Palatal Myoclonus (syn. Palatal Tremor)

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Key Words
Palatal tremor • Palatal myoclonus • Guillain-Mollaret triangle • Hypertrophic inferior olivary nucleus • Neuronal coupling

Abstract
Symptomatic palatal tremor is caused by a lesion in the triangle of Guillain and Mollaret and is associated with hypertrophic olivary degeneration that has multiple causes. Essential palatal tremor has no currently demonstrable cause and no accompanying physical or radiological signs. But it is probable that an organic genesis will become apparent. I suggest that some examples of palatal tremor may depend on an ‘upper motor neurone type’, i.e. supranuclear, lesion in the striatum or rostral brainstem releasing medullary activation with denervation hypersensitivity of olivary neurones.

History
In 1886 H.R. Spencer (1860–1941) described myoclonic movements of the pharynx and larynx, and noted the similarity to nystagmus. The inferior olivary nucleus (‘the olive’), in certain pathological conditions, is apt to show a peculiar type of abnormality whose main feature is a conspicuous enlargement. Early references to such ‘hypertrophy’ of the olive are those of Oppenheim in 1887, Thomas [4], and Marie and Guillain [5]. There was renewed interest in 1928 when van Bogaert and Ber...
trand [6] showed the constant presence of these olivary changes in ‘rhythmic and synchronous’ PM.

Over the next 40 years, Guillain and Mollaret [7], Bender et al. [8], Nathanson [9] and many others reported small groups of patients with PM.

**Clinical Features**

Certain features separate this form of myoclonus from other varieties of myoclonus (table 1). Careful study in the above case report and in the patients described by Nathanson [9] have shown that the myoclonus can persist during phonation and sleep in symptomatic PM. Intravenous barbiturates which can abolish nystagmus do not affect PM, nor is it influenced by caloric stimulation, the inhalation of carbon dioxide or the advent of coma. It may persist indefinitely but is not a cause of disability, although patients may complain of a ‘clicking’ sound in the ear, made by the spasmodic contractions of the levator veli palatini part of whose origin is from the cartilaginous auditory tube. Associated disordered, synchronous eye movements include nystagmus, vertical and pendular or horizontal and torsional or hypermetropic saccades: so-called ‘oculopalatal myoclonus’. PM may be also accompanied by synchronous contractions in the oropharynx, larynx, oesophagus and diaphragm.

**Aetiology**

The aetiology in about 70% of cases has been a vascular infarct. Multiple sclerosis, metastatic and astrocytic tumours, syringobulbia, and trauma have been invoked as occasional causal lesions. Of 287 cases with PM from the literature 75% were symptomatic and no cause was found in the 25% ‘essential’ cases. Myoclonic contractions are located in the levator veli palatini muscle (facial or nucleus ambiguous innervation) in symptomatic, and in tensor veli palatini (trigeminal innervation) in ‘essential’ cases. MRI may be normal, but a hyperdense signal in the region of the inferior olive on T2 or proton density images is often seen in symptomatic PM [10].

A heterogeneous syndrome of progressive ataxia with palatal tremor and dysarthria has been reported [11]. Rare instances of an association with Alexander’s disease and Rosenthal fibre encephalopathy [12], supranuclear palsy, and, prominent bilateral activation of the putamen [13] have been associated with PM.

**Possible Mechanisms**

Palatal myoclonus (syn. palatal tremor) is of two types: symptomatic and essential. (1) Symptomatic palatal tremor is caused by a lesion in the triangle of Guillain and Mollaret and is associated with hypertrophic olivary degeneration visible on MR images. (2) Essential palatal tremor has no currently demonstrable cause, and no accompanying physical or radiological signs, and is almost certainly heterogeneous.

