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Authors
Shoeibi, A
Litvan, I

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Prominent tongue and jaw tremor in a patient with probable Progressive Supranuclear Palsy

Ali Shoeibi, M.D.1,2 and Irene Litvan, M.D.1
1Movement Disorder Center, UC San Diego Department of Neurosciences, La Jolla, CA, USA
2Department of Neurology, Mashhad University of Medical Sciences, Mashhad, Iran

Keywords
Progressive supranuclear palsy; tongue and jaw tremor; case-report

Isolated tongue tremor (without palatal and limb tremor) has rarely been reported in patients with Parkinson disease, and in those rare cases the tremor was Levodopa responsive.1, 2 This is the first report of an isolated tongue and jaw tremor not responsive to Levodopa in a patient with progressive supranuclear palsy (PSP).

Case report

This 82-year-old female presented with progressive parkinsonism for 8 years before evaluation when her family noticed decreased facial expression. Thereafter, she had difficulties writing and her gait slowed with decreased associated movements. A year later, she developed dysarthria and drooling. Later, she developed jaw and tongue tremor. Balance progressively worsened. Two years later, she was diagnosed with idiopathic Parkinson’s disease. She was started on carbidopa/levodopa which was gradually increased up to 1250 mg with 1000 mg of entacapone, but without success in the treatment of tremor or parkinsonian features. Gabapentin (unknown dose) had also been unsuccessful in the treatment of tongue tremor. Three years before evaluation, she developed freezing and falls, she started using a cane and later a walker. She denied vivid dreaming, constipation, depression, and motor fluctuations. She had no previous history of trauma or exposure to

Correspondence: Irene Litvan, M.D., FAAN, FANA, Tasch Endowed Professor in Parkinson Disease Research, UC San Diego Movement Disorders Center, Department of Neurosciences, Altman Clinical Translational Research Institute, 9452 Medical Center Drive, 2nd floor West 114, La Jolla, CA 92037, Phone: (858) 822-5871, FAX: (858) 822-5743, ilitvan@ucsd.edu.

Ali Shoeibi: 1C, 2A, 2B, 3A
Irene Litvan: 1A, 1B, 1C, 2A, 2B, 2C, 3B

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psychoactive or antidopaminergic medications. There was absence of non-motor features or family history of tremor.

On her first examination, blood pressure was normal while lying and standing. Her Montreal Cognitive Assessment showed mild cognitive impairment (20/30) with prominent abnormalities in executive function. Blepharospasm, eyelid opening apraxia, vertical supranuclear ophthalmoparesis (slow hypometric vertical saccades with normal horizontal saccades and horizontal but no vertical optokinetic nystagmus), and absent convergence were noted. She had moderate to severe akineto-rigid symmetric parkinsonism with predominant axial involvement and a 3-4 hertz jaw tremor associated with a 4-5 hertz resting and postural tremor of the tongue (Video). Tongue tremor was continuous and interfering with eating and speaking.

She walked with a walker and was unable to safely stand without assistance. Brain MRI was normal. Her symptoms continued to slowly progress. Table 1 shows the evaluations at the first and last visit four years later.

**Discussion**

This is the first report of a prominent tongue and jaw tremor without palatal and limb tremor in a patient with PSP. However, there are rare reports of palatal tremor in PSP patients. Our patient meets both the William's definition and recently described probable MDS PSP-parkinsonism criteria. She presented with a PD phenotype (bradykinesia and rigidity with axial predominance, and levodopa resistance (A2) that later converted into a PSP-Richardson phenotype (clear limitation of the range of voluntary gaze in the vertical more than in the horizontal plane, O1).

Central oscillators in the Guillain-Mollaret triangle, thalamus, or basal ganglia are thought to generate tongue tremor. We hypothesize that the mechanism of tremor in this patient might be similar to that of palatal tremor, but here involvement of special unknown regions of the red nucleus or central tegmental tracts might produce an autonomous neuronal rhythm generator in the inferior olive which only affects the group of motor neurons of the ambiguous or hypoglossal nuclei which are responsible for tongue and not palatal movements. Although, the patient's brain MRI was normal, the areas mentioned above can be affected in PSP. Likely involvement of unknown rare red nucleus or central tegmental tract areas have caused this patient's isolated tongue and jaw tremor. This type of involvement should be very rare because there are no previous reports of isolated tongue and jaw tremor in PSP patients.

An alternative possibility to consider is that this patient has both, PSP and Parkinson's disease (PD) but this patient does not have the typical non-motor PD features. Moreover, not only the occurrence of prominent tongue without limb and palatal tremor is uncommon in PD, but to our knowledge the association of these two disorders is extremely rare.

**Supplementary Material**

Refer to Web version on PubMed Central for supplementary material.
Acknowledgments

The authors thank the patient and family for their time and for their involvement in the project.

References

Table 1
Evaluations at the first and last visits

<table>
<thead>
<tr>
<th>Clinical finding (clinical scale)</th>
<th>First visit Score in item(s)</th>
<th>Last visit* Score in item(s)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Urinary problems (MDS-UPDRS: item 1.10)</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Chewing and swallowing problems (MDS-UPDRS: item 2.3)</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Speech problems (MDS-UPDRS: item 3.1)</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>Neck rigidity (MDS-UPDRS: item 3.3a)</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>Limb rigidity (MDS-UPDRS: items 3.3b- 3.3e)</td>
<td>2-3</td>
<td>2-3</td>
</tr>
<tr>
<td>Arising from chair (UPDRS: item 3.9)</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>Gait problems (MDS-UPDRS: item 3.10)</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>Freezing of gait (MDS-UPDRS: item 3.11)</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>Postural instability (MDS-UPDRS: item 3.12)</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>Tremor of limbs (MDS-UPDRS: items 3.15a – 3.17 d)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Lip/jaw tremor (MDS-UPDRS: item 3.17e)</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Cognitive state (MoCA)</td>
<td>20/30</td>
<td>16/30</td>
</tr>
</tbody>
</table>

* Four years after first visit

MDS-UPDRS: Movement Disorder Society-Unified Parkinson Disease Rating Scale, MoCA: Montreal Cognitive Assessment