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Diagnosis of Tensilon-Negative Ocular Myasthenia Gravis By Daily Selfie

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Abstract: The initial symptoms of myasthenia gravis are usually ptosis and diplopia. The diagnosis is often confirmed by testing for anti-acetylcholine receptor antibodies or by observing the effects of intravenous edrophonium (Tensilon) injection. However, these standard tests may be negative in patients with isolated ocular findings. We present the case of an 83-year-old woman with negative serologic and Tensilon testing. She was asked to photograph herself daily. The resulting sequence of daily selfies captured striking fluctuations in her ocular alignment and ptosis. Daily selfies may be a useful strategy for confirming the clinical diagnosis of ocular myasthenia gravis.

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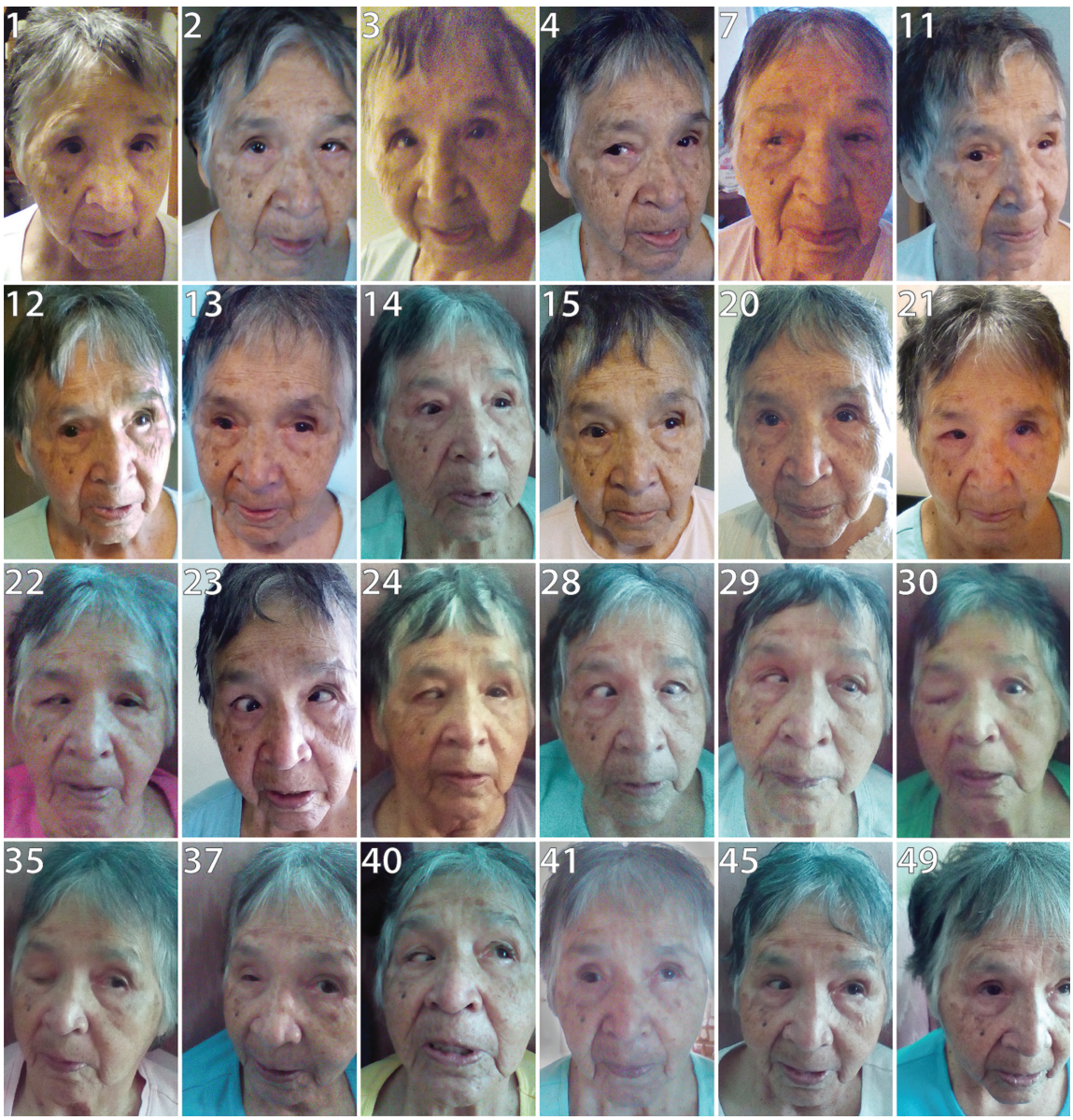

FIG. 1. Selfies to capture serial changes in ptosis and eye misalignment. The patient remembered to take a selfie on 24 of 49 mornings (numbers indicate the day since last clinic exam). Although her ocular deviation was unclear on some days, owing to uncertainty regarding her gaze angle and the camera position, the images provide objective documentation of fluctuating ptosis and eye misalignment. Her initial exotropia was replaced by esotropia, that became marked on days 23–29, followed by subsequent return of exotropia on day 41. There was also a right ptosis that became complete on days 30–35.

