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Vertical Oscillopsia: A Case of Superior Oblique Myokymia

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INITIAL PRESENTATION

Chief Complaint: "Shaking vision in my left eye"

History of Present Illness (HPI)

A 39-year-old male presents to our general ophthalmology clinic for a third opinion regarding intermittent episodes of objects bouncing up and down in his vision in his left eye. The episodes have been occurring for the past two months and last for several seconds to minutes. Bright lights, caffeine, and looking down to read seem to exacerbate these episodes. At times, these symptoms have made it difficult for him to work. He recalls having similar difficulties with his vision 3 years prior, but his symptoms resolved within a week after their onset. He denies double vision, redness, or pain with eye movement.

Past Ocular History

- Refractive error, contact lens wearer since age 14
- No eye trauma or surgeries

Past Medical History

- Guillain-Barré Syndrome (GBS) and subsequent pulmonary embolism, which required hospitalization for 4 months, 15 years prior to presentation
- Obstructive sleep apnea

Intraocular Pressure (IOP)

- OD: 12
- OS: 10

Pupils

- OD: 5 mm in dark → 4 mm in light, no afferent pupillary defect (RAPD)
- OS: 5 mm in dark → 3 mm in light, no RAPD

Confrontation visual fields: Full to counting fingers both eyes (OU)

Slit lamp exam

OU

- External/Eyelid: Meibomian gland dysfunction, no blepharospasm, no facial spasms
- Conjunctiva: Clear and quiet
- Cornea: Clear
- Anterior chamber: Deep and quiet
- Iris: Normal architecture
- Lens: Clear

Dilated fundus examination (DFE)

OU: Normal apart from intermittent vertical-torsional movements of fundus OS

DIAGNOSIS

Superior oblique myokymia

CLINICAL COURSE

Based on the patient's history of intermittent, brief episodes of vertical oscillopsia and the low-amplitude vertical-torsional movements of the left eye on examination, the patient was diagnosed with superior oblique myokymia. Since the patient had previously sought the opinion of two other eye care providers, he was particularly pleased to learn of a diagnosis and to hear that treatment options were available. He was started on carbamazepine 100 mg orally three times a day and scheduled to follow-up in the neuro-ophthalmology clinic.

He returned 1 month later with significant improvement of his symptoms. He experienced several side effects (drowsiness, dizziness, and upset stomach) while taking carbamazepine 100 mg three times a day, so he had reduced the dose. When he returned for his follow-up, he was taking 50 mg two times a day, which he felt provided 70% relief of symptoms. There was further discussion with the patient regarding other treatment options and he expressed interest in trying a topical agent. He was started on 1 drop of timolol in the left eye twice a day. He was told that if the timolol was effective, the carbamazepine could be tapered to an even lower dose or perhaps discontinued. An MRI of the brain with contrast and an MRA of the head were recommended to evaluate for an underlying structural lesion—most commonly, a small vascular loop compressing the left fourth nerve. The MRI and MRA were normal and did not show any vascular abnormalities. The patient was scheduled to return for routine follow-up in 4-6 months.

DISCUSSION

Superior oblique myokymia (SOM) is an uncommon disorder characterized by rapid, low-amplitude, high frequency contractions of the superior oblique muscle, which results in monocular vertical-torsional oscillopsia. Duane first described the disease in 1906 and he termed it a 'unilateral rotatory nystagmus' (1). In 1970, Hoyt and Keane were the first to use the term superior oblique myokymia after describing the clinical presentations of five patients (2). Affected patients are typically healthy, young to middle-aged adults without ocular or neurologic disease. Patients may report visual disturbances such as spontaneous image tilt, a fluttering or trembling sensation, and recurrent episodes of vertical-torsional oscillopsia, often described as "shaking," "shimmering," "vibrating," "jiggling," "dancing," or "jumping" (1, 2, 3, 4). The episodes are brief, only lasting a few seconds to minutes, and recur sporadically. A patient may experience multiple episodes in one day for several weeks and then have symptoms disappear suddenly. The symptoms may recur weeks, months, or even years later, with a different frequency and duration (2). The characteristic symptoms allow for the diagnosis to be strongly suspected based on the history alone. The pathognomonic eye movements can often be observed with mild to moderate magnification at the slit lamp, although it is relatively uncommon to see these movements in clinic. The movements can occasionally be elicited by having the patient gaze in the direction of action of the superior oblique muscle—down and in (3, 5, 6).

The pathogenesis of SOM remains uncertain, although several mechanisms have been proposed (2, 7, 8, 9). At present, the pathogenesis is thought to be similar to that of other paroxysmal cranial nerve disorders (such as trigeminal neuralgia and hemifacial spasm) and due to compression of the nerve by a vascular loop near the nerve root exit zone (10). Vascular compression is defined by the absence of a detectable layer of cerebrospinal fluid between the fourth nerve and an adjacent blood vessel (typically a branch of the superior cerebellar or posterior cerebral artery) most easily seen on thinly sliced (1-2 mm) MRI images (6, 10). SOM can occasionally be caused by a structural lesion (e.g., tumor) or brainstem demyelination (8, 10, 11, 12, 13, 14). Although an underlying structural lesion is not often identified, most neuro-ophthalmologists will recommend an MRI with and without contrast and, if available, an MR angiogram with contrast to evaluate for an underlying structural lesion and possible vascular compression of the fourth nerve (3).

