Bilateral Involvement in Hypertrophic Olivary Degeneration

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Summary

Hypertrophic olivary degeneration (HOD) is a form of trans-synaptic degeneration with hypertrophy of the inferior olivary nucleus in response to neurologic insult to the dentatorubro-olivary pathway. In the central nervous system, the degeneration of an anatomical structure is usually characterized by neuronal loss replaced by proliferation of glial elements. Unique to the inferior olivary nucleus is transneuronal degeneration resulting in hypertrophy of the targeted region. HOD is a relatively rare disorder that may mimic many diseases including tumors and demyelinating processes. Herein we report on a case of HOD with bilateral involvement and we present the follow-up findings.

Key words: Hypertrophic olivary degeneration, HOD, Guillian-Mollaret triangle, dentatorubro-olivary pathway

INTRODUCTION

Hypertrophic olivary degeneration (HOD) is a degenerative disorder of the inferior olivary nucleus that occurs after damage to the dentatorubro-olivary pathway. Bilateral involvement is extremely rare. Herein we report on a case of HOD with bilateral involvement and we present the follow-up findings.

A 46-year-old man presented to emergency room with headache and vertigo. He was conscious and had normal orientation. Neurological examination showed bilateral vertical optokinetic nystagmus, restricted horizontal look to the right and the left but unrestricted look up and down. Restriction in bilateral horizontal conjugate gaze. Vertical gaze was normal. He had bilateral mild palatal myoclonus; the palatal arc was normal but he could not get his tongue out. He had dysarthric speech, dysdiadochokinesia and ataxia. He could not do the coordinated movements. No motor deficit or lateralized paresis was
found. He walked with assistance because of ataxia. He mentioned that he had experienced a short period of unconsciousness which was attributed to pontine hemorrhage 6 months earlier.

Magnetic resonance imaging (MRI) disclosed T2 hypointense, faintly T1 hyperintense spicular contoured, oval lesion at the pontine tegmentum. T1 weighted post contrast image showed partial contrast enhancement in the lesion while gradient echo T2 weighted image revealed exaggerated signal loss due to magnetic susceptibility effect. The lesion was placed predominantly at the dorsal right side of the tegmentum and extended to left partially. There was some increased T2 signal around the lesion. These findings suggested a cavernoma with probable previous hemorrhage. Sections through the bulbus level showed that both of the inferior olivary nuclei had increased signal on T2 weighted images. Furthermore the right nucleus was enlarged. The abnormally increased T2 signal was continuous between the pontine and the bulbar lesions. There was no contrast enhancement and no restricted diffusion. Altogether these findings were interpreted as bilateral hypertrophic olivary degeneration associated with pontine cavernoma. A small meningioma of 1 cm in diameter in the right cerebello-medullary angle was considered coincidental because it simply protruded into the cistern without any contact with the brainstem.

After the diagnosis of HOD the patient received phenytoin 100 mg daily. He underwent a 6-month follow up imaging which showed no remarkable change. The follow up neurological examination showed clear improvement in palatal myoclonus. There was slight improvement in dysarthric speech, dysdiadochokinesia and ataxia. He was still walking with assistance.

![Image](https://example.com/image1)

**Fig 1:** MR images at the level of bulbus. A, Axial T2-weighted image at the level of the medulla shows hyperintensity of both olivary nuclei (arrows) and expansion of the right medullary olive. The cerebellopontine angle meningioma is marked with 'asterix'. B, Contrast-enhanced axial T1-weighted image at the same level shows enhancement in the meningioma (asterix). There is partial contrast enhancement in the olivary lesions (arrow).
Fig 2: MR images at the level of pons. A, Axial gradient echo T2 weighted image shows magnetic susceptibility effect of predominantly right sided dorsal tegmental cavernoma (arrow). B, Axial fast spin echo T2 weighted image at the same level. The hypointensity is predominant peripherally, producing a "rim shape". The central part is relatively hyperintense. Note some increased signal around the cavernoma (arrows) likely representing gliosis from previous hemorrhage. C, T1 weighted image shows foci of hyperintensity within the lesion (arrows), typical of a cavernoma.

Fig 3: Drawing shows the triangle of Guillian and Mollaret.
DISCUSSION

Hypertrophic olivary degeneration is a form of trans-synaptic degeneration with hypertrophy of the inferior olivary nucleus in response to neurologic insult to the dentatorubro-olivary pathway. These dentatorubral olivary connections were first described by Guillian and Mollaret in 1931 in association with oculopalatal tremor and HOD⁸.

