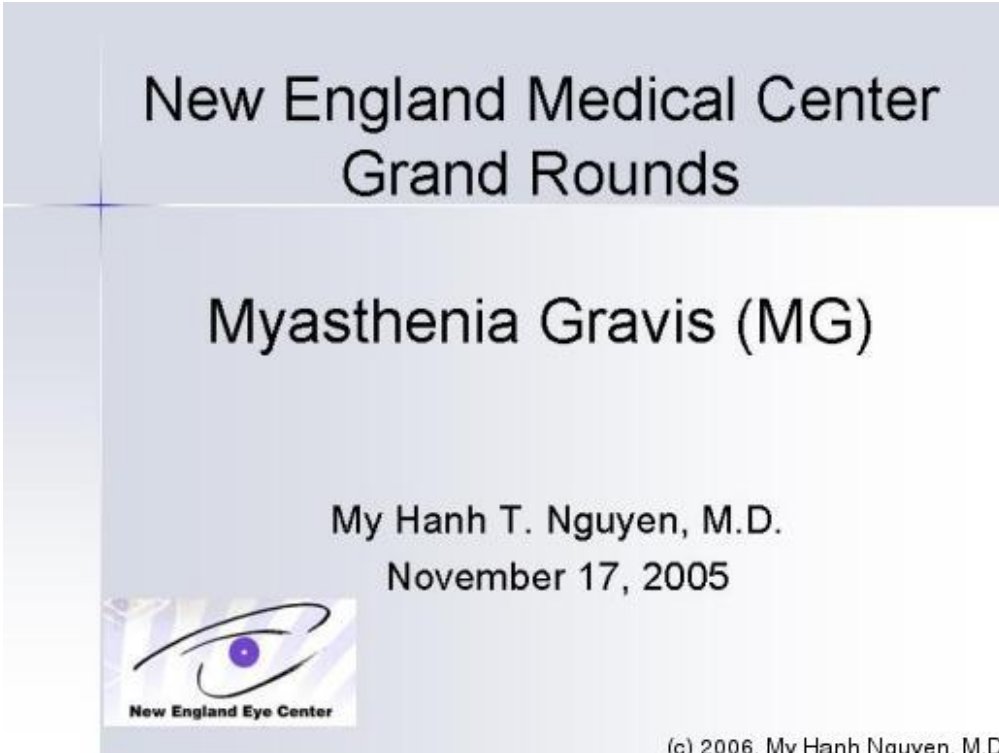



1. New England Medical Center Grand Rounds - Myasthenia Gravis ...



New England Medical Center
Grand Rounds


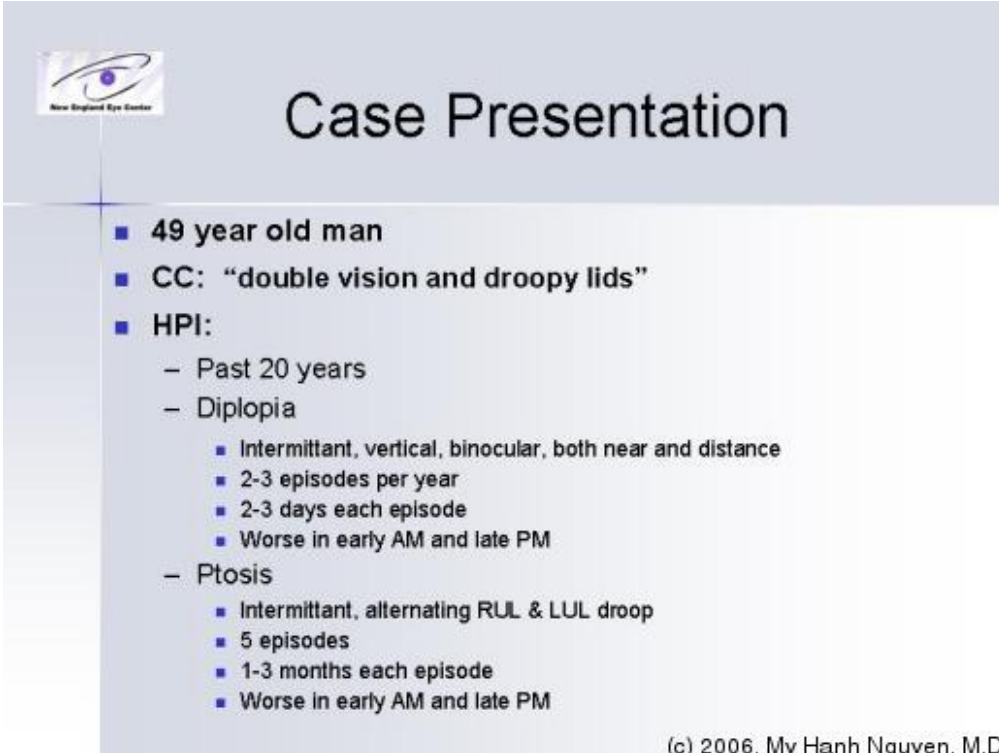
Myasthenia Gravis (MG)

My Hanh T. Nguyen, M.D.
November 17, 2005



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2. Case Presentation




Case Presentation

- 49 year old man
- CC: “double vision and droopy lids”
- HPI:
 - Past 20 years
 - Diplopia
 - Intermittant, vertical, binocular, both near and distance
 - 2-3 episodes per year
 - 2-3 days each episode
 - Worse in early AM and late PM
 - Ptosis
 - Intermittant, alternating RUL & LUL droop
 - 5 episodes
 - 1-3 months each episode
 - Worse in early AM and late PM

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3.

