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Case Report

Tongue Thrusting: A Sign of Myasthenia Gravis

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Abstract

Tongue thrusting is a sign of Myasthenia Gravis. MG has a variety of presentations, including bulbar symptoms. Tongue thrusting can be misdiagnosed as a brainstem stroke and anaphylaxis. Early identification should prompt appropriate antibody testing and treatment. We include three cases of antibody positive MG presenting with tongue thrusting as a clinical feature.

Introduction

Myasthenia gravis (MG) may present with tongue thrusting mimicking signs of brainstem stroke and anaphylaxis. Tongue weakness from bilateral hypoglossal nerve dysfunction results in failure of tongue protrusion. Tongue weakness from MG may present as paradoxical protrusion of the tongue during speech. A protruded tongue is usually associated with macroglossia. In MG, it may be associated with tongue atrophy. Tongue weakness and heaviness from bulbar MG may be mistaken for an anaphylactic reaction to antibiotics or anesthetics. Fifteen percent of MG patients present with bulbar symptoms [1]. A diagnosis of bulbar MG should be considered when palatal weakness and tongue swelling, thought to be from anaphylaxis, fails to respond to appropriate treatment. Double vision, lid ptosis, difficulty swallowing, nasal speech and bilateral extremity weakness may appear when acetylcholine motor endplate receptors are degraded by an antibody/complement autoimmune bombardment. Of note, bulbar MG may present with isolated dysphonia and dysphagia. Recognition of these symptoms will allow early diagnosis

and treatment. We present a series of three adult patients with varied presentations of bulbar MG where tongue thrusting was present. We encourage trainees to consider tongue thrusting as a useful sign of bulbar MG.

Case Reports

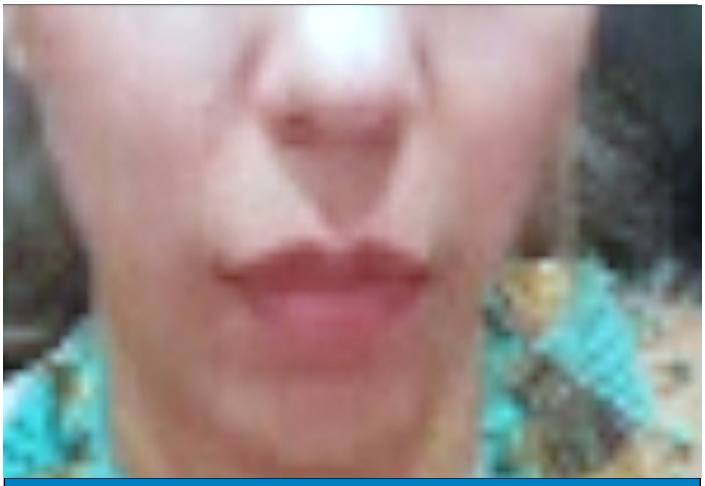
Case: #1

A 30-year-old female presented with six months of undiagnosed bilateral lid ptosis, fatigable speech, evening dysphagia and weakness of bilateral hand grip. She had increasing difficulty swallowing liquids and solids during the day. She manually removed food from her mouth when she could not swallow. While drinking, there was nasal regurgitation of fluid. She allowed saliva to drool from her mouth to prevent choking. Her speech became nasal and slurred after a brief period. Prolonged use of her hands made them heavy and clumsy. She could not fasten the clasp of her bra. She acquired a lisp with anterior tongue thrusts (Video 1). She was unable to position her lips to suck through a straw or whistle. She was short of breath. Clinically diagnosed with MG, she was treated with five alternate day plasmaphereses and high dose intravenous methylprednisolone. Once stabilized, a robotic thymectomy removed hyperplastic thymic tissue. An antibody panel revealed elevated Ach binding: 32.9%, (Normal: 0.0-0.4nmol/L) blocking: 63% (Normal 0-26 %), modulating: 95% (Normal <45 %) antibodies. Anti-Musk antibodies were negative (Positive >0.04 nmol/L). After six months of misdiagnosis, she was thrilled to have a correct diagnosis and successful treatment plan.

