

Primary isolated aberrant misdirection of the oculomotor nerve in a young girl: Case report and review of literature.

Author

Raed Shatnawi

MD, FRCS (Glasg)

Consultant Pediatric Ophthalmologist

Prince Hamza Hospital

Assistant Professor of Ophthalmology

Hashemite University Medical School

Amman - Jordan

Co-Authors

Raed Shatnawi, Anas Injarie, & Yassir Abou-Rayyah

Clinical and Academic Department of Ophthalmology

Great Ormond Street Hospital, London WC1N 3JH, United Kingdom.

Abstract: Aberrant misdirection of the oculomotor nerve is a condition related to the critical central nervous system problem. In this case, a 5-year-old girl was diagnosed with aberrant misdirection of oculomotor nerve with right upper eyelid ptosis but without any pathology of central nervous system. The case having a great significance in terms of its uniqueness and can challenge the pediatric ophthalmologist and oculoplastic surgeon.

1. Introduction

Aberrant misdirection of the oculomotor nerve is most commonly associated with underlying serious central nervous system pathologies such as aneurysms and meningiomas^{1, 2}. It is very rare to have an isolated misdirection of the oculomotor nerve without underlying pathology^{3, 4}. In this case, where the patient has misdirection of the oculomotor nerve without any underlying pathology related to central nervous system.

2. Case Report

A 5-year-old girl was referred to the hospital for possible surgical management of right upper eyelid ptosis with an abnormal eye movement. She was born through normal vaginal delivery with uncomplicated pregnancy and postnatal period.

Past medical history having a significance in terms of asymptomatic murmur that was investigated

thoroughly including echocardiography, which was normal. There was no significant family history related to ocular motor nerve. No history of head or ocular trauma was observed. The patient's mother informed that the ptosis appeared since patient's early childhood, which is variable in severity since then.

On examination, visual acuity was 0.08 in the right eye and 0.04 in the left eye (crowded LogMar). Examination of both anterior and posterior segment were unremarkable with normal cycloplegic refraction in both eyes. The patient was orthotropic with full range of eye movements in all positions of gaze. There was a right upper eyelid ptosis in primary gaze position. The palpebral fissure height in primary gaze was 8 mm in the right eye and 12 mm in the left eye. Lid margin to reflex distance was -1.0 mm in the right eye and 4 mm in the left eye. Levator muscle function in the right eye was 8.5 mm, and in the left eye it was 13 mm. Bell's phenomenon was normal in both eyes with normal corneal sensations.

No eyelid winking movements were associated with jaw movement, but the other signs were observed. The patient has more ptosis on abduction. She has a lid retraction on adduction, down gaze, and convergence. There was no globe retraction on attempted abduction.

Pupillary reflexes were normal and symmetrical with no significant anisocoria. The examination of other cranial nerves and neurological system were within normal limits.

On the basis of above findings, involvement of the oculomotor nerve was suspected. Magnetic resonance imaging (MRI) of the brain and orbit was recommended to monitor the compressive lesions of the oculomotor nerve. The MRI was reviewed by two separate neuroradiologists, and it was reported to be normal with no suspicion of any orbital or intracranial lesions. No space-occupying lesions or aneurysms could be identified.

At this stage, a diagnosis of an isolated primary aberrant misdirection of the right oculomotor nerve was made. The diagnosis and different surgical options were explained to the family. They decided not to proceed with any surgery due to unpredictable surgical outcomes.

3. Discussion

The oculomotor nerve travels in the brain under the posterior cerebral artery, above the superior cerebral artery and adjacent to the posterior communicating artery. The close proximity of the oculomotor nerve to the blood vessels accounts for the frequent involvement of this nerve by vascular disorders, such as aneurysms of the posterior communicating artery. Once the oculomotor nerve penetrates the dura, it continues anteriorly in the lateral wall of the cavernous sinus. In the anterior cavernous sinus, the oculomotor nerve divides into two divisions: superior and inferior. The superior division innervates the levator and superior rectus muscles, whereas the inferior division innervates the inferior rectus, the medial rectus, and the inferior oblique muscles⁵. In the patient, the lid retraction occurred on abduction and down gaze, which indicates that the aberrant misdirection occurred between the inferior division of the oculomotor nerve and the levator muscle or its innervating branch.

