Glucose utilization and symptomatic ocular myoclonus

Yakushiji et al. studied FDG-PET in a patient with symptomatic ocular myoclonus before and after successful treatment. The glucose utilization in the inferior cerebellar vermis decreased with the clinical improvement.

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Ocular oscillations and the inferior olive

Commentary by R. John Leigh, MD

Since Guillain and Mollaret defined the syndrome of oculopalatal myoclonus or tremor in 1931, it has remained a neurologic mystery.¹ Affected patients develop spontaneous oscillations that variably affect the eyes, palate, and other branchial muscles during the weeks following brainstem or cerebellar stroke (see video clip on the *Neurology* Web site at www.neurology.org). The characteristic pathologic finding is hypertrophic degeneration of the inferior olivary nucleus.

The inferior olivary nucleus contributes to motor learning, although there is debate as to whether this is achieved by providing an error signal or by synchronizing the timing of cerebellar Purkinje neuron discharge. A distinctive feature of inferior olivary nucleus neurons is electrotonic coupling between their dendrites due to gap junctions (connexons).

It is postulated that the syndrome of oculopalatal tremor arises because of interruption of inhibitory projections from the deep cerebellar nuclei, via the central tegmental tract, to the inferior olive.² Loss of cerebellar inhibition is followed by hypertrophic degeneration with development of connexons between cell bodies, causing large groups of inferior olivary nucleus neurons to synchronize, creating a 1- to 2-Hz oscillator. This synchronized signal is sent to the cerebellar cortex on climbing fibers, perhaps stimulating the cerebellar motor learning network to produce a maladaptive response consisting of pendular ocular oscillations.

Yakushiji et al. provide novel and important findings to test this hypothesis. Their patient with ocular oscillations following pontine hemorrhage showed

increased regional cerebral metabolic rate of glucose utilization (rCMRGlu) on PET scanning in the right inferior olivary nucleus. Treatment with clonazepam suppressed the ocular oscillations, and this was associated with decreased rCMRGlu in the contralateral cerebellar vermis but not in the inferior olivary nucleus. Thus, the pathogenesis of oculopalatal myoclonus or tremor is becoming clearer with two potential treatments: blocking the maladaptive cerebellar response (as in this report) or by giving drugs that block gap junctions in the inferior olivary nucleus.

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