

Case Report — Ocular Myasthenia Gravis

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Abstract

Myasthenia gravis is a chronic disease producing muscle weakness. Ocular muscle involvement occurs in a large percentage of myasthenic patients. A case of myasthenia gravis with initial symptoms of diplopia and extraocular muscle involvement is presented. The ocular signs of myasthenia gravis and diagnostic procedures are emphasized.

Abrégé

Myasthénie Gravis est une maladie chronique sérieuse caractérisée par une faiblesse musculaire affectant très souvent les muscles extrinsèques de l'oeil, ce qui entraîne diplopie et ptosis. Ce travail décrit les symptômes oculaires et les tests diagnostiques pour cette condition.

Introduction

Myasthenia gravis is a chronic disease characterized by weakness of voluntary muscles following their use. It affects primarily the muscles of facial expression, mastication, swallowing, ocular motility and the eyelids.^{1a} Muscle strength returns rapidly after a period of rest. Smooth muscle and cardiac muscle are not affected.² There is a failure of neuromuscular transmission, possibly a depletion of acetylcholine or an increased breakdown of acetylcholine due to overactivity of cholinesterase. About 10% of the patients exhibit thymoma and 5% have dysthyroidism.^{3a} Ocular muscle involvement occurs in 90% of myasthenics and accounts for the initial complaint in about 75% of the cases.

Approximately 20% of myasthe-

nics exhibit purely ocular involvement and extraocular muscles can be involved individually or in any combination.^{3b}

History and Background

Patient E.S. was first examined by the author in 1975 for routine ocular assessment. He was 53 years of age at that time. He was using spectacles for reading only and had no visual complaints. General health history and medication use was negative.

At that time he was found to be a low hyperopic astigmat and presbyope. The refractive status was O.D. +0.75 -0.50 x 030, 20/20; O.S. +0.75 -0.25 x 120, 20/20; add +2.25, 0.37M O.U. This was not significantly different from his habitual correction. Since he preferred not to wear bifocals and unaided vision was O.D. 20/25, O.S. 20/25, O.U. 20/20, he was advised to retain his present reading glasses.

There were no abnormalities of ocular health and ocular motility was normal. Intraocular pressure and visual fields were unremarkable.

E.S. was examined annually for the next two years during which no changes were found. In 1978 there was a slight change in refraction and the following prescription was written: O.D. +1.25 -0.25 x 060, 20/20; O.S. +1.25 -0.50 x 105, 20/20; add +2.25 O.U. Since unaided visual acuities were now O.D. 20/60 and O.S. 20/30, bifocals were advised.

In 1979, E.S. was seen on an emergency basis. He was 57 years old at this time. He reported vertical diplopia on right gaze which had been noticed sporadically for the previous ten days. It had become more troublesome on the day prior to the consultation.

Clinical Findings

No change in refraction or visual acuities was noted. Ophthalmoscopy

and external health examinations were unremarkable. Pupils responded normally and there was a negative Marcus-Gunn sign. The three-step method for evaluating ocular deviations revealed the following:

primary position

12 prism dioptres right hyperdeviation

gaze left

ortho (i.e. decrease in deviation)

gaze right

20 prism dioptres right hyper (i.e. increase in deviation)

head tilt left

hyperdeviation decreased

head tilt right

hyperdeviation increased

Diagnosis

Analysis of the three-step method would implicate the left inferior oblique as the muscle primarily involved. Telephone consultation with a neuro-ophthalmologist resulted in immediate referral as this type of deviation can be indicative of an ischemic attack of the vertebrobasilar vascular supply or myasthenia gravis.

During the ophthalmological assessment a peculiar twitching of the right upper lid was noted. This was suspected to be Cogan's lid "twitch" sign which is elicited by having the patient rapidly redirect his gaze from downward to the primary position. If the lid appears to twitch upward and then settle back to its original position, myasthenia gravis is suspected.^{3c}

A Lancaster red-green test was conducted with Tensilon injected intravenously. This resulted in a decrease in the ocular deviation. Since the Tensilon test is considered diagnostic for myasthenia gravis the patient was referred to a neurologist for definitive diagnosis and treatment.^{1b}

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to the easy way, — lots of money; and a few believe that the world owes them a living: "I'm a big Doctor, and I'm a university graduate." But I know that they're quality people, and that if it came to a fight, they would fight very well. They would be fighting on a more sophisticated level than we were able to do, because they are better educated. We all seem to forget, even though it may seem a nasty kind of sociological thing to say, current graduates have brothers who are lawyers, aunts who are physicians, uncles who are judges and, on the average, they come from a higher social stratum. Those

connections are valuable and give current graduates a better chance to deal with opposition. I'm not at all pessimistic on that score.

The other thing I would like to mention along the same line relates to inspiration and dedication. I think it was Hitler who said that if you had a hundred people who were absolutely committed to a cause, you could do anything. What happened at the School was that, for 15 years or so, we had five or six people, who worked like beavers to make this place go. We met almost every day, and almost every night, and we worked weekends. There was no-

thing that anybody wouldn't do for the good of the school, in spite of personal problems, age, needs or wants. That dedication is bound to taper off in time, and it was successful because this small group of single-minded people agreed on the goal. They brought different kinds of skills, but a common aim: the enhancement of optometry. A small group of people like that, with the determination and the commitment and the will to hang together for a cause, constitutes a very powerful force.

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Management

The diagnosis of ocular myasthenia gravis was confirmed by the neurologist. Treatment was undertaken using Mestinon. After two months of treatment the diplopia cleared completely. Medication was discontinued and there was no recurrence of symptoms.

The final diagnosis was given as ocular myasthenia gravis currently in remission.

Followup

E.S. remained free of symptoms for one year. Treatment with Mestinon was resumed when occasionally diplopia reappeared, however it was

not as severe as during the initial occurrence. He is currently controlled with medication.

Discussion

Myasthenia gravis is a condition which often presents initially with ocular signs. Ptosis, diplopia and lid twitches are among the classical early signs of the disease. There is no affect on pupillary reflexes, visual fields or visual acuities. The patient described above is unusual in that his condition has remained purely ocular; more often there are varying degrees of systemic involvement. Periods of remission are common as was the case with E.S.

The patient presented with recent

onset diplopia resulting from a noncomitant deviation. This is always a serious symptom as it may be caused by intracranial pathology or neurological disease. Prompt medical referral is always indicated in such cases.

References

1. Walsh, Thomas J. "Neuro-Ophthalmology: Clinical Signs and Symptoms" Lea and Febiger, Philadelphia, 1978 (a) p. 86. (b) p. 82.
2. Cecil, R.L. and Loeb, R.F., "A Textbook of Medicine" ed. 10. W.B. Saunders Co., Philadelphia, 1959, p. 1474.
3. Glaser, J.S. "Neuro-Ophthalmology" Medical Dept. Harper and Row, Hagerstown, Maryland, 1978 (a) p. 269. (b) p. 269. (c) p. 38.

*"The world is moving so fast these days that the man who says
it can't be done is generally interrupted by someone doing it."*

— Elbert Hubbard

*Have A
Happy Day!*

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