Pearls & Oy-sters: Progressive ataxia and palatal tremor

Imaging and disease course

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Pearls

- Palatal tremor (PT) (or myoclonus) is a rare movement disorder comprising 2 forms: essential PT (EPT) and symptomatic PT (SPT).
- The rhythmic movement of the PT may be unilateral or bilateral with partial or complete rhythmicity. During sleep, tremor ceases in EPT but not in SPT.
- Treatment for SPT includes medication such as valproate, clonazepam, or trihexyphenidyl; botulinum toxin has been successful in only a few reported cases.

Oy-sters

- Examination of the palate and careful review of imaging should be done in patients who present with progressive ataxia to recognize the clinical syndrome of progressive ataxia and PT.
- MRI may demonstrate olivary pseudohypertrophy with contrast enhancement in PT and should not be confused with malignancy or stroke.

A 52-year old man presented to the movement disorders clinic with concern for 3 years of progressive worsening gait and slurred speech. He would veer to the left while walking and steer to the left while driving. His voice would quiver occasionally. His wife became aware of clicking noises while he was asleep. Proximate to his visit, he noticed slurring in his voice and difficulty with multiple motor tasks. He denied history of stroke, cancer, trauma, or neurosurgical procedures; his family history was unremarkable.

General examination was unremarkable, and vital signs were within normal limits. Pertinent findings on neurologic examination included a persistent, rhythmic tremor of the soft palate that extended to include other muscles in the throat, with externally visible movements in the anterior neck. The patient could arise somewhat slowly from a chair with his arms crossed. His gait was slightly wide-based; he was able to walk on heels and toes. He had difficulty standing in tandem position with eyes closed but was able to hold the position. He was mildly dysarthric. There was slight dysmetria on finger-to-nose testing, significant dysdiadochokinesis worse on the left than the right, bilateral hypometric saccades, but normal smooth visual pursuit. Extraocular movements were otherwise intact. MRI of the brain showed increased T2 hyperintensity and hypertrophy of bilateral inferior olivary nuclei (ION); there was gadolinium enhancement of the ION (figure).

The patient was diagnosed with progressive ataxia and palatal tremor (PAPT). Trials of clonazepam and lamotrigine to treat the PT were ineffective. Nasal sumatriptan 20 mg slowed the rhythmicity for 3 minutes, but after 10 minutes, it returned to baseline. He was followed from October 2004 to April 2012, during which there was no significant change in

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Axial brain MRI with (A) T2-weighted and (B) precontrast and (C) postcontrast T1-weighted sequences shows T2 hyperintensity, hypertrophy, and patchy enhancement of bilateral inferior olivary nuclei. Follow-up MRI 13 years later with (D) T2-weighted and (E) noncontrast T1-weighted sequences demonstrates atrophy with residual T2 hyperintensity.

PT. When the patient was seen again in May 2018, he complained of problems with fine motor control in his hands when using tools or writing and increased difficulty with motor learning. His wife reported that the clicking noises had stopped. On examination, the tremor had resolved spontaneously, but the patient continued to have significant dysdiadochokinesis, worse on the left than the right, bilateral hypermetric saccades on looking from right to midline, and difficulty standing or walking in tandem longer than 5–6 seconds. No palatal myoclonus was observed. Follow-up MRI of the brain showed atrophy of bilateral ION with some residual T2 hyperintensity, as shown in the figure.

Discussion

PT (formerly known as palatal myoclonus) is a rare movement disorder consisting of rhythmic movements of soft palate and comprises 2 forms: EPT and SPT.¹ The name palatal tremor was formally coined in 1990 at the First International Congress of Movement Disorders based on characteristics of the movements, which were considered to

be more regular and continuous, like a tremor, and less shock-like, which would be typical of myoclonus. There are some features that are consistent with myoclonus, such as variability.² PT may be unilateral, bilateral, or asymmetric, with partial or complete rhythmicity at a reported frequency of 100–150/min.^{3,4} During sleep, PT mostly ceases in EPT in about 50% of the cases but is invariably present in all patients with SPT.^{2,5}

