

Plethora of Cranial Nerve Palsies: A Problematic Presentation of Myasthenia Gravis

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Introduction

- Myasthenia gravis is an autoimmune disease of the neuromuscular junction, commonly involving pathogenic antibodies that bind to different components of the neuromuscular junction (1).
- Initially myasthenia was thought to primarily affect young females between 20-40 years of age, recently have seen increasing frequency of late onset myasthenia gravis. (2)
- Diagnosis of myasthenia gravis can be underdiagnosed in this older population as symptoms can mimic other motor neuron diseases or even brainstem strokes.
- Common presentation of MG include hoarseness, and ptosis; more recently have seeing an increase in association with hearing function and MG (3).

Physical Exam

Neurologic Exam:

- CN1: Decreased sense of smell
- CN II: Normal vision
- CN III, IV, VI: notable ptosis of L eye; normal EOMI, denies diplopia, PERRLA
- CN V: Intact, no decrease or increase in sensation
- CN VII: L eye droop, when swallows dribbled out of left side of mouth noted
- CN VIII: no changes in balance, straight arm test negative; decreased hearing noted in L ear
- CV IX: decreased taste and smell
- CN X: Hoarse voice noted
- CN XII: Tongue movement in all directions with normal strength



Image: Kurukumbi, M., Weir, R.L., Kalyanam, J. et al. Rare association of thymoma, myasthenia gravis and sarcoidosis: a case report. *J Med Case Reports* 2, 245 (2008). <https://doi.org/10.1186/1752-1947-2-245>

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Case Report

Clinical History

- A 84 year-old- female with history of hypothyroidism and recent femur fracture, presenting with 1 week history of left sided ptosis and hoarseness.
- Patient had symptomatic improvement of hoarseness with steroids, but returned. MRI brain was negative for acute intracranial event
- At this visit, in addition to ptosis and hoarseness, patient complained of dribbling out of side of mouth with swallowing, dysphagia associated with the sensation that food was getting stuck in her chest, as well as fatigue with mastication. Mentioned difficulty with word finding as well as occasional slurred speech.

Assessment & Plan

- MRI brain negative, ruling out intracerebral stroke.
- With numerous cranial nerves involved had concern for other sinister pathologies, including Brain Stem stroke, or skull base tumor, CT skull base was ordered for further evaluation.
- Myasthenia gravis was also in the differential so Myasthenia Panel was ordered.
- With patient's complaints of dysphagia, and weakness with mastication, where concerned about her swallowing mechanism, barium swallow was ordered.

Results

- Myasthenia panel returned first, all antibodies positive, as noted in Laboratory section. This essentially confirmed results of myasthenia.
- Patient was then started on high dose steroids, pyridostigmine, and was referred to neurology.
- CT skull base, was changed to CT chest to evaluate for Thymoma; this returned as negative.
- Symptom improvement and resolution was achieved from everything, except dysphagia. Dysphagia remained despite increased steroids. Patient received IVIG, which did result in improvement of dysphagia.
- With history of thyroid disease, did increase monitoring of TSH to every 6 months, in the setting of new autoimmune disease.
- With continue use of steroids, as well as recent fragility fracture, patient was started on Alendronate for additional bone protection.

Laboratory Results

Myasthenia Gravis Panel

	Results	Normal
AChR Binding Abs	44.50 H	0.00 – 0.24 nmol/L
AChR Blocking Abs	43 H	0 - 25%
AChR Modulating Abs	57 H	0-20%
Anti-striation Abs	1:160 H	Neg : <1:40

Conclusion & Key Learning Objectives

- It is important to keep Myasthenia Gravis on differential for elderly patients, as can present in a similar manner as a plethora of other pathologies.
- It is highly likely that there are patients in our community with Myasthenia that have been misdiagnosed.
- Diagnosis is important as treatments are available, and improve quality of life; and complications results from delay of treatment can be lethal.

References

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