Case report

Cardinal features of superior oblique myokymia: An infrared oculography study

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ARTICLE INFO

Article history:
Received 18 November 2016
Received in revised form 21 April 2017
Accepted 20 June 2017
Available online 23 June 2017

Keywords:
Superior oblique myokymia
Diplopia
Oscillopsia
Trochlear nerve

ABSTRACT

Purpose: Superior oblique myokymia (SOM) is a rare eye movement disorder characterized by unilateral oscillopsia and binocular diplopia. Our study aimed to better understand SOM using infrared oculography.

Methods: We examined and recorded five patients with SOM.

Results: Binocular infrared oculography showed that in primary gaze, all patients exhibited torsional oscillations, which worsened in infraduction and abduction and improved in supraduction and adduction. Saccades showed increased downward saccade amplitudes but normal peak velocities. During fixation in primary gaze, removal of target led to extorsion and supraduction, unmasking underlying superior oblique weakness.

Conclusions and importance: Our data suggest both weakness and activity-dependent hyperactivity of the trochlear motor unit, supporting a model of injury followed by aberrant regeneration.

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1. Introduction

Superior oblique myokymia (SOM) is characterized by unilateral, intermittent contractions of the superior oblique muscle resulting in oscillopsia and binocular diplopia.1–4 It has been attributed to altered membrane thresholds of the neurons in the trochlear nucleus,2 associated with microvascular compression of the trochlear nerve and trochlear nerve palsy.3,5–9

2. Material and methods

2.1. Subjects

We studied 5 consecutive patients with SOM who were Caucasian women in their third to fifth decades, approved by the Stanford Institutional Review Board. None had a history of trochlear nerve palsy, and 3 had remote history of minor head trauma. Four experienced headaches, 2 of which were migrainous. Three were right-sided and 2, left-sided. All experienced monocular oscillopsia, and 2 had diplopia. In 4 patients, the same symptoms occurred more than 10 years prior and resolved spontaneously. Symptoms were typically worse with stress. Examination was significant for normal visual acuities and unremarkable anterior and posterior segments. Small, unilateral oscillations were often observable through the slit lamp or on direct ophthalmoscopy. Ocular alignment measured a 1–4 prism-diopter ipsilateral hyper- (2 patients) or hypotropia (3 patients) in primary gaze.2 Infraocular worsened symptoms and alignment in all patients. All patients exhibited a small contralateral head tilt, and ipsilateral head tilt worsened symptoms in one patient. Magnetic brain imaging with magnetic resonance angiography was unremarkable with no evidence of vascular compression in all except one, who had a slightly prominent vessel adjacent to the ipsilateral orbital trochlear nerve-muscle complex. In our patients, no medicine was effective for all patients, and multiple medications were tried as monotherapy. Treatment with carbamazepine or oxcarbazepine improved symptoms in 2 of 4 patients. Clonazepam helped 2 of 2 patients. Gabapentin helped 1 out of 2 patients. Pregabalin was ineffective in 2 patients. One patient was able to stop medication after lifestyle modifications to reduce stress.
2.2. Eye movement recordings

Eye movements in all patients were recorded using 60-Hz three-dimensional infrared oculography (3-D VOG, 60-Hz, SensoMotoric Instruments, Boston, MA, USA). Patients were asked to maintain primary gaze and to look in various eccentric gazes. One patient’s saccade amplitudes and velocities at different gazes were analyzed using 500-Hz two-dimensional infrared oculography (iView-X Hi-Speed, SensoMotoric Instruments, Boston, MA, USA).

2.3. Data analysis

Data were exported and analyzed using Excel and IgorPro. Statistical significance was determined using the Student’s t-test.

3. Results

In primary gaze, 60-Hz infrared oculography showed unilateral torsional oscillations (0–6°) lasting 20 ± 6 s (range 2–42 s), occurring on average twice per minute. Patients spent 72 ± 18% (range 47–85%) of time in torsional oscillations and 57 ± 10% (range 26–85%) in intorsion (N = 5 patients). Fig. 1 shows an example of a patient with left SOM who, in primary gaze, developed 2° torsional oscillations at 1-Hz. This oscillation intensified over seconds to 4° at 1–5 Hz, with a new onset intorsion. Scatter plot of this patient demonstrated significant torsional deviations of the affected eye compared with the unaffected eye (inter-ocular difference p < 0.00001 for each patient). Torsional mismatch was observed in all patients with an average difference of 2.4 ± 0.8° during events in primary gaze (N = 5 patients).

The intorsion and torsional oscillations varied depending on gaze (Fig. 2). Torsional oscillations increased during infraduction and abduction, which correlated with greater torsional scatter. Oscillations disappeared during supraduction and adduction. Downgaze, which triggered the worst symptoms, led to intorsion 78% of time and, on average, 4.9 ± 0.7° of intorsion of the affected eye compared with the unaffected eye (p < 0.00001 for each patient, N = 5 patients). Fifty-five percentage of downgaze was accompanied by sustained intorsion after returning to primary gaze (range 0.7–17.6 s, average duration 7.5 s).

We analyzed reflexive saccades in one patient using 500-Hz binocular two-dimensional infrared oculography. The downward saccades exhibited significantly larger amplitudes in the affected eye compared with the unaffected eye (p = 0.007) (Fig. 3A–B). There was no inter-ocular difference in the upward saccade amplitudes (p = 0.4), and inter-ocular peak velocities were similar in up, down, left, and right gazes (Fig. 3C–D, p = 0.6, 0.7, 0.2, 0.9, respectively).

