Unilateral symptomatic palatal tremor due to pontocerebellar infarction

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Introduction
Palatal tremor (PT) is an uncommon movement disorder which is classified as essential and symptomatic forms. Patients with symptomatic palatal tremor (SPT) often have demonstrable lesions in the Guillain Mollaret triangle.[1-3] Electrophysiological studies in PT are sparse.[1,4,5] We recorded blink reflex and sensory evoked potentials in a young man with SPT from pontomedullary infarction extending to cerebellar peduncles.

Case Report
A 34-year-old farmer had acute onset of vertigo and ataxia without dysphagia, dysphonia, weakness or sensory impairment 5 months before admission with partial improvement after 1 month. Two months prior to the evaluation, his voice had changed without dysphagia, nasal regurgitation or auditory clicks. Examination revealed asymmetric gaze-evoked nystagmus (right > left), lower motor facial weakness, impaired corneal reflex and impaired hearing on right side with right unilateral PT (video). There was right-sided limb, stance and gait ataxia with normal power and sensation.

Magnetic resonance imaging revealed old right pontocerebellar infarction with left inferior olivary nucleus (ION) hyperintensity in T2 and FLAIR sequences [Figure 1]. Flow voids were normal in the major intracranial vessels. Facial nerve conductions revealed reduced amplitudes on right side with marginally increased latencies. Blink reflex study revealed prolonged latencies with reduced amplitudes in right side; the blink reflex in left side was normal [Table 1, Figure 2]. The right auditory evoked potentials (AEP) revealed normal wave I with poorly defined wave II and III; wave IV and V were absent while left AEP was normal. Bilateral median nerve somatosensory evoked potentials were normal. Treatment with clonazepam did not improve the tremor.

Discussion
Palatal tremor (PT) earlier known as palatal myoclonus is a rhythmic contraction of the palatal muscles. Differentiation between essential and symptomatic palatal tremor (SPT) is based on the clinical and radiological findings.[1-3] The SPT occurs with lesions in the brainstem including red nucleus and central tegmental tract (CTT) from infarctions, hemorrhages, encephalitis, demyelination, etc.[1-3] In a pooled analysis of the patients with PT, nearly three-fourth had SPT. Lesions due to cerebrovascular diseases was the commonest cause of SPT.[1] Pharmacotherapy of PT with clonazepam, valproate, sumitriptan and trihexyphenidyl has had variable success.[3,6] Chemodenervation with botulinum toxin injection to palatal muscles using electromyographic guidance has been found to be useful in the treatment of PT.[3,7]

Table 1: Facial nerve conductions and blink reflex in the patient

| Stimulation        | Right     | Left     |
|--------------------|-----------|----------|
| CMAP latency       |           |          |
| Nasalis            | 2.80 ms   | 2.45 ms  |
| Orbicularis oculi  | 3.65 ms   | 3.15 ms  |
| CMAP amplitude     |           |          |
| Nasalis            | 1.89 mV   | 3.29 mV  |
| Orbicularis oculi  | 1.04 mV   | 1.53 mV  |
| Blink reflex       |           |          |
| R1 latency         | 14.0 ms   | 9.5 ms   |
| R1 amplitude       | 0.15 – 0.2 mV | 0.8 – 1.0 mV |
| R2i latency        | 34.5 ms   | 32.0 ms  |
| R2c latency        | 30.3 ms   | 36.5 ms  |

CMAP = Compound muscle action potential; R2i = Ipsilateral R2 response; R2c = Contralateral R2 response

Images in Neurology

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Generation of SPT has been traditionally linked to the appearance of hypertrophic degeneration of the ION which is considered to be the central pacemaker for generation and sustenance of SPT.\(^8\) ION outflow lesions including the inferior cerebellar peduncle are not associated with SPT while lesions in the CTT are associated with SPT.\(^2\) In patients with unilateral SPT, the ION hypertrophy is seen on the opposite side.\(^2,3\) The major ION inputs are from CTT and spinocerebellar afferents. In our patient, the PT was seen on the right side with ION hyperintensity on left side. The causative lesion was the right pontocerebellar infarction which probably interrupted the ION afferents.

The SPT occurs about a month or later after the inciting event and persists for years. Meta-analysis of literature on magnetic resonance imaging in SPT patients revealed olivary hyperintensity by 1 month which persisted up to 4 years. The olivary enlargement occurred by 6 months and resolved by 3–4 years.\(^9\) Occurrence of the SPT with contralateral olivary hyperintensity 3 months after the posterior circulation infarction in our patient is consistent with the published literature.

A study of brainstem auditory evoked potentials (BAEPs) in 20 patients with SPT revealed abnormality in six patients including one patient with brainstem infarction.\(^5\) Our patient had abnormal BAEP suggesting impaired conduction in the auditory pathway central to the auditory nerve considering the poorly defined wave II and III with absent wave IV and V which correlates with the lateral medullary and pontine infarction. Somatosensory evoked potentials are preferentially transmitted through the medial lemniscal pathway in the brainstem and were probably spared considering the lateral location of the lesion in our patient.

Blink reflex evaluates the trigeminal afferent, facial efferent, oligosynaptic and polysynaptic brainstem pathways extending through pons to medulla oblongata.\(^{10}\) Blink reflex studies in six patients with SPT revealed prolongation of R1 in three patients. Enhanced blink reflex recovery curve was seen in two patients with SPT and two out of the five patients with EPT had enhanced blink reflex recovery curve.\(^1\) Enhanced blink reflex recovery was also seen in a patient with intestinal lymphoma who had PT.\(^4\) In our patient, the blink reflex evaluation revealed efferent defect in view of the reduced amplitudes of R1 and ipsilateral R2, explained by the pontomedullary infarction. As the right orbicularis oculi direct response latency was only marginally increased, the blink reflex latency prolongations is likely to be central in origin. While the reduced amplitude of the R1 is expected from the axonal nature of the lesion, the exact reason for the latency prolongation is not clear. The BAEP and blink reflex latency abnormalities have been documented earlier; however, the amplitude abnormalities in the blink reflex have not been reported.

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