The pathway, known as the Guillian-Mollaret triangle, connects the dentate nucleus of the cerebellum with the fibers ascending to the contralateral red nucleus in the superior cerebellar peduncle (Fig 3). These fibers decussate within the brachium conjunctivum and synapse in the contralateral red nucleus of the midbrain. The fibers from the red nucleus descend in the central tegmental tract to the inferior olivary nucleus of the medulla. The olive then sends fibers to the contralateral dentate nucleus via the inferior cerebellar peduncle, completing the triangle. Lesions that disrupt the afferent pathways to the olive (dentatorubral and rubro-olivary pathways) result in HOD⁸.

Lesions in the contralateral cerebellum and superior cerebellar peduncle or the ipsilateral brain stem involving the red nucleus and central tegmental tract can cause HOD. Infarcts, hemorrhages, traumatic brain injury, tumor, and surgery for vascular lesions have all been reported as causes of HOD⁴,⁵. When both the central tegmental tract or superior cerebellar peduncle are involved, bilateral involvement results.

In the central nervous system the degeneration of an anatomical structure is usually characterized by neuronal loss replaced by proliferation of glial elements. Unique to the inferior olivary nucleus is transneuronal degeneration resulting in hypertrophy of the targeted region². Pathologically, cell body enlargement, vacuolation of the cytoplasm, demyelination, astrocytic proliferation and fibrillary gliosis have been described which follows 15-20 months after the onset of primary lesion²,³.

Common clinical findings associated with HOD include palatal myoclonus, dentatorubral tremor, ocular myoclonus and have been described since 1886. Palatal myoclonus usually develops 10-11 months after primary lesion, although it may not always be seen⁴. Diagnosis of HOD on MR imaging is made by the presence of a T2-hyperintense nonenhancing olivary lesion in association with another lesion in the afferent pathway of the olive at Guillian-Mollaret triangle.

In our patient, a small cavernoma was present in the pontine tegmentum which lies predominantly at the right side. There was some MRI evidence of prior hemorrhage and no past history of surgery. We assumed that this cavernoma with its previous hemorrhage component was the cause of the HOD. In a metaanalysis the hyperintensity associated with HOD appeared as early as one month after the brainstem injury. The authors stated that the hyperintensity may persist for several years. The hypertrophic reaction on the other hand usually appears 10 to 18 months after the insult while cases as early as 6 months after the injury have also been reported⁴. It seems that the bulbar hyperintensity and hypertrophic olivary change appeared 6 months after the brainstem injury in our patient. It is likely that the HOD in our patient developed as a result of hemorrhage together with the cavernoma rather than the cavernoma alone.

The various differential diagnoses of signal hyperintensity on T2 weighted images at the brainstem includes tumours, demyelinating lesions, infarction, and inflammatory processes. The demyelinating, ischemic or inflammatory lesions tend to show evolutilional changes within days and weeks, which is
characterized by swelling in the acute phase and shrinkage and persistence of high signal in the late phase. Auffray-Calvier E. et al. described evolution of HOD in 12 patients and observed hyperintensity of the olive on proton density and T2-weighted images as early as 3 weeks after the ictus which persisted up to 3.5-13 years\(^7\). Kitajima M. et al followed 11 patients-eight with pontine tegmental hemorrhages and three with cerebellar hemorrhages in the dentate nuclei described hyperintense areas of the olive appeared 3 weeks after ictus. Hypertrophy appeared after 5-15 months\(^5\).

The lack of contrast enhancement in HOD also helps to exclude some inflammatory and contrast enhancing tumoral processes. Amyotrophic lateral sclerosis and adrenoleukodystrophy demonstrate hyperintensity of corticospinal tracts but not the inferior olivary nucleus\(^6\). The bilateral HOD is extremely rare. However familiarity with the clinical and radiological findings, which are very characteristic, would preclude many unnecessary diagnostic tests.

Tegmental cavernoma in our patient was on the central tegmental tract. The abnormal high signal areas around the cavernoma extended to the right superior cerebellar peduncle. This extension may be the cause of bilateral olivary nucleus disease rather than unilateral involvement. The high signal of the left olivary nucleus without enlargement suggests that it happened recently.

Bilateral mild palatal myoclonus and bilateral vertical optokinetic nystagmus in our patient are both the clinical findings of Guillain-Mollaret triangle disease. Dysarthric speech, dysdiadochokinesia and ataxia are all cerebellar findings. All these findings can be explained by damage to the superior cerebellar peduncle or feed-back pathway of cerebellum which pass through the inferior olivary nucleus using Guillain-Mollaret triangle. Damage of rubrocerebellar-dentatorubral fibers may cause extremity ataxia, dysmetria and dysdiadochokinesia.

In conclusion HOD is a relatively rare disorder that may mimic many diseases including tumors and demyelinating processes. Recognition of the imaging findings of this rare entity may preclude unnecessary examinations and interventions.

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