

Congenital Nuclear Syndrome of Oculomotor Nerve

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A patient is reported who suffered from a fixed, non-progressive encephalopathy caused by a lesion involving the left portion of both the mesencephalon and basal ganglia; the lesion was caused by an acquired prenatal vascular insult. The clinical expression of third cranial nerve palsy corresponds to a nuclear syndrome of the left oculomotor nerve, affecting both eyes asymmetrically, later developing into aberrant reinnervation.

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Introduction

Ocular cranial nerve palsies are usually a sign of a group of disorders that is acquired later in life and most commonly is vascular, traumatic, or neoplastic in origin. Congenital causes account for fewer than 10% of all affected patients according to some series [1].

When present since birth, paralysis of the third cranial nerve was usually believed to be an isolated phenomenon, unrelated to other neurologic or systemic disorders [2]. In such cases, a peripheral lesion was presumed to be based, in part, on the common observation of a later development of aberrant reinnervation [3]; however, with the availability of advanced scanning techniques, the association of congenital oculomotor palsy with central nervous system (CNS) lesions is being increasingly reported [4]. It is unknown whether these associated CNS lesions are dysplastic or encephaloclastic, but evidence points to the latter.

Congenital oculomotor palsy is usually unilateral. To our knowledge, only 5 bilateral patients have been reported [5,6]. In one of them, the bilateral symptoms

appeared to have been caused by a unilateral brainstem lesion [5]. Such an event was described in 2 adult patients by Pierrot-Deseilligny et al., naming the disorder as the "nuclear syndrome of the oculomotor cranial nerve" [7].

We report a patient with bilateral congenital oculomotor nerve palsy, in whom the neuroradiologic findings support the hypothesis of a central mechanism, mainly a unilateral vascular infarct of the brainstem acquired prenatally.

Case Report

This term boy was born weighing 3,400 gm to a 38-year-old mother by a normal, spontaneous vaginal delivery after a normal pregnancy. Apgar score at 1 min was 5 which was attributed to nuchal cord entanglement. With only manual ventilation, the 5-min Apgar score was 8. The neonatal period was otherwise uneventful except for jaundice (serum bilirubin 18.2 mg/dl) that was controlled after 72 hours of phototherapy.

The patient was referred at 3 days of age because of ptosis and ophthalmoplegia. The primary position of the left eye was exotropic, with no upward, downward, or adduction movement. A ptosis of the left eyelid caused a narrow palpebral fissure 1 mm wide. The left pupil was mydriatic and nonreactive.

Primary position of the right eye was hypotropic and exotropic. Adduction to the midline was possible, but not beyond. Ocular elevation was absent. A moderate ptosis of the right eyelid spared the pupil. Size and reaction to light of the right pupil was normal. During doll's eye maneuvering, horizontal nystagmus of the right eye was observed. There was also cervical muscular hypotonia without localizing signs. Alertness, crying, sucking, and other primitive reactions were normal, as were the remaining cranial nerves. Optic fundi examination revealed coloboma of the papilla bilaterally. The occipito-frontal circumference was 36 cm. A dilated left lateral ventricle was identified by ultrasonography. The initial lumbar puncture was traumatic. Electroencephalography was normal.

At 7 months of age, right hemiparesis was detected. Independent gait was attained at 20 months of age. Mental retardation was later evident with an I.Q. of 55.

At present, the patient is 4 years of age and suffers from a right spastic hemiplegia associated with a permanent flexion of the head towards the right shoulder, with a chin-up posture. The left eye remains fixed in a primary position of exotropia (Fig 1). The upper eyelid appears retracted and the pupil miotic without reaction to light. The right eye, in primary position of hypotropia, is unable to adduct beyond the midline. The right pupil reacts normally and ptosis of the eyelid covers the upper border of the pupil, in contrast to the neonatal period when ptosis was much more severe on the left side. When the patient cries, a complex phenomenon appears which reverses the eyelid positions; whole closure over the left eye is attained while the right eye remains open. When attempting lateral gaze, horizontal nystagmus of the right eye occurs. When displacing the head laterally, the eye passes through the midline for a few seconds.

MRI demonstrated atrophy of the left portion of the mesencephalon, with a wedge-shaped loss of tissue over the left cerebral peduncle that reached the cerebral aqueduct (Fig 2). Left cerebral hemiatrophy manifested with the disappearance of thalamic and lenticular nuclei, as well as the internal capsule. The left hemispherical gray-white matter junction was indistinct.

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Figure 1. Patient at 4 years of age. The left eye is exotropic and the upper eyelid is retracted. The right eye (R) is hypotropic with more severe ptosis. All of these findings were the reverse of those observed in the neonatal period.

Discussion

Congenital unilateral oculomotor palsies are seldom associated with other neurologic deficits. Balkan and Hoyt reported hemiparesis contralateral to cranial nerve palsy in 4 of 10 patients, suggesting that a brainstem infarct could be the cause, although they lacked clinical-neuro-radiologic correlation [8]. Such correlation has been provided by MRI. Hamed reported an associated neurologic



Figure 2. T₁-weighted MRI revealed a wedge-shaped infarct of the left cerebral peduncle (arrow) and hemiatrophy of the left cerebral hemisphere.

disorder in 10 of 14 patients, 1 of whom exhibited corresponding profound midbrain damage on MRI [4]. White et al. described 2 patients with associated cerebellar hypoplasia [9].

Bilateral congenital third cranial nerve palsy is seldom encountered. Good et al. reported a 3-month-old patient with bilateral but asymmetric congenital oculomotor palsy whose MRI disclosed unilateral absence of caudate and lentiform nuclei, as well as hypoplasia of the extraocular muscles innervated by the ipsilateral third cranial nerve [6]. The authors presumed an early CNS insult arising before the oculomotor nerve fibers reached the appropriate extraocular muscles (before the sixth week of gestation). Based on the MRI findings, they favored a developmental anomaly of the brain rather than a destructive process as the original cause. Their patient did not demonstrate an aberrant reinnervation of the pupil at age 3 months.

In 1981, a nuclear syndrome of the third cranial nerve was first described by Pierrot-Deseilligny et al. in 2 adults who presented with oculomotor nerve palsy on one side and isolated paralysis of the superior rectus muscle on the other [7]. The bilateral clinical expression was due to a unilateral lesion at the site of the oculomotor nucleus, and could be explained by previous experimental studies [10] that established a contralateral innervation of the superior rectus muscle, as well as bilateral innervation of the levator palpebral muscle. All other extraocular muscles have ipsilateral innervation.

CNS lesions of arterial territories, occurring prior to birth, have been widely documented ever since the introduction of MRI [11]. The presence of a vascular lesion specifically located at the site of the oculomotor nucleus was established by Norman [12] at autopsy in a 30-week preterm infant, in whom thrombosis of the basilar artery was demonstrated. That region of the brainstem is supplied exclusively by paramedial branches of the basilar artery. A basilar thrombus could also occlude other arterial branches, such as the mesencephalic portion of the posterior cerebral arteries, causing infarction of the basal ganglia, as can be seen in the "top of the basilar artery syndrome," in adults. This condition could have occurred in our patient, leading to a nuclear syndrome of the third cranial nerve as well as a lesion in the homolateral basal ganglia.

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