



CASE REPORT

Focal Unilateral Palatal Myoclonus Causing Objective Clicking Tinnitus without Uvula Elevation Diagnosed by Concurrent Auscultation

Chindhuri Selvadurai, Sara Maguire Schaefer

Movement Disorders, Department of Neurology, Yale New Haven Health Hospital, New Haven, CT, USA

ABSTRACT

Palatal myoclonus generally entails a visible elevation of the palate and uvula and may be accompanied by myoclonus of other oropharyngeal muscles. A 55-year-old man presented with left ear clicking and hyperacusis. Examination showed arrhythmic left lateral soft palate contraction in the tensor veli palatini region without elevation of the uvula, which correlated with an audible click by auscultation with a stethoscope over the left ear. This is a rare case of focal, unilateral palatal myoclonus without visual uvula elevation with concurrent auscultation, demonstrating the importance of careful examination to detect focal myoclonic contractions.

Key Words Botox; Myoclonus; Palate.

Palatal myoclonus may occur secondary to brainstem lesions in the Guillain-Mollaret triangle between the red nucleus, inferior olivary nucleus, and dentate nucleus. If a lesion is found, pseudohypertrophy of the inferior olive is the most common. When a structural abnormality is not identified, it is termed “essential myoclonus”. Patients usually present with subjective or objective unilateral clicking sounds, sometimes have pain and ear fullness, and usually have complete bilateral palate and uvula elevation on examination.¹ Palatal myoclonus may be accompanied by myoclonus of other muscles in the throat, tongue, or face. Treatment for this condition includes oral medication, surgery, or botulinum toxin injections.² We report this case as it is a unique presentation not previously reported that emphasizes how a detailed neurologic examination can aid in diagnosis.

CASE REPORT

A 55-year-old previously healthy man presented to the neu-

rology clinic with ear clicking for almost one year. After attending a rock concert, he developed bilateral ear fullness, followed by left ear clicking and hyperacusis. The symptoms of clicking interfered with his mood and ability to sleep, causing extreme distress. It is unknown whether the clicking itself persisted during sleep. He was seen first by a dentist, who prescribed a dental guard to treat temporomandibular joint pain. He then saw an otolaryngologist who treated him with antibiotics for an ear infection. The patient then consulted a psychiatrist who diagnosed him with anxiety, which was treated with 0.25 mg clonazepam four times daily. None of these three treatments provided relief of the clicking or hyperacusis. Finally, two neurologists considered a diagnosis of palatal myoclonus; however, they did not visualize full palatal elevation on examination and thus did not diagnose or treat him. Upon arrival at our subspecialty clinic, his examination was notable for arrhythmic left lateral soft palate contraction in the region of the anterior soft palate that did not completely raise the palate or uvula. This contraction

Received: February 3, 2020 Revised: April 8, 2020 Accepted: April 28, 2020

Corresponding author: Chindhuri Selvadurai, MD

Movement Disorders, Department of Neurology, Yale New Haven Health Hospital, New Haven 06519, CT, USA / Tel: +1-203-785-4085 / E-mail: chindhuri.selvadurai@yale.edu

© This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (<https://creativecommons.org/licenses/by-nc/4.0>) which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

correlated with an audible click by auscultation with a stethoscope over the left ear. His physical and neurologic examinations, including gag and cough reflexes, were otherwise completely normal (Supplementary Video 1 in the online-only Data Supplement). Magnetic resonance imaging of the brain with and without contrast, as well as computed tomography angiogram of the head, were both normal. Ultimately, he received 2.5 units of Onabotulinum toxin A injected into the affected muscle by his otolaryngologist and had mild improvement in clicking and hyperacusis. He was able to sleep better and felt less anxious overall.

DISCUSSION

The five muscles of the soft palate include the tensor veli palatini, levator veli palatini, musculus uvulae, palatoglossus, and palatopharyngeus.³ Importantly, regarding our case, the tensor veli palatini tightens the anterior soft palate, flattens the arch, and opens the Eustachian tube. If it is affected unilaterally, it will deviate the soft palate laterally, and overactivity explains the audible click noted by the patient and heard on examination. The levator veli palatini tightens the posterior soft palate and raises the soft palate.

