

A case of palatal myoclonus

HISTORY

A 72 year-old woman was seen in clinic complaining that she had lost the ability to sing. Associated with this complaint was an 8-year history of dizziness, falling and hearing strange noises in her ears. She had also noticed some mild slurring of speech. There had been a slow progression of symptoms over time, but the initial symptoms appeared to come on rather abruptly.

She had a past history of gastro-oesophageal reflux and took omeprazole. She was on no other drugs and there was no family history.

EXAMINATION

Systemic examination was normal. Neurological examination revealed bilateral rhythmic oscillations of the soft palate (video). Other cranial nerves were normal except rather jerky pursuit eye movements. The limb examination was normal and there were no cerebellar signs.

INVESTIGATIONS

Other than raised serum cholesterol, her routine laboratory tests were normal. MRI of the brain was normal. Further cardiovascular work-up including echocardiography was unremarkable.

DIAGNOSIS

The diagnosis was essential palatal myoclonus.

DISCUSSION

Palatal myoclonus (PM; also known as palatal tremor) is a form of segmental myoclonus characterised by vertical oscillations of the soft palate at a frequency of 1-3 Hz. Movements may be unilateral or bilateral, and other muscles may become

involved, such as pharyngeal, laryngeal, lower facial, eye, neck and respiratory muscles.

PM may be classified as essential (in which no cause is identified) or symptomatic. Symptomatic PM is classically caused by a lesion in Mollaret's triangle (see figure) and appears to arise due to interruption of the dentato-rubral-olivary pathways. The commonest aetiology is ischaemia, although trauma, demyelination or other focal lesions have been described. Pathologically inferior olivary hypertrophy is often seen and generation of symptomatic PM may depend on olivary neuron hyperactivity (released from inhibitory inputs). Persistence of PM in these cases however is likely to be related to other brainstem mechanisms (Nishie et al, 2002)

If restricted to the soft palate (typically essential PM), it may be asymptomatic. Often, however, an annoying clicking noise is heard by the patient, caused by opening and closing of the eustachian tube. Symptomatic PM is usually associated with other brainstem signs and myoclonus may also be seen in limb and trunk muscles. PM often develops with considerable latency between the precipitating clinical event and onset of symptoms.

Treatments include carbamazepine, phenytoin, L-5-hydroxytryptophan, lamotrigine, diazepam, barbiturates and injections of botulinum toxin into palatal muscles.

REFERENCES

Nishie M, Yoshida Y, Hirata Y, Matsunaga M *Generation of symptomatic palatal tremor is not correlated with inferior olivary hypertrophy* (2002) *Brain* 126, 1348-1357



Alastair Wilkins is ACNR's Case Study Co-ordinator. He is Specialist Registrar in Neurology in East Anglia, having trained in Cambridge, Sheffield and London, and has just finished a PhD investigating potential mechanisms of axon loss in multiple sclerosis.



Figure: Mollaret's triangle: triangular area bounded by red nucleus, inferior olive and dentate nucleus of the cerebellum

Video Clip: There is also a video clip on the web site to accompany this case report.

Correspondence address:
Alastair Wilkins, Addenbrooke's Hospital, Cambridge.
E-Mail. aw255@cam.ac.uk