Cerrahpaşa Med J 2022; 46(1): 75-77 CASE REPORT

Tongue Atrophy and Myopathy Mimicking EMG Findings in a Case with Acethylcholine Receptor-Positive Myasthenia Gravis

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Cite this article as: Asan F, Tütüncü M, Gündüz A, Adatepe N. Tongue atrophy and myopathy mimicking EMG findings in a case with acethylcholine receptor-positive myasthenia gravis. *Cerrahpaşa Med J.* 2022;46(1):75-77.

Abstract

Myasthenia gravis has various clinical presentations and can mimic multiple diseases. Isolated oro-facial muscle weakness and tongue atrophy at the time of presentation are rare. Besides the clinical picture, electrodiagnostic findings also can mimic various conditions and cause diagnostic difficulties. Our patient was a 28-year-old female admitted with non-fluctuating, progressive fatigue while talking and difficulty in swallowing. Her examination revealed tongue atrophy and fasciculations, ptosis, and facial weakness. Repetitive nerve stimulation revealed decrement in two muscle-nerve pairs. Electromyography demonstrated low amplitude and polyphasic motor unit action potentials at facial and neck muscles. She was diagnosed as having myasthenia gravis and was treated with pyridostigmine and methylprednisolone. Our case demonstrates that myasthenia gravis can present with tongue atrophy with fasciculations, oro-facial weakness, and electromyography findings mimicking myopathy. All these features were separately described during the disease course but our patient showed these features at presentation time. Repetitive nerve stimulation was the hallmark in correct diagnosis.

Keywords: Myasthenia gravis, myopathy mimicking EMG findings, oro-facial muscle weakness, repetitive nerve stimulation, tongue atrophy, tongue fasciculations

Myasthenia gravis (MG) is characterized by fatigable, fluctuating, and painless weakness of different muscle groups. Besides these hallmarks, MG can mimic a lot of clinical pictures. The diagnosis of MG is supported by electrophysiological tests and serum antibody levels. Serum anti-acetylcholinesterase (AchR) and anti-muscle specific kinase (MuSK) are the main antibodies studied for the diagnosis of MG. Repetitive nerve stimulation and single-fiber electromyography (EMG) are the main electrophysiological tests for the diagnosis of MG. In addition to typical presentation and electrophysiologic results, atypical presentation with electrophysiological results can create confusion in diagnosis. For this reason, we want to describe such a patient with diagnosis of MG.

Case Presentation

A 28-year-old woman who was born in Central Asia region was admitted with difficulty of swallowing and speaking for almost 8 years. Ten years before the admission, she suffered an episode of right-sided ptosis and diplopia for 1 year. Though slowly, ptosis and diplopia resolved completely. Thus, she was administered no medicine. Eight years before the admission, she started to experience progressive fatigue while talking. There was no other problems like diplopia, ptosis, swallowing problems, and extremity weakness at that time. In the last year, the symptoms during speaking worsened and trouble in swallowing, especially with liquids, added to it. When she had a right-sided peripheral facial paralysis 5 months before the admission, administration of oral methylprednisolone improved weakness during speaking and swallowing as well as peripheral facial paralysis. However, trouble in speaking and swallowing recurred after cessation of methylprednisolone treatment. Besides these complaints, she could not close eyelids. None of the symptoms fluctuated throughout the day. In her neurologic examination, pupil reflex and ocular movements were bilateral normal. Eyelids and neck flexion were weak, she could not whistle, and gag reflex was hypoactive. Bilateral sternocleidomastoid (SCM) muscles were atrophic. Tongue was atrophic and furrowed (Figure 1) with visible fasciculations; there was no deviation but protrusion was weak. Other muscle strength, deep tendon reflexes, sensory examination, and cerebellar examination were completely normal.