Case: #2

A 43-year-old female with known MG presented with years of prominent tongue thrusting, tongue atrophy and palatal weakness. She had exertional shortness of breath that was limiting her activity. She was never hospitalized. She had childhood surgery for strabismus with a left esotropia. An amblyopic left eye prevented a complaint of binocular diplopia. She never experienced nasal regurgitation of fluid or droopy eyelids. She was acetylcholine receptor antibody negative and MUSK antibody positive. She had intermittent difficulty swallowing liquids and solids. Physical exam revealed an atrophic tongue, profound weakness of frontalis muscles with inability to wrinkle her forehead or raise her eyebrows. She was unable to fully abduct her right eye. Eyelid closure was strong. There was a narrowed left palpebral fissure

where she had previous strabismus surgery. Her tongue was atrophic with a napkin ring sign at the tip. While speaking, her tongue thrusted forward through her teeth with a prominent lisp (video 2). She limited her treatment to steroids before she agreed to begin rituximab. Patient was anxious prior to starting immunosuppressive medication but she was pleased when her symptoms improved. A few months post rituximab, her tongue gained



Video 1: Tongue Thrusts

Video 2: Tongue Thrrusting

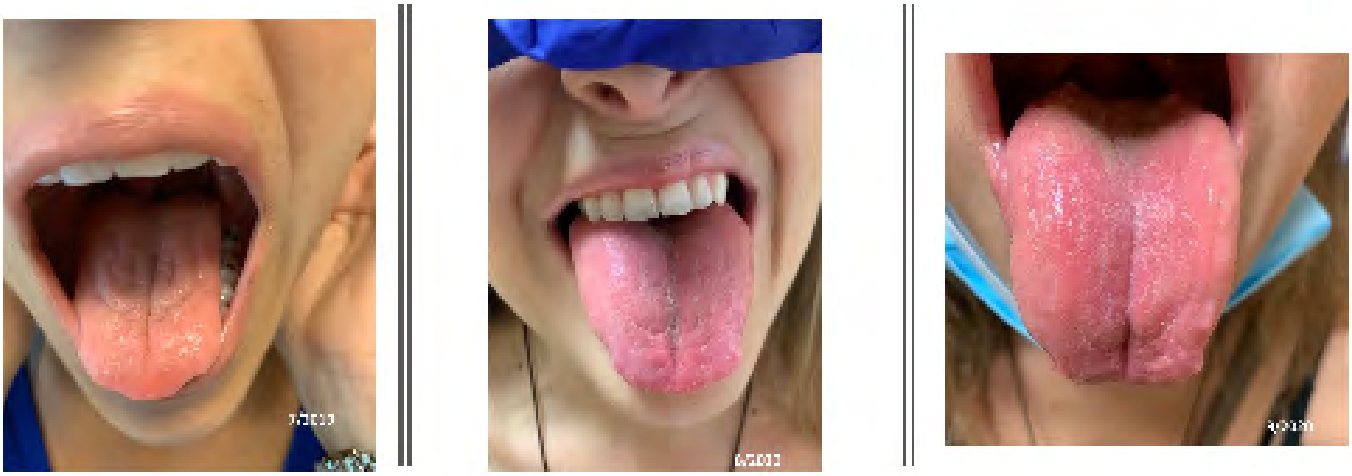


FIgure 1: Reversal of tongue atrophy (Before treatment and 1 & 3 months after treatment with rituximba, steroids and IVIG)

bulk and her tongue thrusts diminished (Figure 1).