Lastly, patients were recorded while fixating on a small light target in the dark. Upon removal of the target, there was expected gradual deviation of the eyes due to loss of the fixation. Interestingly, the affected eyes exhibited relative extorsion and supraduction compared to the unaffected eyes—changes found in trochlear nerve paresis (Fig. 4A–C, N = 5 patients). Oscillations also diminished over seconds. Among all patients, there was, on average, 3.9 ± 1.8° of extorsion of the affected vs. the unaffected eyes without visual feedback (p = 0.02). This extorsion in the absence of vision was significantly different from the intorsion in primary gaze (2.4 ± 0.8°, p = 0.01) and downgaze (4.9 ± 0.7°, p = 0.002) with visual feedback.

Fig. 1. Evolution of torsional oscillations in left superior oblique myokymia in primary gaze. Top. Three-dimensional eye positions. OD: black, OS: gray traces. Y-axis tick: 5-deg. Bottom left. Power analysis of the torsional oscillations. Bottom right. Torsional versus vertical eye position scatter plot.
Fig. 2. Torsional oscillations during infraduction and abduction in left superior oblique myokymia, with improvement in supraduction and adduction. **Top.** Three-dimensional eye positions. OD: black, OS: gray traces. Y-axis minor tick: 10-deg. **Bottom left.** Torsional versus horizontal and **Bottom right.** torsional versus vertical scatter plots. CW = clockwise, CCW = counterclockwise.

Fig. 3. Increased amplitudes but normal peak velocities of the affected eye in downgaze. **Top left.** Eye position traces in left superior oblique myokymia. OD: black, OS: gray. **Top right.** Pooled saccade amplitudes (**p = 0.007**). **Bottom left.** Velocity traces of **Top left and Bottom right.** Pooled peak saccade velocities.
4. Discussion

Our patients demonstrated 6 cardinal features of eye movement abnormality in superior oblique myokymia including: 1) involuntary intorsion and torsional oscillations; 2) episodic events lasting seconds; 3) worsening with infraduction and abduction—positions where the superior oblique is not activated; 4) overshooting of saccades on infraduction; 5) extorsion and diminished oscillations that were unmasked upon removal of a visual target, consistent with underlying weakness; and 6) improvement with membrane stabilizers used to treat neuropathic conditions. These features localized the lesion to the trochlear nerve, fascicle, or nucleus but not to the superior oblique muscle or neuromuscular junction.

The primary action of the superior oblique is intorsion, which explains the predominantly torsional oscillations in primary gaze. In eccentric gazes, eye movements worsened on infraduction and abduction—secondary and tertiary actions of the superior oblique, and improved with elevation and adduction—positions that did not require trochlear nerve action. Consistent with the idea that these oscillations are triggered by activation of the trochlear nerve, all patients exhibited a small contralateral head tilt regardless of alignment, as an ipsilateral head tilt triggers worse oscillopsia. Small ipsilateral hypertropia (2 patients) may be due to paresis, and hypotropia (3 patients), to overaction, as hypotropia correlated with worse symptoms and ocular motor findings. This relationship between nerve activity and ocular alignment is dynamically demonstrated in the unmasking experiment, where loss of the fixation target led to elevation and extorsion of the affected eye, features consistent with trochlear nerve paresis.

The trochlear nerve activity in SOM, which behaved simultaneously as a hyperactive and a weakened system, is compatible with the nerve injury-regeneration model suggested by electromyography studies and oculographic data.1,2,5,6,8 Although absent in our patients, history of head trauma, superior oblique palsy,1 microvascular compression,7,9 intracranial mass,10,11 and evidence of superior oblique atrophy seen on MRI12 have been associated with SOM. Microvascular compression has also been observed in hemifacial spasm (cranial nerve VII),13 which has been associated with SOM14 and trigeminal neuralgia (cranial nerve V)13.

Aberrant regeneration after partial nerve injury may result in neurons that lack appropriate supranuclear control.1,3,5,6,8 Electromyographic demonstration of rapid phasic firing of abnormal and prolonged motor units in the superior oblique muscle without corresponding inhibition in the inferior oblique muscle suggests altered membrane activation thresholds of the trochlear motor neurons. Consistent with this finding, membrane-stabilizing medications provide some relief for patients with SOM.1,3,14 Carbamazepine, phenytoin, propranolol, betaxolol, gabapentin, and clonazepam have been shown to be effective in 80% of patients initially, with sustained benefit in 45% of patients.15 If medical therapy is ineffective, extraocular muscle surgery can be considered including simultaneous superior oblique myotomy and inferior oblique myectomy, combined superior oblique tenectomy and inferior oblique myectomy, and the Harada-Ito procedure.17 In our study, all patients improved with stress reduction and treatment with membrane-stabilizing medications.

5. Conclusions

By examining and conducting eye movement recordings of five patients with SOM, we showed both weakness and activity-dependent hyperactivity of the trochlear motor unit, supporting a model of injury followed by aberrant regeneration.

Patient consent

Our study conforms to the World Medical Association Declaration of Helsinki (June 1964) and subsequent amendments. The research protocol was approved by the Stanford University Institutional Review Board, Stanford University School of Medicine,
Informed consent was sought and granted from all subjects.

Acknowledgements and disclosures

Funding

Y.J.L. is supported by the Career Award in Biomedical Sciences from the Burroughs Wellcome Foundation.

Conflict of interest

None of the authors has a financial or proprietary interest to disclose in relation to the content of this article.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship. Sumeer Thinda and Yi-Ren Chen contributed equally to this manuscript.

Acknowledgements

None.

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