Introduction

Myasthenia Gravis (MG) is a chronic disease of voluntary muscles that can be life threatening. Auto-antibodies act against the acetylcholine receptors at the post-synaptic neuromuscular junction. MG affects roughly 20,000,000 in the United States. The hallmark of MG is variability and fatigability of symptoms. This is often seen in a diurnal pattern where the symptoms are better in the morning and worse through the day. Symptoms can also vary due to factors such as stress, pregnancy, temperature, emotional stressors, surgery, and viral illness.1,2 Signs and symptoms of MG include ptosis, double vision, blurry vision, slurred speech, difficulty breathing, difficulty swallowing or chewing, extreme weakness, and chronic muscle fatigue. The natural course of this disease can eventually affect the respiratory muscles, which may result in death. The prevalence of MG varies due to factors such as stress, pregnancy, hypothyroidism, and genetic factors. Auto-antibodies act against the acetylcholine receptors at the post-synaptic neuromuscular junction.

Diagnosis

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Case Description

63 year-old female with a PMHx of DM and HTN presents with blurry vision and a drooping eyelid for one week. Patient states that the droopiness is better in the mornings and gradually gets worse throughout the day. Patient also reports that a similar episode of right drooping eyelid occurred five years ago, but did not seek medical attention. She says it resolved with warm compresses and artificial tears after one week. Patient denies any shortness of breath, proximal arm or leg weakness, difficulty swallowing, or difficulty breathing. Her only medications are Metformin, Lisinopril, and HCTZ.

Examination:

BCVA: OD 20/70-2, OS 20/50-2
Pupils: 3mm OU, PERRL, No RAPD
EOM: Full, Ortho OU.
CVF: Full OU, right upper lid had to be held open
JVP: 14/14 mmHg by Goldmann tonometry
Lids: Ptosis OU, OD>OS
Palpebral Fissure Width: OD 2mm, OS 10mm
MRD: OD 0mm, OS 6mm
Levator Function: OD15mm, OS 15mm

Lab Results:

Acetylcholine Receptor Antibodies: 30.80 H
Blocking Antibodies: 28.40 H

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3rd Nerve Palsy
CPEO
Thyroid Eye Disease
Myasthenia Gravis

Oculopharyngeal Dystrophy
Levator Dehiscence
Intracranial mass lesion
Myotonic Dystrophy

Miller-Fischer Variant of Guillain-Barré
Eaton-Lambert Myasthenic Syndrome

Some patients with presumed ocular MG may actually have subclinical generalized MG. Not all symptoms are clinically evident or reported by the patient. One study showed that 50% of ocular MG patients had a positive skeletal muscle biopsy from outside the ocular region.2,3

Treatment of MG should be tailored to the individual patient. Cholinesterase inhibitors, immunosuppressive therapies, or steroid therapy may be used. Ocular myasthenia does not respond as well to immunomodulatory therapy as generalized MG does. However, it is reported in the literature that ocular MG sometimes responds to oral prednisone therapy.5,6 When imaging reveals a thymoma, surgical removal is indicated, though performing a thymectomy on purely ocular MG is less likely to be therapeutic.5 This patient was referred to a neurologist. He started the patient on Mestinon. Three months after the first visit, the neurologist had decreased her dose of Mestinon secondary to GI upset. The patient also reports that her eyelid droop was better, but not resolved. She will continue to meet with her two-weekly follow-ups and we will continue to monitor her closely to determine response to treatment and stability of lid position. She may still require repair of the levator dehiscence in the future.

Here, our patient is over the age of 50, female, and has high antibody titer. These are all factors that point towards generalized MG versus ocular MG. It is also uncertain whether her previous episode of ptosis was actually a symptom of MG or an unrelated cause, as she did not seek medical attention and it resolved with warm (as opposed to cold) compresses. Thus, we can be sure if her symptoms started five years ago, or one week ago. Follow-up is essential for this patient as her case is statistically more likely to progress to generalized disease.

References