Five muscle-nerve pairs were studied for repetitive nerve examination which were accessory nerve-trapezius, musculocutaneous nerve-biceps brachii, facial nerve-nasalis, median nerve-abductor policis brevis, and axillary nerve-deltoid muscles. More than 10% decrement in amplitude and area was observed in accessory nerve-trapezius and musculocutaneous nerve-biceps brachii pairs at 2 Hz and 3 Hz stimulations. Orbicularis oris, orbicularis oculi, frontalis, SCM, deltoid, first digital interosseous, biceps brachii, infraspinatus muscles, and tongue were examined by needle EMG demonstrating fibrillation potentials only in the tongue. Low amplitude and polyphasic motor unit action potentials (MUAP) with rapid recruitment were determined especially in the neck and facial muscles.

Following EMG findings, the patient was diagnosed with possible MG and pyridostigmine 60 mg 3 times daily was started. Anti-AchR antibody level was 4.5 nmol/L. Thorax computed tomography without IV contrast administration showed soft tissue lesion (possible thymic hyperplasia or thymic remnant) in the anterior mediastinum. One month under pyridostigmine, her symptoms markedly improved (Figure 2), and she was also given oral methylprednisolone 1 mg/kg/day. After 3 months with oral methylprednisolone, her symptoms and also tongue atrophy markedly improved (Figure 3).

Received: April 12, 2021 **Accepted:** September 20, 2021 **Available Online Date:** November 7, 2021
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DOI: 10.5152/cjm.2021.21024

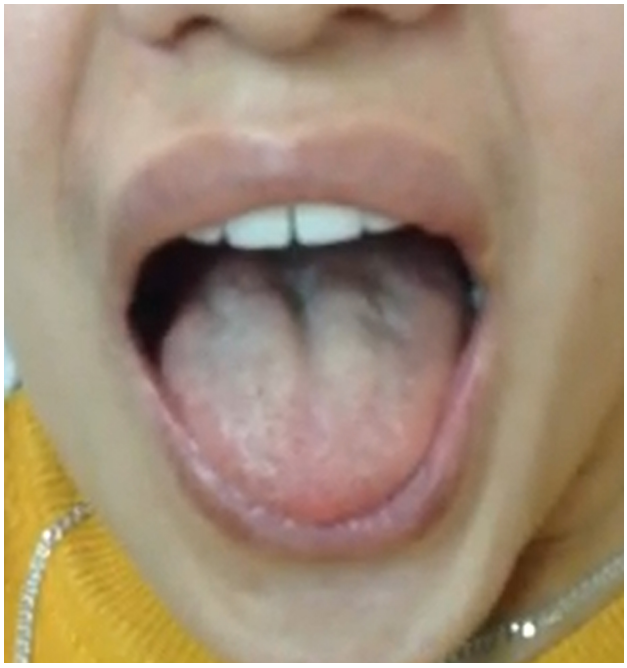

Figure 1. Morphology and maximum protrusion of tongue at presentation time.

Discussion

Our patient was admitted to our hospital with a 10-year term of non-fluctuating, progressive facial, and bulbar muscle weakness which was suggested initially as a kind of myopathy. Despite all non-fluctuating symptoms, the dramatic response of all complaints to oral steroid treatment which was given for the treatment of peripheral facial paralysis is the main key feature suggesting MG.

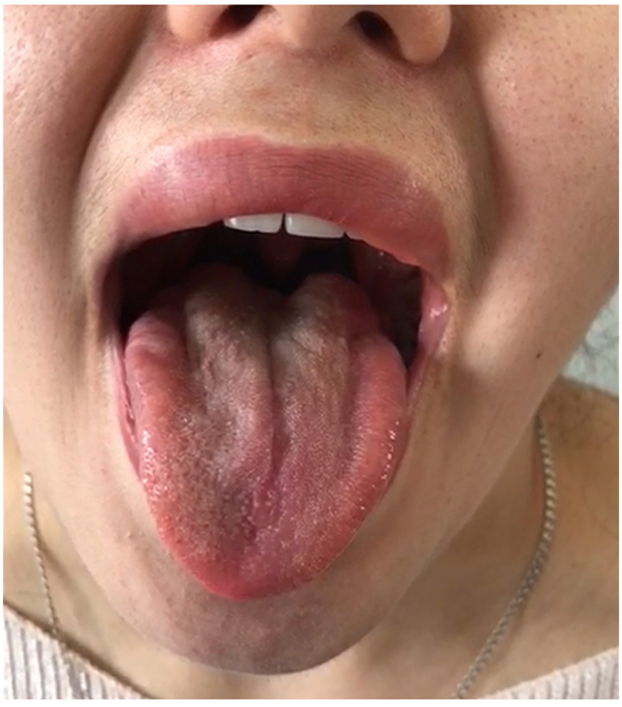

Figure 2. Morphology and maximum protrusion of tongue after 1 month of pyridostigmine treatment.