An 83-year-old woman, living alone, was evaluated for a 6-month history of intermittent ptosis and diplopia. On initial examination, her extraocular eye movements were full. However, there was ptosis of her right upper eyelid and 20 prism-diopters of right exotropia. The ptosis became more pronounced after prolonged upgaze. Magnetic resonance imaging of the brain, acetylcholine receptor antibody levels, and a Tensilon test were negative. On examination 2 weeks later, her ptosis seemed slightly improved but the right exotropia was still present.

Although testing had been negative, we still suspected the diagnosis of ocular myasthenia gravis. The finding of ptosis that worsened after sustained globe elevation was typical. However, we still lacked evidence for fluctuation in her ocular deviation. The patient was asked to photograph herself daily with her cell phone, to track her eye alignment and eyelid position. At a follow-up appointment 7 weeks later, review of the images confirmed that her right ptosis was variable, and that her ocular deviation had changed, even switching from exotropia to esotropia (Fig. 1). After no response to treatment with pyridostigmine, her symptoms improved after a course of oral prednisone.

The diagnosis of ocular myasthenia gravis can be elusive. Ptosis and diplopia are early symptoms in the majority of patients with myasthenia gravis. Half of these patients will develop generalized muscle weakness, shortening the list of diagnostic possibilities substantially (1). Isolated ocular symptoms can be a challenge to interpret, owing to the fact that abnormal ocular motor function is a feature of many disease processes including cranial nerve palsies, stroke, demyelinating disease, and mitochondrial cytopathies.

Physicians use serologic tests, brain imaging, and electromyography to investigate ptosis and diplopia. Yet, these tests can overlook patients with pure ocular myasthenia gravis (2). Standard laboratory assays for acetylcholine receptor antibody levels are often negative (3,4). Tensilon testing, which some physicians are reluctant to perform, lacks sufficient sensitivity to identify many patients (5). Additional testing such as antibodies against muscle-specific kinase and electromyography may also be normal, particularly in patients with only ocular symptoms (2).

In this situation, physicians must rely on clinical findings. Observation of fluctuations in eyelid position, ocular motility, or eye alignment made during serial clinic visits provide strong circumstantial evidence for ocular myasthenia gravis. The problem is that serial examinations provide only a glimpse of daily events. To fill in the gaps, one may query patients about changes in ptosis or diplopia during the course of each day. However, patients' accounts can be difficult to interpret, especially as diplopia is an all-or-none phenomenon. Changes in ocular movement or misalignment can be difficult for patients to perceive or to describe with reliability.

The first photographic self-portrait was taken by Wheatstone in 1840. It was a daguerreotype that required him to remain motionless for several minutes (4). Today, photographs can be taken easily and quickly using a variety of electronic devices available to nearly everyone. Daily autophotography can provide an objective record to support or refute the diagnosis of ocular myasthenia gravis, essentially by collapsing dozens of clinic observations into a single follow-up visit. Scrutiny of serial images may reveal changes in eye position and ptosis. When other testing has failed, we advocate daily selfies to provide evidence in patients with suspected ocular myasthenia gravis.

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STATEMENT OF AUTHORSHIP
Category 1: a. Examination and diagnosis of patient: E. L. Guterman; b. Drafting of manuscript: E. L. Guterman and J. V. Botelho; c. Drafting and critical revision of manuscript for important intellectual content: J. C. Horton.

REFERENCES

1. **Vincent A,** McConville J, Farrugia ME, Newsom-Davis J. Seronegative myasthenia gravis. Semin Neurol. 2004;24:125–133.
2. **Vernino S**. Unraveling the enigma of seronegative myasthenia gravis. JAMA Neurol. 2015;72:630–631.
3. **Peeler AE,** De Lott LB, Nagia L, Lemos J, Eggenberger ER, Cornblath WT. Clinical utility of acetylcholine receptor antibody testing in ocular myasthenia gravis. JAMA Neurol. 2015;72:1170–1174.
4. **Wade NJ**. The first scientific "selfie"? Perception. 2014;43:1141–1144.
5. **Kupersmith MJ,** Latkany R, Homel P. Development of generalized disease at 2 years in patients with ocular myasthenia gravis. Arch Neurol. 2003;60:243–248.