EPT occurs without an obvious structural lesion in the brain^{6,7} and is characterized by rhythmic movement of anterior soft palate, attributable to contractions of the trigeminal nerve–innervated tensor veli palatini muscle, and not uncommonly auditory clicks.⁷ SPT is often associated with a brainstem or cerebellar lesion involving the dentato-rubro-olivary pathway (Guillain-Mollaret triangle).^{1,3,5,6} In SPT, palatal movements result from activation of the levator veli palatini muscle. EPT is often associated with ear clicks; however, levator veli palatini activity can also result in audible clicks. Therefore, presence of ear clicks alone cannot differentiate between EPT and SPT.^{5,7} Extrapalatal manifestations are seen with this movement disorder, including ocular disorder and extremity and gait ataxia.^{5,7}

The most common etiologies of PT are stroke, trauma, demyelinating lesion, and posterior fossa tumors.⁷ Histopathologic examination in PT shows enlarged and vacuolated neurons, increased number and size of astrocytes, and fibrillary gliosis in the region of hypertrophic inferior olivary degeneration.⁵ These changes appear on MRI as olivary hypertrophy. It has been reported that MRI T2 changes occur 1 month after the inciting event and are usually present for 3–4 years.⁶ It is not unusual for the affected ION to atrophy over time.⁷

Contrast enhancement is generally associated with malignancy, infection, or subacute phase of ischemic infarction. We are aware of only one case describing hypertrophic olivary degeneration (HOD) with enhancement, which occurred after surgical excision of a medulloblastoma, and therefore the mechanism could have been related to the surgical intervention.⁸ Our patient has no confounding history of tumor, demyelinating lesion, inflammatory processes, Alexander disease, stroke, or surgical intervention, making the point even stronger that contrary to prior reporting,^{8–10} contrast enhancement of the ION may occur with HOD, and therefore enhancement does not distinguish between HOD and other diagnoses such as malignancy or infectious processes.

PAPT as described here is a subgroup of SPT that is further subdivided into sporadic and familial forms.⁷ Features used to distinguish sporadic PAPT, as in this case, include abnormal eye findings (saccadic pursuit, pendular nystagmus), olivary degeneration, and progression of ataxia. Familial PAPT does not demonstrate olivary hypertrophy. Reported causes of familial PAPT include Alexander disease, polymerase gamma mutation, and spinocerebellar ataxia type 20.⁵ The most problematic symptoms in PAPT include visual disturbance, dysarthria, dysphagia, appendicular and gait ataxia, and other features associated with cerebellar syndromes.⁷ There is no reported effective treatment to prevent or halt the progression of ataxia, the most disabling symptom of PAPT.

Despite recognition of the changes on MRI scan and the histopathology, the natural history of PAPT is unknown. We describe a patient who had spontaneous resolution of PT but continued progression of gait ataxia. This supports data that pathologic changes in the ION are associated with the development but not the maintenance of PT.¹¹ In our review of the literature, we found few cases that describe PAPT and this case is unique in that the patient had spontaneous resolution of PT but continued progression of ataxia and persistent atrophy of the ION as judged by MRI. The patient's imaging was unique in demonstrating HOD with gadolinium enhancement in the absence of tumor, stroke, or surgical intervention.

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Disclosure

The authors report no disclosures relevant to the manuscript. Go to Neurology.org/N for full disclosures.

Appendix Authors

Name	Location	Contribution
Swati Pradeep, DO	University of Kentucky, Lexington	Case concept and design, acquisition of data, interpretation of data, manuscript writing
Parneet Grewal, MD	University of Kentucky, Lexington	Case concept and design, acquisition of data, interpretation of data, manuscript writing
Samia Jahan, MBBS	University of Kentucky, Lexington	Case concept and design, acquisition of data
Flavius D. Raslau, MD	University of Kentucky, Lexington	Neurodiagnostic evaluation, critical revision of manuscript for intellectual content
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