The above signs are called Fuchs' sign which is a lid retraction on abduction and a pseudo-Graefesign which is a lid retraction in down gaze⁶. Most cases with aberrant misdirection occur as a result of aberrant regeneration subsequent to oculomotor nerve palsy or trauma. In most published cases, the aberrant regeneration of the oculomotor nerve occurred after compressive trauma to the oculomotor nerve such as aneurysms and tumors^{1, 2}, or sometimes associated with ophthalmoplegic migraine⁷. There are only a few reports that described the occurrence of misdirection of the oculomotor nerve without underlying central nervous system pathology^{3, 4}.

There are few theories in the literature that considered this subject and tried to explain this phenomenon. One theory suggested that the newly formed axons of the oculomotor nerve reach to the

muscles not originally connected to them⁸. While the other theories tried to link this phenomenon to congenital cranial dysinnervation disorder⁹, some authors believed that this congenital synkinetic movement reflects an incomplete nerve cell apoptosis that occurs during the embryonic phase of development¹⁰. Whatever the case but this case is unique in its presentation and signs. Therefore, the subsequent management may give a challenge to both the pediatric ophthalmologist and the oculoplastic surgeon.

4. References

- [1] Güresir, Erdem, Patrick Schuss, Volker Seifert, and Hartmut Vatter. "Oculomotor nerve palsy by posterior communicating artery aneurysms: influence of surgical strategy on recovery: Clinical article." *Journal of neurosurgery* 117, no. 5 (2012): 904-910.
- [2] Lyons, C. J., F. Godoy, and E. ALQahtani. "Cranial nerve palsies in childhood." *Eye* 29, no. 2 (2015): 246-251.
- [3] Bursztyn, Lulu LCD, and Inas Makar. "Congenital Trochlear–Oculomotor Synkinesis." *Journal of Neuro-Ophthalmology* 34, no. 1 (2014): 64-66.
- [4] Ishiyama, Akihiko, Yoshiaki Saito, Kenji Sugai, Masayuki Sasaki, Hiroko Yamazaki, and Masanori Kohama. "Concurrence of multiple types of eyelid synkinesis in a patient with congenital anomalies." *Brain and Development* 34, no. 2 (2012): 140-142.
- [5] Demer, Joseph L., and Robert A. Clark. "Magnetic resonance imaging of differential compartmental function of horizontal rectus extraocular muscles during conjugate and converged ocular adduction." *Journal of neurophysiology* 112, no. 4 (2014): 845-855.
- [6] Kim, Si Bum, Sei Yeul Oh, Moo Hwan Chang, and Sungeun E. Kyung. "Oculomotorneuromyotonia with lid ptosis on abduction." *Journal of American Association for Pediatric Ophthalmology and Strabismus* 17, no. 1 (2013): 97-99.
- [7] Chugh, J. P., Prachi Jain, R. S. Chouhan, and Ashok Rathi. "Third Nerve Palsy: An Overview." (2012).
- [8] Sayed, Sayed A., and Mahmoud Rabea. "Misdirection (aberrant regeneration) of the third cranial nerve." *Journal of the Egyptian Ophthalmological Society* 106, no. 3 (2013): 150.
- [9] Gutowski, Nicholas J., and J. K. Chilton. "The congenital cranial dysinnervation disorders." *Archives of disease in childhood* (2015): archdischild-2014.
- [10] Bursztyn, Lulu LCD, and Inas Makar. "Congenital Trochlear–Oculomotor Synkinesis." *Journal of Neuro-Ophthalmology* 34, no. 1 (2014): 64-66.