

Palatal Myoclonus-A Case Report

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Abstract

Palatal myoclonus is usually due to a brainstem or cerebellar lesion disrupting the dentato-rubro-olivary pathway. Rarely it may be caused by a cortical lesion. The precipitating factor in 70% of all cases is an infarct.

We describe an unusual case of a patient with palatal myoclonus who had an old ipsilateral cerebellar infarct and a new contralateral subcortical (corona radiata) infarct. We postulate that the new infarct caused disinhibition of the old cerebellar infarct, resulting in palatal myoclonus. Magnetic resonance imaging (MRI) of the brain did not show any hypertrophy of the inferior olivary nucleus. Her myoclonus proved refractory to clonazepam, valproate and phenytoin.

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Introduction

Palatal myoclonus is a rare movement disorder characterised by rhythmic recurring jerky movements of the soft palate, pharynx, larynx and rarely diaphragm, trunk and limb muscles. It usually develops secondary to brainstem or cerebellar disease (symptomatic palatal myoclonus) interrupting the central tegmental tract or dentato-olivary fibres.¹⁻³ Some patients, however, fail to show evidence of a structural lesion (essential palatal myoclonus). The ratio of symptomatic to essential palatal myoclonus is 1.27:1.¹ Movement disorder specialists feel that a more accurate name for palatal myoclonus is palatal tremor because of the rhythmicity of this condition.^{4,5} The characteristic pathological correlate is hypertrophy of the inferior olivary nucleus. Rarely it may be caused by focal cortical lesions^{6,7} causing rhythmic myoclonic jerking or epilepsy partialis continua. Ictal clonic expression involves the palate, tongue, lips, jaw, larynx, hand and arm. We report a patient with palatal myoclonus who had an ipsilateral old cerebellar infarct as well as a new contralateral subcortical (corona radiata) infarct. We postulate that the new infarct caused disinhibition of the old cerebellar infarct, resulting in palatal myoclonus.

Case Report

VM is an 81-year-old Chinese female with multiple

stroke risk factors of atrial fibrillation, hypertension and ischaemic heart disease. She had a left cerebrovascular accident with right hemiparesis 2 years ago prior to this admission. At that time, she was seen by a general practitioner and refused hospitalisation. The patient required some assistance in ambulation but subsequently recovered with good functional recovery. Her medications included aspirin 100 mg om, adalat 10 mg tds and sorbitrate 10 mg tds.

She presented with inability to express herself and right-sided weakness when she awoke one morning. She was well the night before.

On examination she had global aphasia but was otherwise alert. She had preferential gaze deviation to the left. There was a dense right hemiplegia with sensory loss and right-sided, neglect. There was a right extensor plantar response. She was in atrial fibrillation with a heart rate of 60/min.

Investigations including full blood count, urea, electrolytes and thyroid function tests were normal. Chest radiograph showed cardiomegaly. Electrocardiogram showed atrial fibrillation with poor R wave progression. Computed tomography (CT) scan of the head on admission showed an old right cerebellar infarct (Fig. 1). There were no infarcts in the left cerebral hemisphere.

One day after admission she was noted to have rhythmic

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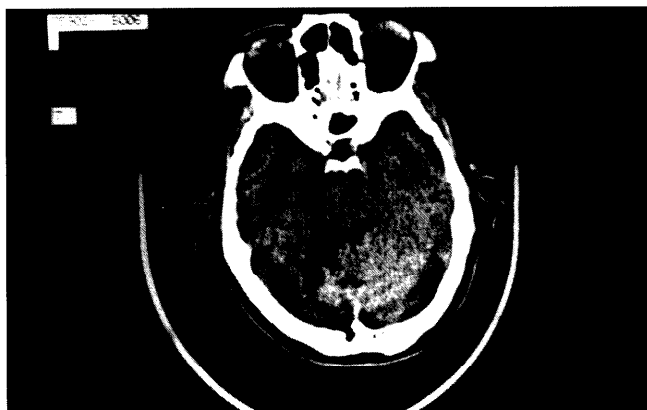


Fig. 1. CT scan of the head shows an old right cerebellar infarct.

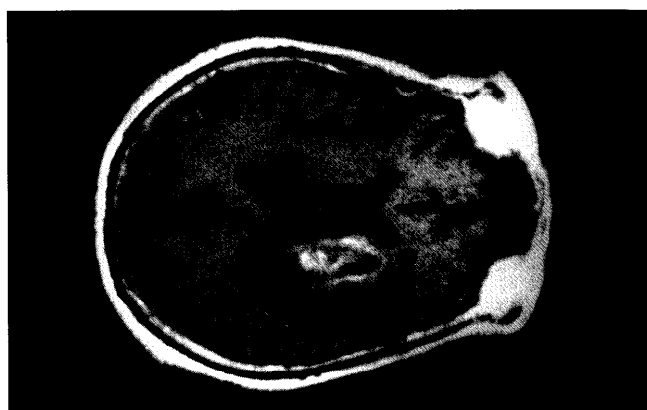


Fig. 2. Axial T1 weighted MRI of the brain shows a haemorrhagic infarct with increased signal in the left corona radiata.

mic clonic movements of the right corner of her mouth including the entire palate, tongue, larynx and right platysma. These occurred continuously at a rate of 150 per minute both during sleep and wakefulness, unaffected by mouth opening. There was no tonic neck deviation and the arms and shoulders were not involved. She remained aphasic and there was no change in consciousness. The patient's husband did not notice any tremor before hospitalisation.

