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Case Report

Case of Bilateral Hypertrophic Olivary Degeneration with a Short Review of the Literature

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Abstract

We report a rare case of bilateral hypertrophic olivary degeneration in a 68-year male patient secondary to lacunar infarcts in the midbrain. MRI examination of the brain without contrast was done as an investigation for gradual onset progressive neurological symptoms like difficulty in speech, imbalance, and ataxia. A history of hypertension and recurrent episodes of minor stroke were noted. MRI showed hypertrophy and T2 signal hyperintensity in bilateral inferior olivary nucleus suggesting hypertrophic olivary degeneration and associated multiple lacunar infarcts. In this case report, we discuss the distinct imaging appearance of hypertrophic olivary degeneration on MRI concerning its temporal evolution. The condition is bilateral making it more interesting. Neuroanatomy and pathology correlation, literature review, and discussion of relevant differential diagnosis reinforces the case and adds overall academic value.

Keywords: *Inferior Olivary Nucleus; Hypertrophic Olivary Degeneration; Gillian-Mollaret Triangle; Palatal Myoclonus*

Abbreviations

HOD: Hypertrophic Olivary Degeneration; MRI: Magnetic Resonance Imaging

Introduction

Hypertrophic olivary degeneration (HOD) is a rare condition that may mimic various other diseases ranging from benign to malignant. On MRI, a non-enhancing focal lesion exclusively limited to the inferior olivary nucleus, that does not show restricted diffusion or contrast enhancement is the classic finding. Anatomical knowledge of the triangular neuronal circuit described by Guillain-Mollaret holds the key for thorough understanding and interpretation of the images. We present a rare case of bilateral hypertrophic olivary degeneration in 68 years old man due to multiple lacunar infarcts in the midbrain disrupting the neuronal integrity of the dentato-rubro-olivary circuit.

Case Report

A 68 years old male patient visited our imaging center for MRI examination of the brain with a clinical history of gradual onset difficulty in speech, imbalance, and ataxia. The patient suffered from hypertension and recurrent episodes of stroke. Eight months before the current clinical presentation he had experienced an event of stroke which needed admission to the hospital. He now presents with worsening of speech, imbalance, and difficulty for coordinated movements for two months. On examination, he was conscious, alert and well oriented. He had nystagmus, bilateral palatal myoclonus, and dysarthria. He needed help to walk. MRI imaging of the brain without administration of intravenous contrast showed T2 hyperintense signal and enlargement of bilateral inferior olivary nuclei (Figure 1A, 1B).

There were small multiple old lacunar infarcts in midbrain at the level inferior colliculus corresponding to the level of the decussation of the superior cerebellar peduncles (Figure 2A, 2B). Supratentorial periventricular white matter showed multiple lacunar infarcts. There was a mild diffuse loss of cerebral white matter with thinning of gyri and widening of sulcal spaces keeping with mild diffuse cerebral atrophy (Figure 3A, 3B).

There was neither hydrocephalus, mass lesion nor features of demyelination. No abnormal areas of restricted diffusion were seen in the brain. Clinical features indicated multi-infarct state without concerns for ongoing infection. The combination of the patient's history, clinical findings, and MRI findings suggested bilateral hypertrophic olivary degeneration with background brain showing multiple lacunar infarcts and mild diffuse cerebral atrophy.

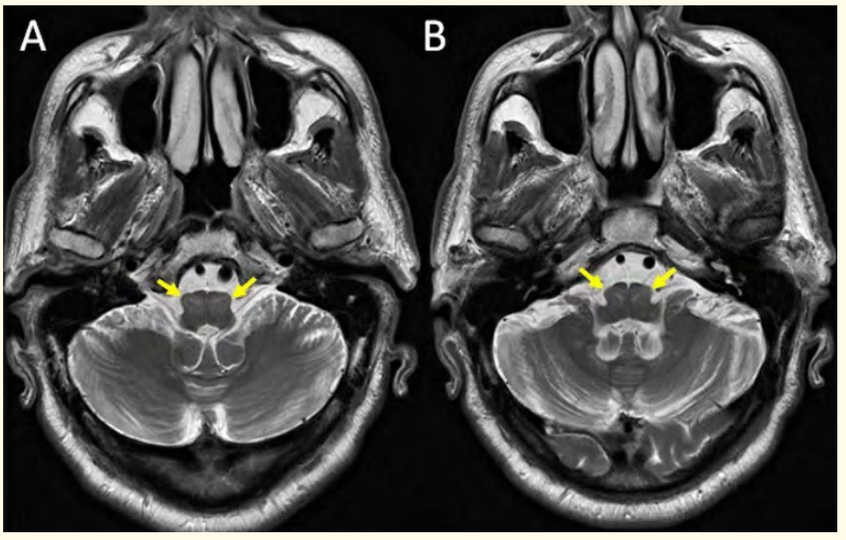

Figure 1: *(A) T2W Axial MRI image of the brain at the level of medullary olives show symmetrical increased signal and enlargement of bilateral inferior olivary nuclei within the medulla (arrows). (B) T2W Axial MRI image at the level of inferior cerebellar peduncles and medulla show symmetrical increased signal and enlargement of bilateral inferior olivary nuclei within the medulla (arrows).*