Case Presentation




Case Presentation

- **POHx:** No surgeries, trauma, or infections OU
- **PMHx:** MVA with LOC 30 years ago
- **Medications:** ASA
- **Allergies:** NKDA
- **Fam Hx:**
 - Glaucoma
 - DM
- **Soc Hx:**
 - Mechanic
 - Denies tobacco & ETOH use

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4.

Office Examination




Office Examination

- BCVA: 20/15 OD, 20/15 OS
- T: 15 OU
- Pupils: 5 mm → 3 mm OU, no RAPD

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5. External Exam



External Exam

	OD	OS
■ IPF	9	11
■ MRD	3	5
■ LF	14	16





Image courtesy of New England Eye Center, 2005.

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6. Office Examination



Office Examination

SLE:


LLL:	wnl OU
C/S:	white & quiet OU
K:	clear OU
A/C:	deep & quiet OU
I:	round, flat OU
L:	trace NS OU
V:	no cells OU

DFE: M/V/P normal OU

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7.

Differential Diagnosis?




Differential Diagnosis?

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8.

Differential Diagnosis




Differential Diagnosis

- Myasthenia gravis
- Third nerve palsy
 - Multiple sclerosis
 - Aneurysm
 - Ischemic vasculopathy
 - Pituitary adenoma and apoplexy
 - Other neoplasms
- Ophthalmoplegic migraines
- Viral illness
- Internuclear ophthalmoplegia
 - Multiple sclerosis
 - Brainstem ischemia
 - Brainstem neoplasms

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9. What would you do next?



What would you do next?

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10. Myasthenia Gravis - In the Office




Myasthenia Gravis In the Office

- Edrophonium chloride
 - Competitive inhibitor of acetylcholinesterase
 - 2 mg IV, additional 8 mg if the first does has no effect
 - Side effects
 - Lacrimation, diaphoresis, abdominal cramping, nausea, salivation, vomiting, diarrhea, urination, bradycardia, respiratory arrest, syncopal episodes
 - 0.4 - 0.6 mg IV atropine co-administered or available
 - False positives rare
 - Negative does not exclude diagnosis
- Neostigmine methylsulfate
- Sleep test
- Ice-pack test

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11.

Myasthenia Gravis



Myasthenia Gravis

- Neuromuscular disease
- Autoimmune mechanism: antibodies block acetylcholine receptors
- Prevalence: 85 - 125 per million
- Incidence: 2 - 4 million per year
- Age of onset:
 - 20 - 40 (women)
 - 60 - 80 (men = women)
- Females 3:2
- Symptoms:
 - Muscle weakness
 - Marked fatigability of skeletal muscles
 - Improves with rest

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12.

Myasthenia Gravis




Myasthenia Gravis

- Involved muscles
 - Eyes
 - Oropharynx (dysphagia, hoarseness, dysarthria)
 - Neck extensors (pain, weakness)
 - Limbs (grip, proximal legs)
 - Trunk (dyspnea, respiratory failure)
- Spontaneous remissions
- Relapses
 - 90% in early stages
 - "Symptom free": months to years
- Hallmark: fluctuation and fatigability

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13. Myasthenia Gravis - Ocular Involvement




Myasthenia Gravis Ocular Involvement

- Frequency
 - 50 - 75% of initial presentation: ocular myasthenia gravis
 - 90% of cases
- Findings
 - Ptosis:
 - Unilateral or bilateral
 - Worsens with prolonged upgaze
 - Diplopia: simulate any cranial nerve paresis
 - Supranuclear motility disturbances (pseudo-INO, gaze palsies)
 - Isolated muscle palsy
 - Total ophthalmoplegia
 - Cogan's lid twitch
 - Orbicularis oculi invariably weak
- Ocular MG → systemic MG
 - 80% within first year
 - 90% within three years

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14. Myasthenia Gravis - Management



Myasthenia Gravis Management

- Neurology
- Anti-Acetylcholine receptor antibodies
 - Binding antibodies
 - 90% of generalized MG
 - 50-70% of ocular MG
 - Modulating antibodies: as frequent as binding antibodies
 - Blocking antibodies rare (1%) without binding antibodies
- MRI or CT
 - Thymic disease 75%
 - Thymic hyperplasia 85%
 - Thymomas 10%
- Thyroid function test (5%)
- ANA: systemic lupus erythematosus
- Rheumatoid factor: rheumatoid arthritis

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15. Myasthenia Gravis - Treatment Options




Myasthenia Gravis Treatment Options

- Long acting anticholinesterase
 - Pyridostigmine bromide
 - Neostigmine bromide
- Immunosuppressives
 - Systemic steroids
 - Azathioprine
 - Cyclosporine A
 - Methotrexate
 - Cyclophosphamide
- Surgical thymectomy
- Plasmapheresis
- Intravenous immunoglobulin

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16. Case Summary




Case Summary

- 49 year old man with diplopia and ptosis
- Intermittant, fluctuating, worsens with fatigue
- Positive Edrophonium chloride test
- Diagnosis: myasthenia gravis
- Follow up:
 - Neurology
 - TSH wnl
 - ANA wnl
 - Acetylcholine antibodies
 - AchR binding AB: high
 - AchR blocking AB: wnl
 - AchR modulating AB: wnl

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17.

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