Letters to the Editors Related to New Topics

Choreodystonia in a Patient with Hypertrophic Olivary Degeneration after Pontine Tegmental Hemorrhage

Although hypertrophic olivary degeneration (HOD) is usually associated with rhythmic palatal tremor (PT), arrhythmic movements, such as Holmes tremor, can also be associated with HOD,^{1–3} even in the absence of PT.^{4–6} Here, we describe a patient who developed choreodystonia following pontine hemorrhage and HOD.

In August 2004, a 75-year-old man was admitted with a sudden-onset of right hemiparesis, dysarthria, and horizontal diplopia. Neurological examination revealed horizontal gaze paralysis, mild right hemiparesis, and mild dysarthria. Initial brain CT and MRI showed parenchymal hemorrhage in the left pontine tegmentum (Fig. 1A,B) with a normal-appearing olivary nucleus. His symptoms improved slowly.

Repeated MRI performed in March 2005 (Fig. 1C,D) revealed marked hypointensity intermixed with hyperintensity in the left pontine tegmentum on T2-weighted images. The hypointense lesion extended rostrally to the lower midbrain. In addition, an enlarged left anterior inferior medulla with high signal intensity suggestive of HOD was also observed.

In November 2006, abnormal hyperkinetic movement began to develop in the patient's right hand, and it gradually progressed to involve the entire limb. In January 2007, neurological examination showed slow and irregular involuntary movement combined with dystonic posturing of the right arm (see Video). Choreic movement of the fingers was noted in the right hand at rest. When walking, pronation of the right arm, accompanied by extension and abduction of the right shoulder, was observed along with intermittent rapid pronation of the right forearm. Intermittent oral dyskinesia was also observed. Muscle tone was increased in the right arm and leg. He had no Parkinsonian symptoms. No palatal or ocular tremor was observed. Sensory exam was normal. Repeated MRI performed in January 2007 showed no significant differences from the MRI results obtained in March 2005, except for slight regression of HOD.

His involuntary movement followed a defined vascular event and HOD as revealed by MRI. Therefore, it is logical to suspect that the patient's involuntary movement was related to the pontine hemorrhage and HOD. A noteworthy feature of our patient was localized chorea combined with dystonic posturing in the absence of rhythmic tremor, such as PT. Pseudoathetosis secondary to proprioceptive sensory loss occurring in thalamic stroke⁷ may be considered as a possible pathomechanism of the choreodystonia in our patient. However, his sensory exam was normal, and no new lesion was found in the left thalamus on follow-up MRI. One may argue that involuntary movement in the right hand is actually a phasic dystonia rather than a chorea. Indeed, it can be very difficult or even impossible to distinguish between these two involuntary movements. However, we thought that the patient had chorea as well as dystonia because his distal finger movements were not patterned.

HOD has traditionally been considered to be a pathologic substrate of PT. However, the literature indicates that the structural lesion responsible for the development of HOD is not necessarily the lesion that generates PT. First, although virtually all patients who develop PT after a brainstem insult will have HOD, not all patients with HOD develop PT.5 Second, in addition to PT and rhythmic tremor in adjacent structures, HOD may be accompanied by Holmes tremor,1-6 which usually results from damage to the cerebello-thalamic and nigrostriatal system.8 Furthermore, dystonic posturing has been reported with HOD, even in the absence of PT.^{4,6} It has not been determined whether all of these involuntary movements directly result from the hyperactivity of olivary neurons or whether there is an undetected common etiology that causes both HOD and involuntary movements to occur simultaneously. In any case, these evidences show that the pathomechanism of HOD can be associated with a wide range of involuntary movements.

The pathogenesis of the choreodystonia in our patient is uncertain. Some studies suggest that damage to the cerebellothalamic pathway and the development of the pathologic neuronal circuit may be associated with secondary chorea and dystonia.^{7,9} In addition, olivocerebellar dysfunction and damage to the inferior olive afferents arising from the midbrain, which descends within the central tegmental tract, have been implicated in secondary dystonia.¹⁰ Therefore, it can be postulated that the pontine tegmental hemorrhage observed in our patient damaged a structure that is involved in the development of chorea and dystonia and caused damage to the dentatorubroolivary pathway, resulting in HOD.

The delay between the pontine hemorrhage and the emergence of choreodystonia in our patient was relatively long in comparison with the reported time delays for the onset of PT or other involuntary movements associated with HOD.^{2–6} However, it is not surprising considering that the delay between the insult and the onset of involuntary movements can be longer than 2 years, especially for hyperkinetic movements because of the damage to the cerebello-thalamic pathway.^{7,9} The delay between the insult and onset of involuntary movements may indicate that it takes time for the pathologic neuronal circuit responsible for the involuntary movements to develop after the injury.

LEGEND TO THE VIDEO

Segment 1. The video shows a slow and irregular involuntary movement combined with dystonic posturing of the right arm.

This article includes supplementary video clips, available online at http://www.interscience.wiley.com/jpages/0885-3185/suppmat.

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FIG. 1. A,B: Initial T2-weighted axial MRI shows parenchymal hemorrhage in the left pontine tegmentum with a normal-appearing inferior olive. C,D: Follow-up MRI 7 months after hemorrhage shows T2-weighted hypointensity intermixed with hyperintensity in the left pontine tegmentum and HOD in the left inferior olivary nucleus.

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