The results of two surface electroencephalograms were normal. She was treated with clonazepam 6 mg a day without improvement. Her medication was subsequently changed to valproate 1.8 g a day. However, the myoclonus persisted and proved refractory to treatment. Addition of phenytoin 300 mg a day with a serum level of 22 mg/l did not result in improvement of myoclonus. Axial T1 weighted MRI of the brain performed 7 days after the onset of stroke (Fig. 2) showed a haemorrhagic infarct with increased signal in the left corona radiata infarct, including the old right cerebellar infarct. There was no hypertrophy of the inferior olivary nuclei.

Discussion

This patient was noted to have rhythmic contractions of the glossopharyngeal musculature after a cerebral infarct. Neither sleep nor proprioceptive factors⁸ like mouth opening or attempts to verbalise could suppress the contractions.

A variety of precipitating events have been reported in palatal myoclonus. Vascular infarcts account for about 70% of the cases. Other conditions include multiple sclerosis, encephalitis, neoplasm, hereditary tremor, syphilis, malaria, vertebral artery aneurysms and trauma.

The pathology of palatal myoclonus is rather well established due to the pioneering work of Klien (1907), Guillain and Mollaret (1931), Lapresle and Ben Hamida (1965, 1968), Schenck (1965), Llinas (1984), Llinas and

Yarom (1986), Llinas and Muhlethaler (1988) and many others.

There are two possible causes for palatal myoclonus: the first is an ipsilateral cerebellar infarct causing disruption of the dentato-rubro-olivary pathway. This pathway travels from the dentate nucleus through the superior cerebellar peduncle and crosses to the opposite side in the commissura of Werneck along the internal and dorsal surface of the red nucleus, before it reaches the inferior olivary nucleus via the central tegmental tract.¹⁵ One of the gross pathological features of palatal myoclonus is a hypertrophic degeneration of the inferior olivary nuclei secondary to denervation. Histopathologic changes in the inferior olivary nucleus are neuronal cell body enlargement, vacuolation of the nerve cells, fibrillary gliosis, demyelination and astrocyte proliferation of the white matter. The lesion is considered to be trans-synaptic in origin, as it is mostly associated with a supra-olivary causal lesion, the two preferential sites being the ipsilateral tegmental tract and the contralateral dentate nucleus. Hypertrophic degeneration occurs over a period of weeks or months and palatal rhythmic movements are usually not observed before hypertrophy develops.¹ These hypertrophic cells are regarded to represent the pace-maker for the tremor. The patient's husband did not notice any tremor before her current hospitalisation.

The second cause of palatal myoclonus is partial status epilepticus. Although this had been reported by Soheyil et al⁶ and Tatum et al,⁷ we do not think that this was the aetiology in this patient for the following reasons: firstly, the facial area has a relatively large representation on the motor homunculus and no seizure activity was noted on 2 electroencephalograms.^{9,10} Secondly, epileptic phenomenon of cortical origin may lead to focal clonic movements known as jacksonian march involving the hand, arm and face. Such movements were absent in this patient. Thirdly, the rhythmic contractions failed to respond to anticonvulsants. In addition, the site of the

lesion in our patient was subcortical (corona radiata) rather than cortical. Both patients reported by Soheyli et al⁶ and Tatum et al⁷ had electroencephalogram patterns consistent with *epilepsia partialis continua*. In addition, palatal myoclonus was accompanied by ipsilateral jerking of the arms. Soheyli et al⁶ reported improvement of palatal myoclonus with clonazepam and phenytoin whereas Tatum et al⁷ found limited success with anticonvulsants.

There are two possible pathways⁷ for the cortex to produce palatal myoclonus. Firstly, corticobulbar fibres from the frontal cortex may descend to bilaterally innervate reticular nuclei. These send efferents to the cerebellum and nucleus ambiguus, which innervates the palatal musculature. It is possible that the subcortical infarct acted to further disinhibit the cerebellar infarct, resulting in palatal myoclonus. Secondly, frontal cortical efferents directly innervate the inferior olivary nucleus. Hence, cortico-olivary fibres may activate palatal myoclonus.

Our patient illustrates disinhibition of an old ipsilateral cerebellar infarct by a new contralateral subcortical focus, resulting in palatal myoclonus.

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REFERENCES

1. Deuschl G, Mischke G, Schenck E, Schulte-Monting J, Lucking C H. Symptomatic and essential rhythmic palatal myoclonus. *Brain* 1990; 113:1645-72.
2. Herrmann C, Brown J W. Palatal myoclonus: a reappraisal. *J Neurol Sci* 1967; 5:473-92.
3. Fitzgerald D C. Palatal myoclonus-Case report. *Laryngoscope* 1984; 94:217-9.
4. Shibasaki H. Myoclonus. *Curr Opin Neurol* 1995; 8:331-4.
5. Deuschl G, Toro C, Valls-Sole J, Zeffiro T, Zee D S, Hallet M. Symptomatic and essential palatal tremor. I. Clinical, physiological and MRI analysis. *Brain* 1994; 117:775-88.
6. Soheyli N, Alois E, Otto W W, Rudiger J S. Palatal tremor of cortical origin presenting as *epilepsia partialis continua*. *Epilepsia* 1995; 36:207-9.
7. Tatum W O, Sperling M R, Jacobstein J G. Epileptic palatal myoclonus. *Neurology* 1991; 41:1305-6.
8. Tomkinson A, Craven C, Brown M J K M. Palatal myoclonus affected by neck position. *J Laryngol Otol* 1995; 109:61-2.
9. Thomas J E, Reagan T J, Klass D W. *Epilepsia partialis continua*. *Arch Neurol* 1977; 34:266-75.
10. Juul-Jensen P, Denny-Brown D. *Epilepsia partialis continua*. *Arch Neurol* 1966; 15:563-78.