Treatment

The vast majority of SOM cases follow a benign, relapsing and remitting course (2, 3, 4, 8, 15). In the setting of normal neuro-imaging, observation and reassurance may be appropriate for patients with mild or infrequent symptoms. For patients with persistent or bothersome symptoms, a variety of oral and topical medications may be considered (4). Carbamazepine provides symptomatic improvement in the majority of patients (5, 15). However, it is often poorly tolerated and has potentially severe adverse effects including leukopenia, acute renal failure, thromboembolism, and arrhythmias (4, 5, 7, 16). For this reason, some have proposed using gabapentin as a first line therapy, given the improved tolerability and safer side-effect profile (6, 17). Many other drugs, including oxcarbazepine, phenytoin, clonazepam, baclofen, oral and topical beta-blockers, mirtazapine, and memantine have been tried with varying degrees of success (3, 4, 5, 6, 7, 8, 16, 17, 18, 19). Botulinum toxin injection into the superior oblique muscle has also been proposed as a treatment, but provides only temporary relief and carries a risk of affecting other extraocular muscles (20).

Surgical intervention is reserved for patients with intolerable symptoms who fail to get an adequate response to medical management (5, 20). The first surgical treatments attempted were tenotomy or complete severance of the superior oblique tendon with subsequent recession (reattachment of the eye muscle at a different location to weaken its action) and myectomy—removal of a portion of the muscle belly of the ipsilateral inferior oblique to weaken its action (1, 2, 5, 21). Other studies report a benefit from tenectomy or partial severance of the superior oblique tendon followed by myectomy of the ipsilateral inferior oblique. While this approach is typically very successful in eliminating oscillopsia, it may produce an iatrogenic superior oblique palsy in about a third of patients. Prism correction may alleviate this complication (8, 21, 22, 23). The Harada Ito procedure has also been

used to treat SOM (24). In patients with intractable symptoms who have evidence of vascular compression of the fourth nerve on imaging, microvascular decompression of the fourth nerve could be considered as a last resort (25, 26, 27, 28).

Summary

A 39-year-old healthy male reported having episodic oscillopsia lasting for seconds to minutes for the previous two months. He recalled having similar episodes three years prior to presentation with spontaneous resolution. On slit-lamp examination, low amplitude and high frequency vertical-torsional movements were noted in the left eye, consistent with superior oblique myokymia. In most patients, superior oblique myokymia is a benign, relapsing and remitting condition. Neuroimaging is usually obtained to evaluate for an underlying structural lesion. Superior oblique myokymia often responds to oral medications, such as carbamazepine. Topical beta-blockers have also been used with varying degrees of success. Surgical treatments, such as strabismus surgery and microvascular decompression of the fourth nerve, are reserved for patients with severe and intractable symptoms.

EPIDEMIOLOGY

- Typically, young to middle-aged otherwise healthy adults
- Men and women appear to be equally affected
- May have a history of mild ocular, orbital, or head trauma

SIGNS

- Unilateral, intermittent, vertical-torsional eye movements that occur for seconds to minutes at a time
- Not always seen in clinic, but may be appreciated using magnification with a slit lamp or 20 D lens
- Movements classically precipitated by downward and inward movement of affected eye

SYMPTOMS	TREATMENT
<ul style="list-style-type: none"> • Recurrent brief episodes of vertical-torsional oscillopsia, often described as "shaking", "shimmering", "quivering", "vibrating", "jiggling", or "jumping" images • Episodic image tilt in one eye • Intermittent vertical or mixed vertical-torsional diplopia • Reported trigger factors and associations: stress, fatigue, alcohol, caffeine, nicotine, fluorescent lighting 	<p>Observation and reassurance</p> <p>Consider obtaining MRI brain and MRA head</p> <p>Medical management:</p> <ul style="list-style-type: none"> • Carbamazepine <ul style="list-style-type: none"> ◦ Start at 100 mg orally twice daily ◦ Titrate up to 200 mg orally three times daily, as tolerated • Gabapentin <ul style="list-style-type: none"> ◦ Starting at 100 mg orally daily or twice daily ◦ Effective dose range: 300-600 mg daily, but 600-900 mg may be required in some cases • Topical beta-blocker <ul style="list-style-type: none"> ◦ 1-2 drops daily in the affected eye • Others: Oxcarbazepine, phenytoin, clonazepam, baclofen, mirtazapine, and memantine <p>Surgical Intervention</p> <ul style="list-style-type: none"> • Strabismus surgery • Microvascular decompression of fourth nerve

Differential Diagnosis

- Monocular nystagmus
- Heimann-Bielschowsky phenomenon
- Voluntary nystagmus
- Blepharospasm

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Past Surgical History

- Cholecystectomy
- Motorcycle accident in 1994 with significant trauma to his leg and several orthopedic reconstructions of the left hip and leg

Medications: Fluticasone 50 mcg nasal spray

Allergies: None

Family History: Brother with Charcot-Marie-Tooth syndrome

Social History

- Significant smoking history; typically smoked 1-1.5 packs per day for 20-25 years, but at times, up to 3 packs per day; currently smoking 4-5 cigarettes per day
- No alcohol or illicit drug use

Review of Systems: Negative except as listed in HPI

OCULAR EXAMINATION

Visual Acuity

- Right eye (OD): 20/15
- Left eye (OS): 20/20-2

Ocular Motility: Full, no ocular misalignment on cross cover testing. No nystagmus. Intermittent, low-amplitude, vertical-torsional movements were observed at the slit lamp in the left eye. [Video 1]

from [University of Iowa Ophthalmology](#)

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