Case report

Oculopalatal tremor and hypertrophic olivary degeneration

W L S Mahapitiya[1], A E S Senanayake[2], Darshana Sirisena[2], Manjula C. Caldera[3]

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Abstract

Background: Oculopalatal tremor is characterized by synchronous occurrence of palatal tremor and pendular nystagmus. Oculopalatal tremor (OPT) follows a destructive lesion in the Guillain-Mollaret triangle (GMT) and is associated with hypertrophic inferior olive degeneration (HOD).

Case presentation: We report a case of 45 years-old patient with palatal tremor and pendular nystagmus without ear click. He has had a brainstem stroke 11 months back. He had right side pyramidal drift, left side lower motor neuron type facial weakness and left abducens nerve palsy and mild gait ataxia in addition to oculopalatal tremor. MRI showed hypertrophic inferior olive degeneration.

Conclusion: Ear click is an infrequent finding in symptomatic palatal tremor. Additional clinical features may be present in OPT depending on topography of initial structural lesion. There is a wide variety of time intervals between the occurrence of anatomical lesions and recognition of palatal tremor.

Key words: oculopalatal tremor, pendular nystagmus, Guillain-Mollaret triangle.

Background

Palatal tremor is a rare movement disorder. Initially it was most commonly referred to as “palatal myoclonus”. However, it was subsequently termed as “palatal tremor” considering its continuous, rhythmic nature of the jerks of the soft palate[1]. Palatal tremor is of two types: symptomatic palatal tremor (SPT) and essential palatal tremor (EPT)[2].

Case presentation

A 45-year-old man presented with rhythmic movements in the soft palate for several weeks. He denies clicking sounds in both ears. He was a diagnosed patient with hypertension and has had a stroke 11 months before with sudden onset double vision, mouth deviation to right side and right upper limb and lower limb weakness. CT brain was normal at that time and was managed as an ischemic brainstem stroke at a local hospital. Physical examination revealed rhythmic movements of soft palate and patient was not able to control it voluntarily. Eye examination revealed mild abduction deficit on left eye and pendular nystagmus. There was mild lower motor neuron type facial weakness on left side and a right-side pyramidal drift. His right-side tendon reflexes were exaggerated with extensor plantar response on same side. Left side tendon reflexes and plantar response were normal. His gait was slightly ataxic with tendency to fall to left side. His speech and higher functions were normal.

His blood investigations including full blood count, erythrocyte sedimentation rate, blood picture, liver function tests, renal function tests and electrolytes were normal. Electrocardiogram, echocardiogram and carotid doppler were also unremarkable. MRI brain showed increased T2-weighted signal and enlargement of left inferior olive (Figure 1).

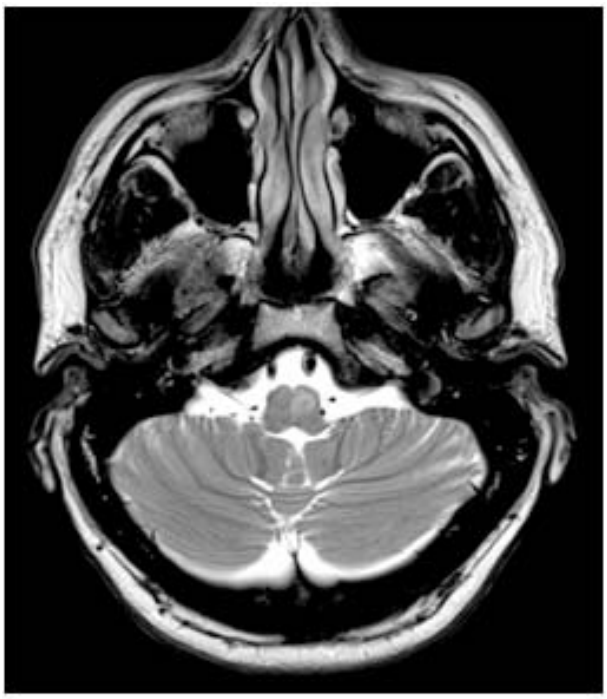

Figure 1. Axial T2-weighted MRI image shows increased signal and enlargement of left inferior olive.

[1]*Department of Neurology, National Hospital, Kandy, Sri Lanka,* [2]*Department of Neurology, National Hospital of Sri Lanka, Colombo, Sri Lanka,* [3]*Department of Neurology, Teaching Hospital Anuradhapura, Sri Lanka.*
Correspondence: W L S Mahapitiya E-mail: shyaman85@gmail.com

Discussion

Rhythmic movements of soft palate in our patient are compatible with palatal tremor. Simultaneously, pendular nystagmus was also observed. His past history of combination of left abducens nerve palsy, lower motor type left facial palsy and contralateral hemiparesis point toward the lesion in the brainstem at pontine level. MRI findings are compatible with hypertrophic inferior olivary degeneration.

