Ocular Neuromyotonia and Myasthenia Gravis

To the Editors:

Myasthenia gravis presents with only ocular symptoms (diplopia, ptosis, and extraocular movement limitation) in approximately 20% of patients.\textsuperscript{1,2} Ocular myasthenia gravis is to be suspected in patients with intermittent diplopia with ophthalmoplegia that does not correspond to a single ocular motor nerve paresis. However, diplopia in only eccentric sustained gaze in myasthenia gravis is uncommon.

A 32-year-old man with a 20-day history of intermittent diplopia while looking to the right side was seen at our clinic. He was referred to us because of right abducens nerve palsy and had no systemic disease or ocular history. Brain magnetic resonance imaging and thyroid function tests were unremarkable. On examination, his visual acuity was 20/20 in each eye and his intraocular pressure was 12 (right eye) and 13 (left eye) mm Hg. Both anterior segment and fundus examinations were normal. Ocular motility examination revealed orthotropia in the primary and secondary positions. Ductions and versions were normal (Figure 1). During the quiescent periods, the patient had no diplopia. Attacks were precipitated especially by sustaining right gaze for more than 10 seconds. During an attack, the left eye showed 25 prism diopters of esotropia in right gaze and 12 prism diopters of esotropia in the primary position and eyelid retraction (Figure 1, Video A, available in the online version of this article). Pupillary function was normal. Over time, the diplopia gradually disappeared. We treated the patient with carbamazepine based on idiopathic ocular neuromyotonia.

After 1 week, the patient developed a headache and had fatigable weakness of the left eyelid. The acetylcholine receptor Ab was 10.699 nmol/L and a mass in the anterior mediastium was noted on a chest computerized tomography scan. Positive findings in a Jolly test were observed at 2, 3, and 5 Hz. A thymectomy was done and biopsy results showed thymoma. The symptoms of ptosis and diplopia after sustained gaze disappeared after 4 months of treatment with pyridostigmine medication.

Spontaneous and transient diplopia after sustained gaze to the right or left side is a characteristic feature of ocular neuromyotonia. Ocular neuromyotonia is caused by an autoimmune disease or neoplasm and myasthenia gravis, thyrotoxicosis, systemic sclerosis, thymoma, and small cell lung cancer are known to be associated with it.\textsuperscript{3,4} Although the exact mechanism of ocular neuromyotonia is unknown, voluntary potentials of the ocular motor nerve with an unstable cell membrane and autoantibodies have been suggested.\textsuperscript{5} Our patient developed ocular neuromyotonia in the left eye after experiencing a sustained right gaze, from a previously healthy state, and was diagnosed as having myasthenia gravis. The paroxysmal esotropia and eyelid retraction of the left eye that occurred in this patient was typical of ocular neuromyotonia affecting muscles innervated by the oculomotor nerve. Synkinetic movement with levator palpebrae is usually related to contraction causing eyelid retraction.\textsuperscript{4} Therefore, this case of ocular neuromyotonia was thought to be caused by autoantibodies for myasthenia gravis.

It is important to consider the possibility of ocular myasthenia gravis in patients who initially present with diplopia with eccentric gaze only and no ptosis. Ocular myasthenia gravis should be considered in patients with ocular neuromyotonia.

REFERENCES

2. Rush J, Shafrin F. Ocular myasthenia presenting as superior


JunWon Jang, MD
Moohwan Chang, MD, PhD
Sungeun Kyung, MD, PhD
Cheonan-City, South Korea

The authors have no financial or proprietary interest in the materials presented herein.

doi: 10.3928/01913913-20150427-09