Original paper

Neuro-Ophthalmology
0165-8107/01/US$16.00

Neuro-Ophthalmology – 2001, Vol. 26, No. 2, pp. 79–83

Accepted 30 April 2002

Migraine aura and diplopia phenomenology associated with congenital nystagmus

L.F. Dell'Osso[1–3]
R.B. Daroff[1,2]
R.L. Tomsak[2]

[1]Ocular Motor Neurophysiology Laboratory, Veterans Affairs Medical Center, and the Departments of [2]Neurology and [3]Biomedical Engineering, Case Western Reserve University and University Hospitals of Cleveland, Cleveland, OH, USA

Abstract An observer with horizontal/torsional congenital nystagmus (CN) made two unique observations. One occurred during a migrainous aura and the other during a decompensated vertical phoria. The otherwise typical migraine aura was perceived as oscillating horizontally over a stable visual field – an oscillating scintillating scotoma. The vertical diplopia secondary to a decompensated phoria oscillated vertically – a diplopic oscillopsia. The implications raised by these observations strengthen the role of efference copy in the usual suppression of oscillopsia in CN. Retinal rivalry is proposed to explain the possible perception of vertical oscillopsia without vertical nystagmus.

Key words Congenital nystagmus; migraine; scotoma; oscillopsia; diplopia

Introduction Clinical phenomena are 'experiments of nature' and, when properly analyzed, may be the source of considerable knowledge of human neurophysiology. This is especially true of ocular motor research in general and of congenital and other types of nystagmus in particular. Migraine auras are usually visual and consist of scintillating lights (photopsias) and scotomata (fortification spectrum). The aura's position may be stationary, but usually migrates slowly across the visual field, with a typical duration of approximately 20 minutes or less. The scintillating aura may be thought to be 'moving' since it might be perceived to move with voluntary eye movements, but it will not continually oscillate. Many auras, particularly in the middle-aged or elderly, are not associated with a headache.[1] Diplopia is the appearance of two *static* images of the visual scene, usually caused by the misalignment of the eyes in one or more planes. CN is usually a horizontal-torsional oscillation of the eyes that, in its purest form, is not associated with an afferent deficit or strabismus, not accompanied by oscillopsia, and does

Correspondence and reprint requests to:
L.F. Dell'Osso, Ph.D.
Ocular Motor Neurophysiology Laboratory
Veterans Affairs Medical Center (127A)
10701 East Boulevard
Cleveland, OH 44106
USA
Tel: (216) 421-3224
Fax: (216) 231-3461
E-mail: lfd@po.cwru.edu

Acknowledgements:
This work was supported in part by the Office of Research and Development, Medical Research Service, Department of Veterans Affairs.

not disturb binocular single vision. This report describes two *unique* observations made by an observer with congenital nystagmus (CN). The first occurred during a migrainous visual aura and the second during a transient period of hypertropia.

Case report The subject was a 60-year-old male with hereditary CN, best-corrected visual acuity of 20/25 OU, 60 arc seconds of stereopsis, a small vertical phoria, and no afferent deficits. Prior to the events in this report, he had never experienced a migraine headache, visual aura, or diplopia. An MRI scan of the head was normal. Quantitative oculographic recordings made over a 30-year period, using both search-coil and video recording systems, verified that the subject's CN was purely horizontal-torsional, without a vertical component.

Observations

OBSERVATION 1 In December of 1999, the subject developed the first of what became recurrent (approximately 12 in 15 months) stereotyped visual events, consisting of a central scotoma capped with a typical scintillating fortification spectrum. These were compatible with the International Headache Society diagnosis of 'migraine aura without headache'.[1]

During each event, the scotoma and visual spectrum *continuously oscillated* horizontally (at ≈2–3 Hz) over the stable visual field. The horizontal oscillation continued as the spectrum migrated slowly to the right lower quadrant, eventually occupying the right lower periphery of the visual field. The duration of the events was 10–12 min and none were associated with a headache. No torsional oscillopsia was perceived.