Case: #3

A 67-year-old woman presented as a stroke alert with slurred speech and generalized symmetric muscle weakness. CT perfusion scans and follow up MRI brain found no evidence of cerebrovascular disease. Even with negative imaging, a brainstem transient ischemic attack remained a plausible diagnosis. A month prior to admission, she had elective arthroscopy to relieve shoulder pain. Increasing pain was thought secondary to post-procedural joint infection. She was treated with antibiotics and had shoulder irrigation and debridement. Immediate post-operative tongue swelling was treated with intravenous antihistamines and glucocorticoids for the possibility of an allergic reaction to antibiotic or anesthetic exposure. She developed CO2 retention but did not require mechanical ventilation. A Chest CTA angiogram excluded pulmonary embolism. Neurological consultation noted hypophonia, tongue thrusting and dysphagia of liquids and solids. MG was clinically diagnosed. Exposure to inhalation anesthetics (isoflurane and halothane) and antibiotics (aminoglycosides, fluroquinolones and macrolides) may have precipitated bulbar symptoms. Plasmapheresis, intravenous glucocorticoids and pyridostigmine had limited efficacy, so rituximab was administered. A MG antibody panel was later reported with elevated binding 6.24nmol/L (Normal 0.0-0.4nmol/L) and blocking antibodies to the Acetylcholine receptor of 57% (Normal 0-26 %) and normal modulating antibodies of 38% (Normal <45 %). MUSK antibody was negative. Prior to discharge, she received a robotic assisted thymectomy and placement of a percutaneous gastrostomy (PEG) tube. She voiced frustration and worry during her admission but with improvement of her symptoms, she became hopeful and optimistic. This occurred by upward titration of pyridostigmine and waiting the necessary time for immunosuppression to work. She was discharged to a rehabilitation facility after a three-week hospital stay. She was able to take adequate nutrition by mouth after resolution of dysphagia over a few weeks. The PEG tube was then removed.

Discussion

MG is an autoimmune disease with myriad clinical presentations making diagnosis challenging. The classic presentation of diurnal ocular and bulbar symptoms may not always be present. Isolated bulbar MG may mimic brainstem stroke, motor neuron diseases, Miller Fisher syndrome, myositis, botulism, or allergic reactions. In generalized MG, confirming antibody titers against acetylcholine receptors occurs in 80% of cases [1]. Ten percent of seronegative cases are associated with muscle specific kinase (Musk) receptor antibodies. A small number have li-

poprotein-related protein 4 in serum [2]. Age, gender, bulbar symptoms, and comorbid disease are clinical predictors of MG prognosis [3]. There is a bimodal distribution of onset occurring in young women in the third decade, and elderly men in the sixth decade [4]. Absence of lid ptosis with diplopia delays MG diagnosis in aged patients. Twenty eight percent of patients with MG develop esophageal dysmotility and a tongue that is unable to propel food towards the posterior nasopharynx resulting in dysphagia [5]. The cardinal symptoms of MG are fatigue and weakness [6]. Tongue thrusting is a curious symptom of MG. Hypoglossal weakness due to stroke or hypoglossal nerve damage causes dysfunction of tongue protrusion. In MG, a weak atrophic tongue produces tongue thrusting, likely from failure of tongue retraction. It is most noticeable during speech. In case #2, treatment of MG produced partial reversal of tongue atrophy and noticeable improvement in tongue thrusting. In case #3, the appearance of tongue thrusting pointed to the diagnosis of MG. There is limited literature regarding tongue thrusting as a clinical symptom of bulbar MG. A delay in the diagnosis of MG may result in adverse outcomes. As in case #3, severe dysphagia may require placement of a gastrostomy tube [5]. MG may be exacerbated by antibiotic or anesthetic exposure [2]. As in case #3, tongue immobility and heaviness after antibiotic exposure may be mistaken for an anaphylactic reaction. Dysarthria and a subjective "swollen tongue" sensation from MG was misdiagnosed as anaphylaxis in a previous report [5]. Our cases had positive serological testing confirming the diagnosis of MG. In seronegative cases, electromyography (EMG) with repetitive stimulation or single fiber analysis may aid in the diagnosis. It may have limited availability in the acute setting. We have successfully treated cases of seronegative and EMG negative (including repetitive and single fiber techniques) based on clinical features alone. In this series, different treatment modalities were used depending on physician or patient preference. Rituximab was used as induction therapy alone or with standard agents. It may slow progression of bulbar MG and improve disease symptoms [7]. The purpose of this report is to point out that tongue thrusting may be a symptom of bulbar MG.

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