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CASE REPORTS

SUPERIOR OBLIQUE MYOKYMIA - A CASE STUDY

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Abstract

Superior Oblique Myokymia is a rare spasmodic contraction of the superior oblique muscle giving rise to monocular vertical and rotary microtremor and accompanied by torsional shimmering oscillopsia. A fifty-five year old woman presented with a long history of right ocular discomfort and a lack of synchronisation between the eye movements of the two eyes without diplopia. This was diagnosed as being due to right superior oblique myokymia. The unusual clinical presentation of this condition is discussed and treatment options outlined.

Key words: *vertical and rotary microtremor, oscillopsia.*

INTRODUCTION

Superior oblique myokymia is an unusual disorder of ocular motility[1]. It was first described by Duane in 1906 (cited in Walker, 1985) as a *unilateral rotary nystagmus*[2], and has also been described by Hoyt and Keane[3] as *intermittent uniocular microtremor*. It is characterised by recurrent episodes of monocular vertical and rotary microtremor, and accompanied by torsional shimmering oscillopsia. The affected eye intorts for seconds during each episode[4]. In some cases the patient is able to define the precise ocular movement which precipitates the attack[2], however for many patients episodes may occur without a change in ocular position[4].

Symptoms may vary between patients, and visual symptoms are often accompanied by physical ocular sensations. The visual symptoms can consist of intermittent oscillopsia or intermittent torsional or vertical diplopia[5]. Patients may also describe ocular sensations of twitching, jumping or moving, and a pulling or drawing sensation may accompany or precede an attack[5].

This condition has no known associations with other neurologic disease. Disordered activation and/or inhibition at the level of the 4th cranial nerve nucleus are proposed as the mechanisms of production of this unique ocular condition[4]. Hoyt and Keane[3] have suggested that this distinctive neuro-ophthalmic condition represents a self limiting motor neuron disorder.

CASE STUDY

A 55 year old woman, J.F., presented with a history of right ocular discomfort which she had experienced for many years. She stated that on an intermittent basis the movements of the right eye did not synchronise with those of the left eye. On further questioning regarding other symptoms such as diplopia, she stated that she experienced blurred vision like a heat haze during the episodes but not diplopia.

J.F. felt that the condition was aggravated by

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stress, driving and bright lights, and alleviated by changing head posture, particularly by adopting a chin down head posture. During history taking she commented that she sometimes thought she was going mad or imagining the symptoms, and felt very frustrated by the fact that previous eye tests had failed to disclose the nature of her problem.

On examination, visual acuity was 6/5, N5 right and left with correction. The Lang's test demonstrated stereopsis to 200 seconds of arc (with and without use of a chin down head posture). On cover testing at 6 meters J.F. was orthophoric initially, but then an intermittent microtorsional tremor of the right eye was noted. At 1/3 metre she had an exophoria initially, with an intermittent microtorsional tremor demonstrable after a short time of cover testing. Each episode of the tremor lasted only a few seconds and J.F. was able to report when it was about to happen because the right eye 'felt different'.

On testing ocular movements J.F. was found to have a downshoot of the right eye on laevoversion followed a few seconds later by a recovering updrifting movement. These unusual eye movements did not occur on every occasion of laevoversion but when they did occur they were quite obvious. Overaction of the right superior oblique and underaction of the left inferior rectus could only be demonstrated occasionally. On dextroversion and dextroelevation there was a definite torsional movement of both eyes (right more than left). Convergence was full with no microtremor noticeable during testing of this function.

J.F. was diagnosed as having right superior oblique myokymia and when this was discussed with her she was very pleased that her unusual symptoms were actually due to a clinical condition rather than to her imagination. She was commenced on Carbamazepine (Tegretol) 100mg three times a day, to be reduced to twice daily over a three week period. This provided immediate relief from her symptoms and she has continued with this regime for the past 12 months without the need for any increase in strength or frequency of the prescribed drug.

DISCUSSION

Superior oblique myokymia has such distinctive signs and symptoms that the diagnosis can be made without subjecting the patient to a whole battery of unnecessary neurodiagnostic tests[4]. Slit lamp examination is sometimes needed to see the fine tremorous ocular movements of this condition[1], otherwise the patient may be diagnosed incorrectly as having a functional problem.

Management of this condition is by drug therapy or surgery. Carbamazepine is usually the drug of choice[1], and it provides dramatic relief of symptoms for most patients. Some patients remain symptom free following its discontinuation but others require a continuing low dosage to remain symptom free[2]. However it is not suitable for all patients because for some it has no effect and for others may cause side-effects. In these cases surgery may be considered[1]. Intrasheath tenotomy of the superior oblique muscle and recession of the insertion of the inferior oblique of the involved eye[3] has proved useful in those cases which are unresponsive to drug therapy.

CONCLUSION

Orthoptists need to be aware that while rare, superior oblique myokymia does exist, and may pass unrecognised unless credence is given to the patient's symptoms and sufficient time is spent during the ocular examination to allow the condition to manifest itself. It is important for the orthoptist to reassure these patients that their unusual symptoms are due to a condition which is benign and self limiting.

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