OBSERVATION 2 A sudden onset of vertical diplopia occurred while the subject rapidly descended a flight of stairs. The lower diplopic image of the visual field, coming from the right eye, appeared to be oscillating vertically over a static image of the visual field from the left eye. Thus, the subject went from his normal stationary binocular perception of the world to one with diplopia and uniocular vertical oscillopsia. At the bottom of the stairs, he looked at a blank wall with a room number sign. The 2-inch high by 6-inch wide sign appeared as two signs, vertically displaced by ≈4 in, leaving a 2-inch separation between them, with the lower one oscillating vertically at ≈2–3 Hz with an estimated peak-to-peak amplitude of <5° (based on the distances involved). During the oscillation, the lower image did not overlap the upper one. Attempts to fuse the images brought them closer by raising the lower image, but did not achieve single vision. Pushing the right eye slightly downward by applying pressure on the upper eyelid also lessened the vertical diplopia. Occlusion of either eye eliminated both diplopia and oscillopsia. The spontaneous vertical diplopic oscillopsia lasted ≈3 min and has not reoccurred.

Discussion These two observations may provide insight into mechanisms that suppress oscillopsia in CN. When an after-image is placed

on the retina of the CN subject, it appears as a horizontally oscillating image over a stationary visual field.[2] Thus, the retinally stabilized afterimage causes oscillopsia, while the constantly moving visual field is perceived as stable (i.e., no oscillopsia). Extraretinal information about the motor signals driving the eyes (efference copy) may play a major role in oscillopsia suppression.[2–4] Models of the ocular motor system with CN *require* the use of efference copy to simulate the responses exhibited by individuals with CN to a variety of input stimuli.[5]

The observations made during the migrainous auras suggest that the image arising from the visual cortex produced the same perception as a stabilized retinal image; that is, oscillopsia in the major plane of the CN, while the visual scene traversing the retina is perceived as stable. We regard the use of efference copy of eye motion as the only reasonable mechanism by which the resulting retinal image motion is negated, producing a stable percept; stabilized retinal (or cortical) images are then perceived as oscillating. A relatively stationary cortical area was responsible for the aura that was perceived as oscillating horizontally, while the visual scene oscillating horizontally across the retina (and cortex) was perceived as horizontally stationary. During retinal image stabilization in the subject with horizontal-torsional CN, the retinally (and cortically) stationary visual scene is perceived as oscillating horizontally. Thus, under both conditions, the stable cortical areas/images produced horizontal oscillopsia. This is supported by Jung's observation that his left-hemifield migrainous fortification spectrum moved in the direction of vestibular slow phases during rotational stimulation.[6] Here, also, the cortically stable fortification spectrum was perceived as moving horizontally in the same direction as the uncompensated vestibular slow phases. The absence of any perceived torsional oscillation of the visual aura was probably due to both the smaller magnitude of the torsional component of the CN (resulting in a small efference-copy signal) and masking due to the scintillation of the complex fortification spectrum.

In contrast to the consistent perceptions during the multiple occurrences of the longer (10–12 min) migrainous aura, the one-time short (3 min) vertical diplopia did not cause oscillopsia in the horizontal plane of the CN of either image of the visual field and there was no migrainous aura. In the horizontal plane, the perception of each visual field image was independently stabilized, presumably using the efference copy from the motor commands to the respective eye. In horizontal CN, the oscillations in the two eyes are phase-locked, with equivalent waveforms and coincident foveation periods,[7,8] although the amplitudes of the two waveforms may differ as has been demonstrated in this subject.[9] Thus, the suppression of horizontal oscillopsia in each eye must occur independently, each using efference copy of the motor commands to that eye; the two stable perceptions are then combined into one binocular image of the world. Because no horizontal oscillopsia occurred during the vertical diplopia, we presume that the horizontal CN was unchanged. What then could be the source of the perceived *vertical* oscillopsia in the deviated eye? We will discuss three possibilities.

One hypothesis for the vertical oscillopsia experienced during transient vertical diplopia would require a singular occurrence of a verti-

cal component in the CN of the deviated eye that was *uncorrected* by efference copy. However, this is inconsistent with the absence of horizontal, vertical, or torsional oscillopsia in individuals with these components to their CN. That is, all components of CN occur within the efference copy feedback loop and oscillopsia in their respective planes is automatically suppressed.[4]

An alternative hypothesis involves either a rivalry or suppression mechanism (modulated by the CN oscillation) between the two disparate images. At each foveation period of the horizontal CN, the fixating eye might change, inducing a vertical shift in perceived position (due to the strabismus), even though neither eye is oscillating vertically. The constant alternation of perceived vertical position could have simulated the effects that would be produced by a vertical oscillation. The frequency of the perceived vertical oscillation was similar to the horizontal CN (≈2–3 Hz). In support of the rivalry hypothesis for the vertical oscillopsia is the observation that when the subject alternates fixation between the eyes (either using an occluder or by winking), the visual field is perceived to jump vertically downward when using the right eye and upward when using the left. This shift was secondary to his right hyperphoria and would manifest when the phoria decompensated to a tropia. Further study, under more controlled conditions, is necessary to explore this putative mechanism.

