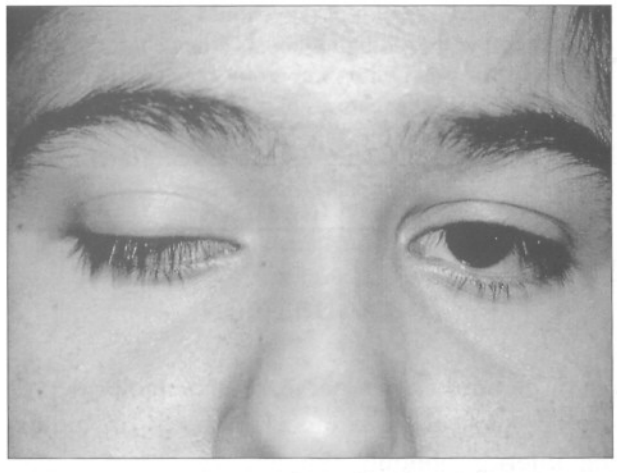

Figure 2. Severe ptosis in the right eye.

supraduction and infraduction (Figure 3). Forced adduction was accompanied by retraction of the upper lid, and abduction was accompanied by lid depression. The right lid became elevated on attempted downward gaze. Results of forced duction tests were unremarkable. Anisocoria was noted in primary position at average room light and under dim illumination. In room light, pupillary diameter was 5-6 mm on the right and 3 mm on the left. Direct and consensual pupillary light reflexes were absent on the right, and the diameter of the right pupil decreased to 4-5 mm on attempted downward gaze and right gaze. Slit-lamp examination revealed no segmental paresis of the sphincter. The patient did not exhibit nystagmus. Fundus examination with a +90 D lens showed atrophy of the optic disc in the right eye (Figure 4).

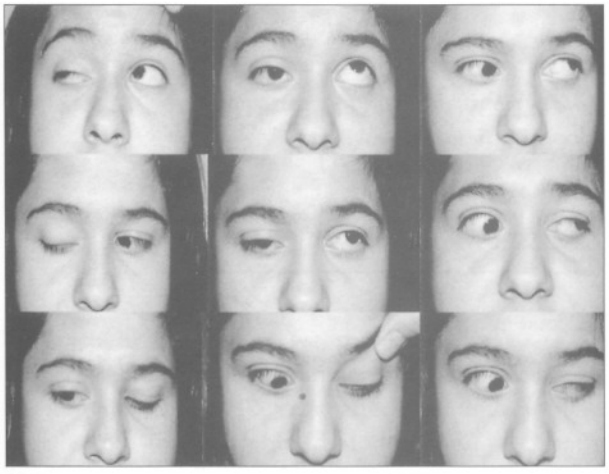

Figure 3. Severely restricted adduction, supraduction and infraduction in the right eye.

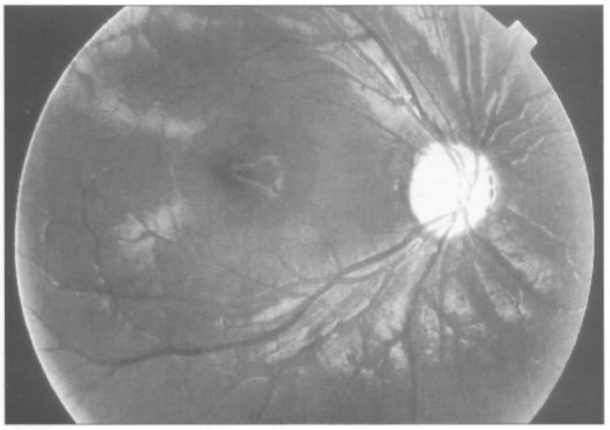

Figure 4. Fundus examination of the right eye revealed optic disc atrophy

DISCUSSION

The incidence of aberrant regeneration of the third nerve after acute third nerve injury is 15% (2). Head trauma and aneurysms of the posterior communicating artery are the most common causes of this condition. Horizontal gaze-eyelid synkinesis, pseudo-Von Graefe phenomenon (elevation of the lid on downward gaze), and limited elevation and depression of the eye with retraction of the globe with attempted vertical movements are common signs of aberrant regeneration of the oculomotor nerve (10). Patients may also show gaze-evoked pupillary constriction (pseudo-Argyll Robertson pupil). The signs of nerve fiber misdirection may appear months to years after the third nerve injury occurs. Misdirection of regenerating third nerve fibers, ephaptic transmission, and central synaptic reorganization are the proposed mechanisms for this condition (9).

Our patient developed aberrant regeneration of the oculomotor nerve after traumatic brain injury. She showed full-blown signs of fiber misdirection, including elevation of the right eyelid on adduction, retraction of this eyelid on downward gaze, limited elevation and depression of the eye and pseudo-Argyll Robertson pupil on downward gaze and adduction. Severe head trauma resulted in traumatic optic disc atrophy and reduced visual acuity (20/400) in the right eye.

Aberrant regeneration of the third cranial nerve should be kept in mind as a possible late complication in patients who suffer severe head injury involving the third nerve. Patients should be warned about the possibility of nerve fiber misdirection and the potential problems that this causes. Follow-up should be done in consultation with ophthalmologists.

Correspondence: Başar Atalay, MD
Başkent University
Faculty of Medicine
Department of Neurosurgery
Fevzi Çakmak Caddesi,
06490 Bahçelievler, Ankara, Turkey
Phone : 212 6868/1368
Fax : 223 7333
E-mail : basara@baskent-ank.edu.tr

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