Palatal myoclonus generally entails a visible elevation of the palate, its five muscles, and uvula and may include myoclonus of other oropharyngeal muscles. It is crucial to rule out any structural lesions (e.g., inferior olivary hypertrophy) before diagnosing essential palatal myoclonus. This is a rare case of a patient with focal, unilateral palatal myoclonus without uvula elevation, and focal contraction only visualized on close inspection with concurrent auscultation. The cause of tinnitus in this disorder is not well understood, but one theory is that abnormal muscle contractions can induce pressure changes in the Eustachian tube, leading to disequilibrium between the atmospheric pressure and the middle ear pressure, which produces oscillations in the tympanic membrane causing tinnitus.^{4,5} Focal palatal myoclonus has rarely been reported.² The differential diagnosis for clicking tinnitus includes focal middle ear myoclonus due to tensor tympani or stapedius contraction with increasing tension. This cannot be visualized on oral examination but rather by examination of the tympanic membrane on otoscopic examination and confirmed by tympanometry.⁶ Unfortunately, we were unable to perform tympanometry or electromyography on our patient, as he did not return for follow-up in our clinic. Treatments such as anticonvulsants, anxiolytics, and surgical interventions are largely unsuccessful. Botulinum toxin can be injected into the levator veli palatini and/or the tensor veli palatini muscles and has been reported to provide symptomatic relief.⁷ This patient elected to see an otolaryngologist

for injections and reported mild improvement in the clicking and tinnitus. The side effects of this treatment include dysphagia, hypernasality, or nasopharyngeal regurgitation.^{8,9} This case illustrates that practitioners should perform careful oral examinations with auscultation to detect focal myoclonic contractions in a patient with ear clicking without obvious palatal or uvula elevation.

Supplementary Video Legend

Video 1. Tensor veli palatini contraction without complete palatal or uvula elevation. A 55-year-old man who presented with left ear clicking and hyperacusis was found to have arrhythmic left lateral soft palatal contraction without uvula elevation, correlating with an audible click by auscultation with a stethoscope. The patient consented, and we obtained a letter of consent from our patient for participation in this educational video.

Supplementary Materials

The online-only Data Supplement is available with this article at <https://doi.org/10.14802/jmd.20010>.

Conflicts of Interest

The authors have no financial conflicts of interest.

Acknowledgments

We acknowledge our patient's time and medical contribution.

Author Contributions

Conceptualization: Chindhuri Selvadurai. Data curation: Sara Maguire Schaefer. Formal analysis: all authors. Investigation: all authors. Methodology: all authors. Resources: Sara Maguire Schaefer. Writing—original draft: Chindhuri Selvadurai. Writing—review & editing: all authors. Approval of final manuscript: all authors.

ORCID iDs

Chindhuri Selvadurai <https://orcid.org/0000-0001-8477-9872>
Sara Maguire Schaefer <https://orcid.org/0000-0003-0796-2272>

REFERENCES

- Zadikoff C, Lang AE, Klein C. The 'essentials' of essential palatal tremor: a reappraisal of the nosology. *Brain* 2006;129(Pt 4):832-840.
- Sinclair CF, Gurey LE, Blitzer A. Palatal myoclonus: algorithm for management with botulinum toxin based on clinical disease characteristics. *Laryngoscope* 2014;124:1164-1169.
- Short MJ, Levin-Goldstein D. *Head, neck and dental anatomy*. 4th ed. New York: Delmar Cengage Learning, 2012;240-243.
- Jamieson DR, Mann C, O'reilly B, Thomas AM. Ear clicks in palatal tremor caused by activity of the levator veli palatini. *Neurology* 1996;46:1168-1169.
- Smith ME, Scoffings DJ, Tysome JR. Imaging of the Eustachian tube and its function: a systematic review. *Neuroradiology* 2016;58:543-556.
- Keidar E, Kwartowitz G. Tensor tympani syndrome [Updated 2020 Jan 31] [Internet]. Treasure Island: StatPearls Publishing; c2019. [accessed on 2019 Dec 20]. Available at: <https://www.ncbi.nlm.nih.gov/books/NBK519055/>.
- Penney SE, Bruce IA, Saeed SR. Botulinum toxin is effective and safe for palatal tremor: a report of five cases and a review of the literature. *J Neurol* 2006;253:857-860.
- Anis MM, Pollak N. Treatment of palatal myoclonus with botulinum toxin injection. *Case Rep Otolaryngol* 2013;2013:231505.
- Kojovic M, Cordivari C, Bhatia K. Myoclonic disorders: a practical approach for diagnosis and treatment. *Ther Adv Neurol Disord* 2011;4:47-62.