Hypertrophic olivary degeneration: A case report

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Abstract
Hypertrophic olivary degeneration is a rare occurrence in which different pathological processes including enlargement and vacuolation of the neurons, demyelination of the white matter, and fibrillary gliosis of the inferior olivary nucleus take place. It mostly develops secondary to a destructive lesion involving the Guillain–Mollaret pathway. The mostly reported destructive lesions causing hypertrophic olivary degeneration are stroke, trauma, tumors, neurosurgical interventions, and gamma knife treatment of brainstem cavernoma. It presents with symptomatic palatal tremor, and typically appears as an expansive nonenhancing nodular lesion that shows increased signal intensity on magnetic resonance imaging (MRI). The identification of hypertrophic olivary degeneration on MRI is of great importance as its MRI appearance is very similar to those of more severe pathologies, including tumors, infarction, demyelinating lesions, and infections. We present a case of hypertrophic olivary degeneration in a patient with a history of ischemic stroke two years before the development of palatal tremor.

Key words: Guillain–Mollaret triangle; hypertrophic olivary degeneration; magnetic resonance imaging

Introduction
Hypertrophic olivary degeneration (HOD) is a rare occurrence in which different pathological processes including enlargement and vacuolation of the neurons, demyelination of the white matter, and fibrillary gliosis of the inferior olivary nucleus take place. It mostly develops secondary to a destructive lesion in brainstem or cerebellum involving the Guillain–Mollaret pathway. This pathway is located in a triangle-shaped anatomical region of which the three corners are formed by inferior olivary nucleus, ipsilateral red nucleus, and contralateral dentate nucleus [Figure 1]. The mostly reported destructive lesions causing HOD are stroke, trauma, tumors, neurosurgical interventions, and gamma knife treatment of brainstem cavernoma.[1]

Case Report
A 62-year-old male admitted to the hospital with the complaints of intermittent clicking in the throat, which persists during sleep, and deficiency in the eye movements. He had a history of stroke two years prior to the admittance. On physical examination, palatal tremor and abducens nerve palsy were recorded. The patient was then referred to the Department of Radiology for a brain magnetic resonance imaging (MRI). MRI demonstrated extensive areas of encephalomalacia in the posteromedial portions of the left cerebral hemisphere, posterolateral portions of both thalami and in the left cerebral peduncle [Figure 2A and B].

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region of the left half of the medulla oblongata, which was hyperintense on T2-weighted and fluid attenuation inversion recovery (FLAIR) images [Figure 2C and D]. The lesion was indistinguishable on T1-weighted images, and did not enhance following contrast administration. No evidence of restriction of diffusion was observed on diffusion weighted images and apparent diffusion coefficient (ADC) mapping [Figure 3]. The lesion was causing expansion of the brainstem [Figure 4]. Combining the history and clinical and imaging findings, the patient was diagnosed as having HOD.

**Discussion**

HOD, a special type of degeneration characterized by nuclear enlargement rather than atrophy, develops weeks or months following the occurrence of a destructive lesion within the Guillain–Mollaret pathway. It occurs as a result of the denervation of olivary neurons by the disruption of the dentatorubral or the rubro-olivary pathways. Denervated neurons enlarge and develop oscillations, which are then modulated by cerebellum causing abnormal motor output in the branchial arches.[1] This type of degeneration is unique in which it causes hypertrophy rather than atrophy of the nucleus. It clinically presents with symptomatic palatal tremor resulting from the rhythmic contractions of the levator veli palatini muscle. These contractions cause unintentional movements of the soft palate and pharynx, which continue during sleep. Palatal tremor develops 1 month to 8 years after the occurrence of the destructive lesion, and persists for life. According to the location of the destructive lesion, different neurological manifestations including contralateral hemiplegia/hemihypoesthesia or spinothalamic syndrome, ipsilateral facial palsy or kinetic cerebellar syndrome, abducens nerve palsy, and internuclear ophthalmoplegia may accompany palatal tremor.

MRI is the method of choice in demonstrating HOD. It typically appears as an expansive nonenhancing nodular lesion, which shows increased signal intensity on T2-weighted and FLAIR images.[2] The signal increase in the olive is visible nearly one month after the onset of palatal tremor, whereas the enlargement of the nucleus takes approximately six months to appear.[1] In the cases with cerebellar lesions, HOD appears in the contralateral
side, whereas it appears in the ipsilateral side in the cases with central tegmental tract lesions. The enlargement of the olivary nucleus resolves in about three to four years; however, the hyperintensity persists for life. In some rare cases, also the increased signal of the nucleus may resolve.

The identification of HOD on MRI is of great importance as its MRI appearance is very similar to those of more severe pathologies including tumors, infarction, demyelinating lesions, and infections. The lack of contrast-enhancement of HOD helps differentiating it from malignant tumors and inflammation. And nuclear enlargement is not a usual finding in cases with demyelinating lesions and chronic infarcts. Diffusion tensor imaging and MR fiber tractography are shown to be useful in demonstrating the disruption of the Guillain–Mollaret pathway in the cases with unclear conventional MRI findings.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

**References**