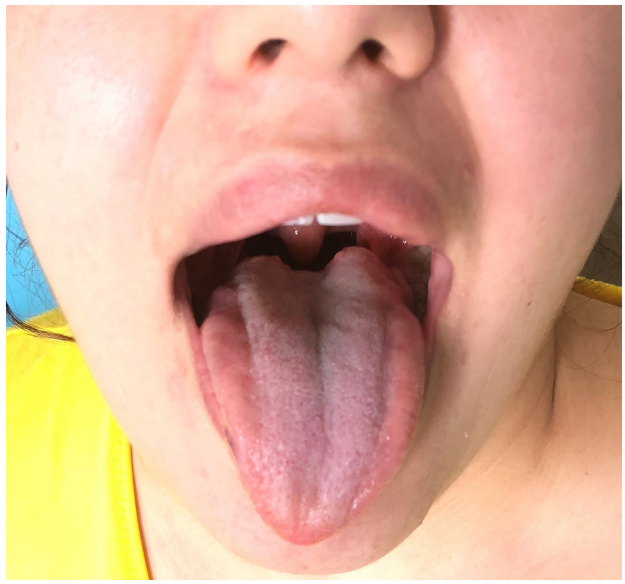

Figure 3. Morphology and maximum protrusion of tongue after 3 months of oral methylprednisolone treatment.

On the other hand, it is not exactly known whether the symptoms were mildly fluctuating at the beginning and had stabilized with atrophy of muscles after that period.

Spinobulbar muscular atrophy (Kennedy disease) is another possible diagnosis for adult-onset, progressive, facial, and bulbar muscle involvement with atrophy. But because of the female gender, it is out of question.

Although atrophy of the tongue is rare in myasthenic patients, it is more prominent in the anti-MusK antibodies positive group than the anti-Ach-R positive group.[1,2] It is unusual to encounter that kind of severe atrophy in Ach-R antibodies positive group at presentation time and to be reported by case reports in literature.[3,4] Some hypotheses have been suggested about the atrophy in patients with MG such as disuse atrophy,[5] long-term steroid treatment,[6] and directly anti-MuSK antibody-induced atrophy[7], but the exact reason is not clear. In our patient, neither long-term steroid treatment nor anti-MuSK antibody was present. For that reason, in clinical practice, we advise to check anti-AchR and anti-MuSK antibody levels in a patient with atrophy of oro-facial muscle and lower motor neuron signs in order to prevent diagnostic problems.

Fasciculations in untreated myasthenic patients are extremely rare and reported in a few patients which were also anti-AchR antibody positive.[2,8,9] Atrophy, fasciculation and short duration, and polyphasic MUAPs with rapid recruitment mimicking EMG findings of myopathy are shown in our patient. All these features are separately described in literature but the unique feature of our patient is having of all these findings at the time of presentation and without any treatment which suggests the disease itself as the primary cause.

We recommend performing a repetitive nerve stimulation test in a patient with oro-facial muscle weakness and atrophy with myopathic findings on EMG even if there is no fluctuation.

Informed Consent: Written informed consent was obtained from patient who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – F.A.; Design – F.A.; Supervision – N.U.A.; Materials – M.T.; Data Collection and/or Processing – M.T.; Analysis and/or Interpretation – A.G.; Literature Search – A.G.; Writing Manuscript – F.A.

Declaration of Interests: The authors have no conflicts of interest to declare.

Funding: The authors declared that this study has received no financial support.

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