Essential palatal tremor

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DESCRIPTION
A 30-year-old man, without any preexisting comorbidities, presented to us with a longstanding complaint of being able to perceive a ‘click’ in his ears. He found it increasingly difficult to concentrate on tasks. He did not have any other accompanying symptoms such as ataxia or vertigo. He had several ear, nose and throat (ENT) consultations where he was prescribed agents for vertigo and tinnitus but in vain. Brain imaging as well as brainstem auditory evoked response (BAER) tests were unfruitful. Examination of the palate revealed a rhythmic, symmetrical, vertical and oscillating movement of the soft palate at a frequency of 30–50 clicks/min, which produced a clicking sound each time (video 1). It was said to worsen with stress. The movement was not voluntarily suppressible and there were no accompanying signs of brainstem or cranial nerve dysfunction. As per his spouse, the ‘click’ was audible even while he was asleep. Considering normal neurological examination and imaging findings, a diagnosis of essential palatal tremor (EPT) was made. He was successfully treated with oral clonazepam (2 mg/day). The patient refused to take botulinum toxin injection. He was followed up for next 6 months during which he showed partial resurgence of the tremor as the medication was tapered, which necessitated resumption of previous doses of clonazepam along with addition of a new centrally acting agent in sodium valproate (400 mg/day).

Palatal tremor is an uncommon hyperkinetic movement disorder comprising rhythmic oscillations of the soft palate. Owing to its rhythmicity, it was classified as a tremor as opposed to earlier label as a myoclonus, in the First International Congress of Movement Disorders in 1990.1 Subsequently recognised as two types, symptomatic palatal tremor (SPT) and essential palatal tremor (EPT), by Deuschl et al in 1990; there are important clinical and pathophysiological differences between the two.2 Unlike SPT, where clicks are rarely present and associated with neurological deficits like dysarthria and nystagmus, palatal movements and ear clicks are the sole manifestations of EPT. The latter is produced by contraction of the tensor veli-palatini, causing sudden opening and closing of the...
The site of lesion in case of SPT is postulated to be the Guillain–Mollaret triangle (figure 1), comprising the dentate nucleus, red nucleus and the inferior olivary nucleus. Lesions anywhere in the dentato-rubro-olivary pathway cause the olive to get disinhibited resulting in compensatory hypertrophy and synchronous firing of its neurons. These signals are relayed to the cerebellar cortex via climbing fibres resulting in oscillations. Pathophysiology of EPT, on the other hand, is obscure. As per Pearce, a supranuclear insult to the striatum or rostral brainstem resulting in disinhibition of olivary nucleus may be at play. Although a functional origin has rarely been suggested owing to occasional suppressibility and distractability, persistence during sleep ruled out such a possibility in the present case.

Patient’s perspective

I was distressed with this clicking sound and was shocked to see my palate in mirror. I rushed to my family physician and he referred me to the neurologist, who consoled me and treated me.

Learning points

- Palatal tremor is a rhythmic oscillation of the soft palate, which even persists during sleep.
- Essential palatal tremor (EPT) is produced due to contraction of tensor veli-palatini, whereas symptomatic palatal tremor (SPT) is attributed to lesion of the Guillain–Mollaret triangle.
- EPT is accompanied by the characteristic audible click which is rare in SPT.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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REFERENCES