Case Report

Hypertrophic Olivary Degeneration and Cerebrovascular Disease: Movement in a Triangle

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Hypertrophic olivary degeneration is a rare kind of trans-synaptic degeneration that occurs after lesions of the dentatorubro-olivary pathway. The lesions, commonly unilateral, may result from hemorrhage due to vascular malformation, trauma, surgical intervention or hypertension, tumor, or ischemia. Bilateral cases are extremely rare. This condition is classically associated with development of palatal tremor, but clinical manifestations can include other involuntary movements. We describe 2 cases: unilateral hypertrophic olivary degeneration in a 60-year-old man with contralateral athetosis and neurologic worsening developing several years after a pontine hemorrhage and bilateral hypertrophic olivary degeneration in a 77-year-old woman with development of palatal tremor, probably secondary to pontine ischemic lesions (small vessel disease). **Key Words:** Hypertrophic olivary degeneration—stroke—palatal tremor—athetosis.

Hypertrophic olivary degeneration (HOD) is a rare condition and a particular kind of trans-synaptic degeneration. It occurs after lesions of dentatorubro-olivary pathway causing olivary hypertrophy. The lesions, commonly unilateral, may result from hemorrhage, ischemia, trauma, surgical intervention, or tumors.[1] Bilateral HOD is extremely rare.[1-2] Clinical findings include palatal tremor, dentatorubral tremor, ocular myoclonus, choreodystonia, ataxia, dysarthria, and diplopia.[1-5]

Case Reports

Case 1

A 60-year-old man complained of involuntary movements in his right hand, which gradually developed over the previous 5 years, accompanied by dysarthria and gait difficulty. He had a brain stem hemorrhage 15 years before, from which he recovered almost completely. Examination revealed a cerebellar speech, slight abduction paresis of the left eye, bilateral intention tremor, and right-sided finger–nose dysmetria. During posture and action, right upper limb, distal predominant, slow writhing movements could be seen and were consistent with athetosis (Video 1). No palatal tremor was observed. Brain magnetic resonance imaging

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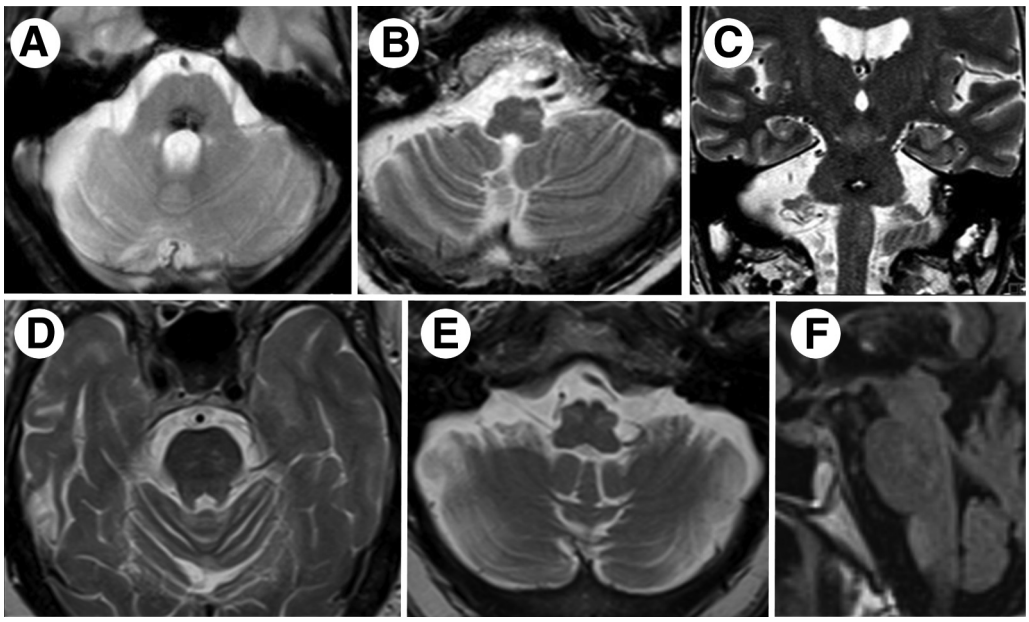

Figure 1. *Brain magnetic resonance imaging: Patient 1 (A-C) and patient 2 (D-F). (A) Axial gradient-echo T2 shows tegmental median pontine hypointensity characteristic of cavernous angioma. (B) Axial T2-weighted imaging shows hyperintensity and hypertrophy of the left inferior olivary nucleus. (C) Coronal T2-weighted imaging shows cavernous angioma and hyperintensity of left inferior olivary nucleus. (D) Axial T2-weighted imaging shows hyperintense foci suggestive of sequelae of ischemic lesions in pons, involving tegmental portion. (E) Axial T2-weighted imaging shows bilateral hyperintensity and hypertrophy of the inferior olivary nucleus. (F) Sagittal T2-weighted imaging shows hyperintensity of the inferior olivary nucleus.*

demonstrated left hypertrophic olivary degeneration and tegmental median pontine cavernous angioma (Fig 1, A-C).

Case 2

A 77-year-old woman, hypertensive, complained of dysphagia and dysphonia that gradually developed over the previous 2 years after a severe traumatic brain injury. Examination revealed palatal tremor. She did not complain of ear clicks. Brain magnetic resonance imaging (Fig 1, D-F) demonstrated bilateral hypertrophic olivary degeneration, sequelae of ischemic lesions in the tegmental portion of the pons, superior cerebellar peduncles, capsular and thalamic, and areas of encephalomalacia in left temporal and right frontal lobes.

Discussion

In the first case, HOD was secondary to pontine hemorrhage and associated with late-onset neurologic worsening. This condition is classically associated with development of palatal tremor, although not always present.[3] In this patient, we observed a long time interval for development of athetosis and suggest it is a manifestation of HOD. We argue that secondary inferior olive nucleus lesion led to disruption in the fine process of motor control and could produce manifestations classically related to dysfunction in the complex network connecting motor cortex and basal ganglia.

In the second case, the etiology is unclear, but it was probably secondary to pontine ischemic lesions (small vessel disease) rather than secondary to traumatic injury. Reports of bilateral HOD are extremely rare, and it was present in this patient in whom lesion involved both the central tegmental tract and the superior cerebellar peduncle.

It is important to recognize HOD to avoid misinterpretation and to enable appropriate clinical management decisions.

Supplementary Data

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.jstrokecerebrovasdis.2014.09.036.

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