Disputes & Debates: Editors' Choice

Robert C. Griggs, MD, FAAN, Section Editor; Steven Galetta, MD, FAAN, Co-Editor

Editors' note: Teaching Video NeuroImages: Olivary enlargement and pharyngeal nystagmus

In "Teaching Video NeuroImages: Olivary enlargement and pharyngeal nystagmus," authors Finlay and Yacovino described a 77-year-old woman with a pontine cavernoma and rhythmic contractions of the soft palate and upper larynx on videofluoroscopy. The accompanying MRI showed right inferior olivary nucleus hypertrophy. Dr. Bhattacharjee comments that the simultaneous palate-pharyngeal tremor may be due to their common innervation by the vagus nerve. He also asks about the interval between the cavernoma expansion, the radiologic signal changes, and the patient's symptoms. Authors Finlay and Yacovino clarify that the olivary hypertrophy was present on the patient's initial MRI and that the palatal tremor was diagnosed 36 months after that study. They also note that synchronic contraction additionally was seen in the bilateral eyelids in their patient, which does not support Dr. Bhattacharjee's theory of common vagus nerve innervation. Dr. Mendez-Guerrero suggests that the term pharyngeal nystagmus, coined a century ago, may be misleading as a central nystagmus now infers a vestibular nuclei or vestibular pathway impairment, which is not present in this disorder. He proposes using the term pharyngeal/laryngeal myorhythmia, denoting a slow (2–3 Hz) rhythmic movement disorder of the cranial musculature. The authors expand on the terminology mentioned by Dr. Mendez-Guerrero and on their use of Spencer's original nomenclature.

Megan Alcauskas, MD, and Steven Galetta, MD
Neurology® 2018;90:753. doi:10.1212/WNL.0000000000005318

Reader response: Teaching Video NeuroImages: Olivary enlargement and pharyngeal nystagmus

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Neurology® 2018;90:753–754. doi:10.1212/WNL.0000000000005316

The interesting Teaching Video NeuroImage by Drs. Finlay and Yacovino¹ demonstrated simultaneous soft palate and pharyngeal tremor and hypertrophic olivary degeneration (HOD). The palatal tremor is secondary to levator veli palatini contraction. The levator veli palatini is mainly supplied by the vagus nerve via the pharyngeal plexus. Most of the pharyngeal muscles are supplied by the vagus nerve, so simultaneous palate-pharyngeal tremor can be explained by the common nerve supply.

The authors demonstrated unilateral olivary degeneration,¹ which is less often found to be idiopathic than bilateral HOD.² However, olivary hypertrophy usually appears 4–6 months after the initial insult and the soft palatal tremor appears after 10–11 months of the initial insult.³ In the presented patient, the insult is most likely either the expansion of the pontine cavernoma or the cavernous bleed. It would be interesting to know the interval among the cavernoma expansion/bleed and the appearance of the radiologic signal changes in the inferior olive and the palato-pharyngeal tremor.

Author disclosures are available upon request (journal@neurology.org).

1. Finlay JB, Yacovino DA. Teaching Video NeuroImages: olivary enlargement and pharyngeal nystagmus. *Neurology* 2017;89:e168.
2. Konno T, Broderick DF, Tacik P, Caviness JN, Wszolek ZK. Hypertrophic olivary degeneration: a clinico-radiologic study. *Parkinsonism Relat Disord* 2016;28:36–40.
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Reader response: Teaching Video NeuroImages: Olivary enlargement and pharyngeal nystagmus

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Neurology® 2018;90:754. doi:10.1212/WNL.0000000000005319

We noted with interest the Teaching Video NeuroImage by Drs. Finlay and Yacovino.¹ The authors examined the phenomenology of a rare slow movement disorder of pharynx and larynx due to a brainstem cavernoma using videofluoroscopy. The term pharyngeal nystagmus, beyond the initial description more than a century ago,² may mislead readers. Central nystagmus implies a vestibular nuclei or vestibular pathway impairment³ that are not related to this movement disorder. Hence, pharyngeal nystagmus was known as palatine myoclonus/tremor, in which the dentate-rubro-olivary tract is commonly involved.⁴ This term is also problematic due to its phenomenologic differences with true myoclonus. Currently, this movement disorder is better considered part of the spectrum of myorhythmias: slow (2–3 Hz) rhythmic movement disorders of cranial musculature or limbs.⁵ We suggest that the term pharyngeal nystagmus be avoided and replaced by pharyngeal/laryngeal myorhythmia.

