Grand Rounds

A 67-year-old woman with shaking of the world

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History

A 67-year-old woman presented with episodic instability of vision due to "shaking of the world" (oscillopsia) affecting only her right eye. It had been particularly noticeable when reading. On a number of occasions, she detected abnormal movement of her right eye when looking in a mirror. With time, the oscillopsia became less troublesome; specifically, it was smaller in amplitude and higher in frequency. She was otherwise well.

Examination

On slit-lamp biomicroscopy, there were fine torsional oscillations of the right eye only. Ophthalmic examination was otherwise unremarkable including extra-ocular motility. The remainder of the Neurologic examination was normal.

Ancillary Testing

Radiographic Studies

Brain MRI prior to referral was normal.

Treatment

As the symptoms had become less troublesome, no treatment was recommended; however, topical beta-blocker therapy was suggested if the symptoms became bothersome again.

Diff DX

The differential diagnosis of monocular oscillopsia or the sign of unilateral nystagmus includes the dissociated nystagmus of internuclear ophthalmoplegia (horizontal monocular), superior oblique myokymia (SOM), spasmus nutans (vertical monocular), and unilateral amblyopia or optic neuropathy (vertical or horizontal).

Diagnosis

In 1970, Hoyt and Keane reported five patients with intermittent monocular oscillopsia and binocular vertical diplopia in whom unilateral repetitive torsional eye movements were seen on slit-lamp examination.¹ They hypothesized that fourth nucleus inhibition was the cause of what they termed superior oblique myokymia (SOM). The earliest similar case was reported as a unilateral rotary nystagmus by Duane in 1906.²

SOM can be readily diagnosed on the typical history and clinical findings. Repetitive contraction of the superior oblique muscle gives rise to high-frequency, low amplitude, tremor with torsional and/or vertical monocular eye movements, which may be exacerbated by ipsilateral head tilt or downward and inward movement of the affected eye, such as for reading. The presence of torsional oscillopsia and/or nystagmus seen in this case is fairly specific for superior oblique myokymia. Right SOM is reported to be more common than left.

The diagnosis in this case is essentially a clinical one, with the vast majority of cases either having no identifiable etiology or being due to fourth-nerve compression by an aberrant artery. Occasionally, these findings have been reported with posterior fossa tumors, dural arteriovenous fistulas, adrenoleucodystrophy, as well as after fourth nerve palsies, brainstem strokes and head trauma.

High resolution MRI has confirmed Bringewald's hypothesis; that is, SOM may result from microvascular compression of the root exit zone of the fourth cranial nerve on the dorsal surface of the midbrain. Similar compression of the fifth, seventh, and ninth cranial nerves is also known to occur in trigeminal neuralgia, hemifacial spasm and glossopharyngeal neuralgia, respectively.^{3,4} It is important to distinguish between

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compression at the root exit zone, which extends up to 1.2mm from the point of exit from the brainstem, and compression of the more distal (cisternal) segment of the fourth nerve. Compression of the distal segment occurs in 60–90% of individuals and is not relevant to the pathogenesis of SOM.⁵ Resolution of SOM following surgical microvascular decompression has been reported.^{6,7}

Conservative treatment is suitable for some patients. Systemic and topical beta-blockers have been reported to be beneficial, the latter presumably due to a direct stabilization of muscle cell membranes.^{8,9} Thus, topical timolol eyedrops are a reasonable first line treatment in patients with troublesome symptoms and no contraindications. Traditionally, carbamazepine has been the preferred oral agent, but other agents besides beta-blockers, such as gabapentin or baclofen also have been reported to be beneficial.¹⁰ More recently, substantial improvement in symptoms with memantine treatment was reported in one patient who had suffered side-effects with gabapentin.¹¹ If symptoms are precipitated by a particular gaze position, prisms may be tried. In cases resistant to medical treatment, superior oblique tenectomy has been advocated, but usually needs to be combined with inferior oblique muscle weakening to compensate for the induced superior oblique palsy. The role of microvascular decompression has yet to be established. There is a tendency for symptoms to spontaneously improve in some cases, therefore conservative follow-up is recommended before any form of surgical therapy is considered.

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