Palatal tremor (PT) comprises of two different entities: symptomatic palatal tremor (SPT) and essential palatal tremor (EPT). Palatal tremor may be unilateral, bilateral or asymmetrical[3]. Palatal movement in EPT results from activity of the tensor veli palatini muscle innervated by trigeminal nerve[4,5]. Tensor veli palatini muscle is attached to the lateral wall of the Eustachian tube and its rhythmic contraction is thought to be associated with repetitive opening and closing of the Eustachian tube, leading to the ear clicks[5,6]. In SPT, there is contraction of levator veli palatine muscle innervated by motor fibres from the facial nucleus or nucleus ambiguous causing palatal movements[4,5]. Levator veli palatine movement is also associated with ear click but it is not seen frequently[5]. Therefore, presence of ear clicks alone cannot differentiate between EPT and SPT.

Palatal tremor and ear click are the sole manifestations in EPT whereas in SPT, additionally, there may be ocular disorders and limb and gait ataxia. Oculopalatal (OPT) tremor refers to synchronous occurrence of palatal tremor and pendular nystagmus[4,7]. Pendular nystagmus is found to be present in 30% of SPT[2]. Vertical pendular nystagmus with varied combinations of torsional and horizontal components are typical for OPT[8,9]. Pendular nystagmus is most frequently asymmetric and dissociated in direction in the two eyes[10]. While PT is mostly asymptomatic, patients with pendular nystagmus may have decreased visual acuity and disturbing oscillopsia[4,10].

The site of the abnormality in EPT is unknown. However, SPT/OPT is believed to arise from destructive lesions in Guillain-Mollaret triangle (GMT), a triangle formed by the contralateral dentate nucleus, the ipsilateral red nucleus and the ipsilateral inferior olivary nucleus (ION)[4,7]. Afferent axons from contralateral dentate nucleus travel through contralateral brachium conjunctivum, cross the midline, turn around ipsilateral red nucleus, and descend in the ipsilateral central tegmental tract to reach the ipsilateral ION[4,7]. Efferent axons from ION cross midline, pass through contralateral inferior cerebellar peduncle to reach contralateral deep cerebellar nuclei. Central tegmental tract lesions are specifically associated with OPT compared to lesions of dentate nuclei/brachium conjunctivum where only PT is observed[8,10]. The most frequent etiology of structural lesions in brainstem or cerebellum is vascular, particularly hemorrhagic or ischaemic strokes. Other less common etiologies include brain trauma, brainstem tumours, surgical or gamma knife removal of brainstem cavernoma and multiple sclerosis[5,11].

SPT or OPT has been described in association with the anatomical observation of hypertrophic inferior olive degeneration (HOD)[4,12,13]. Olivary hypertrophy generally results from transynaptic deafferentation and loss of the inhibitory input from the contralateral dentate nucleus[5,12]. The hypertrophy of the ION appearing over a few months after the disruption of afferent fibers is accompanied by vacuolar changes in neurons and gliosis [5,12,13]. Neurons in the ION eventually become atrophied. However, gross olivary hypertrophy is constant[12,13]. These changes appear on MRI as signal changes on T2-weighted images and olivary hypertrophy with or without contrast enhancement. Increased olivary signal on T2-weighted images first appears approximately 1 month after the initial injury. Olivary hypertrophy develops 4 to 6 months after the acute event and resolved by 3 to 4 years[14,15]. Lesion in the dentate nucleus, superior cerebellar peduncle, or both cause contralateral HOD, but damage to the tegmental tracts leads to ipsilateral HOD[15].

There is a wide variety of time intervals between the occurrence of anatomical lesions and recognition of palatal tremor. It is observed that palatal tremor develops at least 1 month to 8 years after the initial insult (median between 10 and 11 months)[4,13,16], but nystagmus develops much earlier than PT[8]. Once PT or OPT is established, it persists for life[4]. However, few exceptional cases exist where PT or OPT have disappeared after many years[3]. 11 months later, it is noted palatal tremor in our patient.

Additional clinical features may be present in OPT depending on topography of initial structural lesion. Contralateral hemiplegia, contralateral hemi-hypoesthesia or spinothalamic syndrome, ipsilateral facial palsy and ipsilateral kinetic cerebellar syndrome are frequent findings[4]. Patients also frequently have eye movement abnormalities, such as fascicular abducens nerve palsy, internuclear ophthalmoplegia, one and a half syndrome and nuclear abducens syndrome[4,17].

Pendular nystagmus is the most symptomatic consequence of OPT. Hence, most of clinical trial have been mainly performed on acquired pendular nystagmus. Gabapentin and memantine are found to be effective in reducing nystagmus amplitude and frequency irregularity[18]. Some authors have also observed sustained decrease of nystagmus velocity in some patients[4]. Botulinum toxin have been tested for treatment of PT and acquired nystagmus with variable results[19,20,21].

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