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2. Spencer HR. Pharyngeal and laryngeal “nystagmus.” *Lancet* 1886;2:702–704.
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Author response: Teaching Video NeuroImages: Olivary enlargement and pharyngeal nystagmus

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Neurology® 2018;90:754–755. doi:10.1212/WNL.0000000000005344

We appreciate the comments made by Dr. Bhattacharjee regarding our Teaching Video NeuroImage.¹ The cavernoma was an incidental feature on the brain CT conducted after the patient had a fall, and mild hypertrophy olivary degeneration (HOD) was already visible on the subsequent MRI. However, the palatal tremor (PT) was not diagnosed until 36 months after the first studies. Here, PT and HOD are the manifestation of denervation supersensitivity, secondary to lesions involving the unilateral supraolivary central tegmental tract (CTT). According to the anatomical and radiologic changes in HOD, the MRI would likely not detect pseudohypertrophy early on or after a substantial time following Mollaret triangle injury.²

With a cavernoma, a direct expansion, bleeding/gliosis, or direct neurotoxicity induced by the hemosiderosis effect on the CTT are possible.³ The last would explain the progressive and silent deafferentation of the ipsilateral olivary nucleus.

Dr. Bhattacharjee’s observation that the concert of pharyngeal muscle contraction in our case was mainly innervated by the vagus nerve is interesting; however, the same synchronic contraction was

described in the eyelids, facial muscles, and diaphragm, which are innervated by other nerves.⁴ After review of the video-oculographic study, we identified similar bilateral synchronic eyelid contraction.

We also appreciate the clarifying comments made by Mendez-Guerrero et al. regarding our article,¹ and reciprocally comment on some of the terms used.

Similar to congenital nystagmus, the involuntary rhythmic ocular movement in oculopalatal tremor is classified as pendular nystagmus and satisfies the characteristics to be defined as central nystagmus.⁵ However, the mechanism here is not primarily a central vestibular impairment; thus, it is not entirely accurate to attribute all cases of central nystagmus to a vestibular defect.

We agree there is a discrepancy in the terminology of palatal movement: tremor, myoclonus, and myorhythmia are used interchangeably in the literature.⁶ Although it resembles a tremor, the movement is often repetitive rather than oscillatory and only involves pharyngeal constrictor muscles, so it is commonly considered a myoclonus, as in our case.¹

We highlighted the valuable clinical parallels between the movement of the eyes and the pharynx, which Spencer⁷ summarized under the term pharyngeal nystagmus, a typical observation of the clinicians of the time. While the formal terminology has not yet been established, we apologize for the slight departure from strict etymologic accuracy and thank Mendez-Guerrero et al. for prompting us to expand on the content in the case.

1. Finlay JB, Yacovino DA. Teaching Video NeuroImages: olivary enlargement and pharyngeal nystagmus. *Neurology* 2017;89:e168.
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CORRECTION

Alemtuzumab CARE-MS I 5-year follow-up: Durable efficacy in the absence of continuous MS therapy

Neurology® 2018;90:755. doi:10.1212/WNL.0000000000004908

In the article “Alemtuzumab CARE-MS I 5-year follow-up: Durable efficacy in the absence of continuous MS therapy” by E. Havrdova et al.,¹ the title should read “Alemtuzumab CARE-MS I 5-year follow-up: Efficacy in the absence of continuous MS therapy.” The word “durable” was removed by the Editors during the review process and it was later reinserted in error.

Reference

1. Havrdova E, Arnold DL, Cohen JA, et al. Alemtuzumab CARE-MS I 5-year follow-up: durable efficacy in the absence of continuous MS therapy. *Neurology* 2017;89:1107–1116.

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Alemtuzumab CARE-MS I 5-year follow-up: Durable efficacy in the absence of continuous MS therapy

Neurology 2018;90;755

DOI 10.1212/WNL.0000000000004908

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