Experimental stimulation or destruction of the inferior olivary nucleus produces PM in the monkey. In 1931, Guillain and Mollaret [7] described anatomical features related to PM. These comprised the dentate nucleus, red nucleus and inferior olivary nucleus – referred to as the Guillain-Mollaret triangle (fig. 1) [14, 15]. The dentatorubral-olivary pathway comprises efferents from the dentate nucleus which ascend through the superior cerebellar peduncle, cross the midline in the brachium conjunctivum to the red nucleus, and descend via the central tegmental tract to the inferior olivary nucleus. Inferior olivary efferent fibres cross the midline in the inferior

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**Table 1. Features of essential versus symptomatic palatal myoclonus**

<table>
<thead>
<tr>
<th></th>
<th>Essential PM</th>
<th>Symptomatic PM</th>
</tr>
</thead>
<tbody>
<tr>
<td>Main muscle involved</td>
<td>tensor veli palatini</td>
<td>levator veli palatini</td>
</tr>
<tr>
<td>Other signs</td>
<td>no</td>
<td>ocular and/or pharyngeal myoclonus, + ataxic syndrome</td>
</tr>
<tr>
<td>Ear clicking</td>
<td>infrequent</td>
<td>frequent</td>
</tr>
<tr>
<td>Disappears in sleep</td>
<td>yes</td>
<td>no</td>
</tr>
<tr>
<td>MRI</td>
<td>usually no lesion</td>
<td>medullary lesion</td>
</tr>
<tr>
<td>Electrophysiology</td>
<td>abnormal polysynaptic brainstem reflexes</td>
<td>abnormal monosynaptic, oligosynaptic and polysynaptic reflexes</td>
</tr>
</tbody>
</table>

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cerebellar peduncle, terminating in the original dentate nucleus (fig. 1) [16, 17].

In 1935, Julio Óscar Trelles (1904–1990), a neurologist from Lima, Peru, reported that isolated lesions of the inferior cerebellar peduncle do not cause PM because there are no direct connections between the inferior olive and the contralateral dentate nucleus. When damaged they were associated with olivary enlargement [17].

The site of the lesion is of interest. In all pathologically studied cases of symptomatic PM it has involved the dentate nuclei of the cerebellum, the central tegmental tract, and the inferior olivary nucleus [18] on the opposite side. The inferior olivary nucleus shows marked hypertrophy [19, 20], usually after a delay of 1–6 months; palatal tremor may appear within days or after a delay of up to 30 months after an acute vascular lesion [21]. However, there are recorded instances of comparable olivary hypertrophy not accompanied by PM [21]. Hypertrophy of the olivary nucleus can result from transneuronal degeneration caused by any lesion of the ipsilateral central tegmental tract, the contralateral superior cerebellar peduncle or the dentate nucleus.

The inferior olivary nucleus receives a GABA-ergic input densely concentrated in the red nucleus. The GABA-ergic system inhibits electronic coupling of inferior olivary neurons, and if disrupted may result in hypersynchronous firing of these polysynaptic neurons. This may derive from putaminal, ipsilateral brainstem or contralateral cerebellar lesions, with resulting PM. The neurons of the olive are extensively coupled by gap junctions (‘connexons’), which may relate to their capacity to oscillate when there is loss of normal GABA-ergic inhibition [22]. Speculations of the mechanism consider interruption of inhibitory projections from the deep cerebellar nuclei, via the central tegmental tract, to the inferior olive. Blockade of cerebellar inhibition is followed by olivary hypertrophic degeneration with the development of connexons between cell bodies, causing synchronized oscillation of large groups of inferior olivary nucleus neurons. The synchronized signal is sent to the cerebellar cortex via climbing fibres, producing oscillations [23]. Symptomatic PM and its persistence result from denervation hypersensitivity of olivary neurones starting 1–2 months after the onset, because of the inhibitory blockade.

I suggest that essential palatal tremor may depend on an ‘upper motor neurone type’, i.e. supranuclear, lesion in the striatum or rostral brainstem releasing activation with denervation hypersensitivity of olivary neurones.

References


