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MR Imaging of Presumed Olivary Hypertrophy in Palatal Myoclonus

Rhythmic palatal myoclonus is a neurologic sign with one of the most precise pathologic correlations. It almost always is associated with hypertrophic degeneration of the inferior olivary nucleus [1]; however, such hypertrophic degeneration is not always accompanied by palatal myoclonus. The abnormality of the olivary nucleus is thought to be the result of a transsynaptic degeneration due to a lesion either in the ipsilateral central tegmental tract or in the contralateral dentate nucleus. We present a case that we think illustrates the MR detection of hypertrophic degeneration of the inferior olivary nucleus [2].

Case Report

A 68-year-old woman had a right-sided lateral gaze palsy, ipsilateral nuclear facial palsy, contralateral hemihypesthesia, cerebellar incoordination, ataxic gait, insomnia, and complex visual and auditory hallucinations. CT scan was normal, but MR imaging showed an infarct in the middle of the tegmentum pontis, larger on the right side. The patient had a partial recovery.

Two years later she had palatal myoclonus extending to the orbicularis oris, platysma, mentalis, and geniohyoideus muscles. MR imaging confirmed the old ischemic lesion in the right pontine tegmentum (Figs. 1A and 1B), and T2-weighted images showed a hyperintense lesion limited to the ipsilateral medullary olive (Fig. 1C).

Discussion

Our patient had had a stroke of the pontine tegmentum, which had been diagnosed on the basis of clinical findings and confirmed by MR. An infarct in this area would be expected to affect the central tegmental tract producing the ipsilateral hypertrophic degeneration of the inferior olivary nucleus and the palatal myoclonus. The long interval between the stroke and the beginning of the myoclonus is not unusual [1]. The high-intensity lesion on MR corresponds to the location of the inferior olivary nucleus and not to any medullary vascular territory, making the possibility of a new infarct unlikely.

The neuropathologic characteristics of the hypertrophic degeneration (macroscopic enlargement often to the point of being visible to the naked eye) and the high water content of the enlarged and vacuolated neurons and hypertrophic astrocytes allow detection by MR. Nevertheless, some patients with palatal myoclonus have had normal MR findings. This may be due to a lack of imaging capability. Another possibility is that palatal myoclonus becomes clinically evident as soon as the physiologic disturbance is established, which may occur before the appearance of neuropathologic changes and, therefore, before radiologic detection. In our case, MR showed a signal abnormality in the olivary nucleus but did not show convincing evidence of olivary hypertrophy; this may correspond to an intermediate step in the neuropathologic degeneration of the olives before the late hypertrophic final stage. The pathologic basis of the degeneration is the same, whether it is associated with focal lesions of the brainstem or cerebellum or with the degenerative syndrome of progressive ataxia and palatal myoclonus. In cases of the latter, hypertrophic degeneration of the inferior olivary nucleus has been shown on MR images [2, 3].

Neither our case nor any of those previously published has included postmortem findings. Until such confirmation is available, the MR findings can contribute to a better understanding of the natural history of hypertrophic degeneration and palatal myoclonus and prevent confusion of this degeneration with other ischemic or neoplastic conditions.

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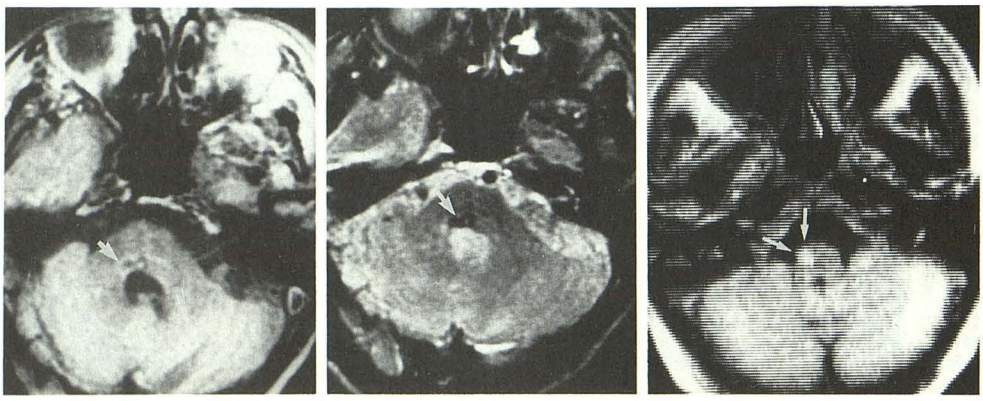

Fig. 1.—Presumed olivary hypertrophy in palatal myoclonus.

***A* and *B*, MR images, 2000/25/2 (*A*) and 2000/100/2 (*B*), at mid level of pontine tegmentum show abnormal signal (*arrows*) of old ischemic lesion, containing hemosiderin, in right tegmentum.**

***C*, Axial T2-weighted MR image, 2000/60/4, at medullary level shows hyperintense signal (*arrows*) in anatomic location of right inferior olivary nucleus.**