A final hypothesis is that a transient uniocular vertical nystagmus was responsible for, or coincided with, the sudden loss of vertical fusion and the vertical oscillopsia of the diplopic image. Although the retinal motion from the horizontal-torsional CN did not induce oscillopsia, an acquired uniocular vertical nystagmus, presumably occurring outside the efference-copy loop,[4] would cause vertical oscillopsia. Superior oblique myokymia (SOM) can induce uniocular vertical nystagmus and uniocular oscillopsia.[10] However, the pendular nystagmus associated with SOM is a low-amplitude (≈0.1°) high-frequency (up to 50 Hz) oscillation that is usually recurrent, lasts less than 10 seconds, and is associated with a *depression* of the affected eye. Shortly after this unique event, we attempted to duplicate the perceptions with a simple technique. Vertical fusion was deliberately broken by placing a 3D vertical base-up (or down) prism in front of the subject's right eye while he fixated a small target. The resulting vertical diplopia and vertical oscillopsia were similar to the perceptions produced by the decompensated vertical phoria. It is not known if this is an idiosyncratic response or if others with CN would have similar perceptions. Studies of these phenomena are now being designed using ocular motility recordings to provide data that might support one of more of these hypotheses; something that was impossible when this singular event occurred and was not part of the simple prism-induced observations. If indeed the loss of vertical fusion alone induced a vertical nystagmus, this would be a further area for study.

In conclusion, the complex perceptual sequela of either a migrainous aura (oscillating scintillating scotoma) or a nonmigrainous decompensated phoria (diplopic oscillopsia) in a visually introspective observer with CN revealed conditions when oscillopsia could no longer be suppressed in the vertical plane only and provides evidence supporting

oscillopsia suppression being dependent on efference copy of the unaffected horizontal motion. However, efference copy was unable to suppress diplopia or the perception of vertical oscillopsia that may have resulted from rivalry or a transient uniocular vertical oscillation.

References

1 Silberstein SD, Saper JR, Freitag FG. Migraine: diagnosis and treatment. In: Silberstein SD, Lipton RB, Dalessio DJ, editors. *Wolf's headache and other head pain*. 7th ed. New York: Oxford University Press, 2001;121–237.

2 Dell'Osso LF, Leigh RJ. Foveation period stability and oscillopsia suppression in congenital nystagmus. An hypothesis. *Neuro-ophthalmology*. 1992;12: 169–183.

3 Bedell HE, Currie DC. Extraretinal signals for congenital nystagmus. *Invest Ophthalmol Vis Sci*. 1993;34: 2325–2332.

4 Dell'Osso LF, Averbuch-Heller L, Leigh RJ. Oscillopsia suppression and foveation-period variation in congenital, latent, and acquired nystagmus. *Neuro-ophthalmology*. 1997;18:163–183.

5 Dell'Osso LF. Nystagmus basics. Normal models that simulate dysfunction. In: Hung GK, Ciuffreda KJ, editors. *Models of the visual system*. New York: Kluwer Academic/Plenum Publishers, 2002; 711–739.

6 Jung R. Translokation corticaler migrainephosphene bie augenbewegungen und vestibularen reizen. *Neuropsychologia*. 1979;17: 173–185.

7 Dell'Osso LF, Gauthier G, Liberman G, Stark L. Eye movement recordings as a diagnostic tool in a case of congenital nystagmus. *Am J Optom Arch Am Acad Optom*. 1972;49:3–13.

8 Dell'Osso LF, Van der Steen J, Steinman RM, Collewijn H. Foveation dynamics in congenital nystagmus I: fixation. *Doc Ophthalmol*. 1992;79:1–23.

9 Dell'Osso LF. Fixation characteristics in hereditary congenital nystagmus. *Am J Optom Arch Am Acad Optom*. 1973;50: 85–90.

10 Leigh RJ, Tomsak RL, Seidman SH, Dell'Osso LF. Superior oblique myokymia: quantitative characteristics of the eye movements in three patients. *Arch Ophthalmol*. 1991;109:1710–1713.