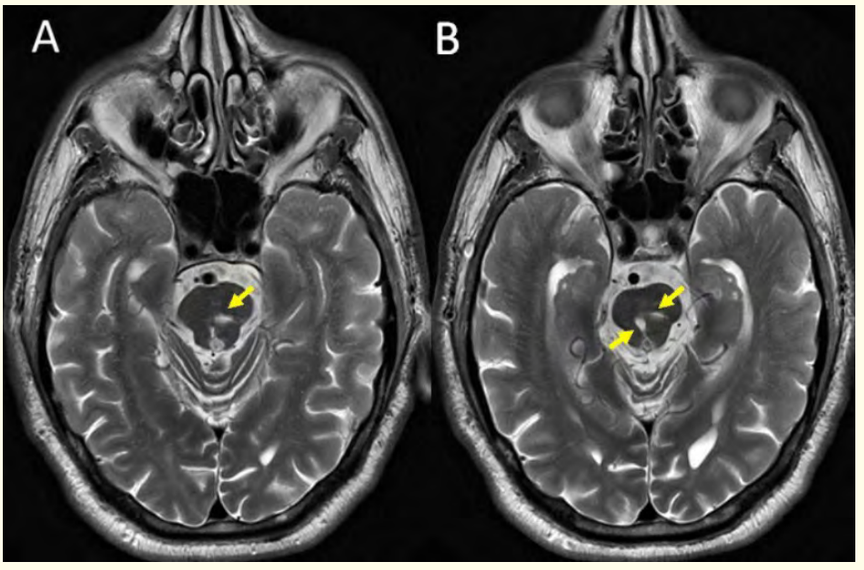

Figure 2: *T2W Axial MRI image of the midbrain at the level of inferior colliculus shows hyperintense signals in the left para-midline region corresponding to the lacunar infarcts of decussation of the superior cerebellar peduncles (arrow). (B) T2W Axial MRI image of the midbrain, slightly above the level of inferior colliculus shows two hyperintense signals in the para-midline region corresponding to the lacunar infarcts at the decussation of the superior cerebellar peduncles (arrows).*

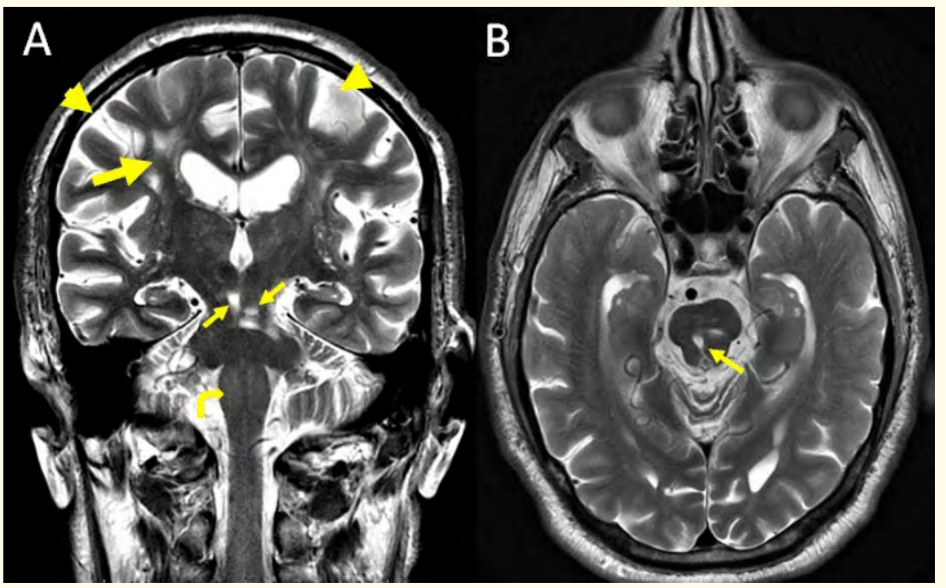

Figure 3: *(A) T2W Coronal MRI image of the brain at the level of the red nucleus shows hyperintense signals in the bilateral para-midline region corresponding to the decussation of the superior cerebellar peduncles (arrows). A diffuse cerebral volume loss is also noted as shown by sulcal widening and thinning of gyri (arrowheads). Multiple deep white matter lacunar infarcts are seen (thick arrow). Bilateral olivary hypertrophy and signal changes are also seen (curved arrow). (B) Axial T2W image of the midbrain, slightly inferior to the level of inferior colliculus shows two hyperintense signals in the bilateral para-midline region corresponding to infarcts at decussation of the superior cerebellar peduncles (arrow).*

Discussion

Hypertrophic Olivary Degeneration is a rare type of neuronal degeneration caused by the disruption of the dentato-rubro-olivary pathway or the triangle of Guillain-Mollaret [1]. It affects all age group, men, and women alike. The dentato-rubro-olivary path is a triangular neural network connecting red nucleus and the inferior olivary nucleus through the ipsilateral central tegmental tract [2]. The inferior olivary nucleus of the medulla is attached to the contralateral dentate nucleus of the cerebellum through the inferior cerebellar peduncle. Superior cerebellar peduncle completes the triangle by connecting dentate and the red nucleus (Figure 4).

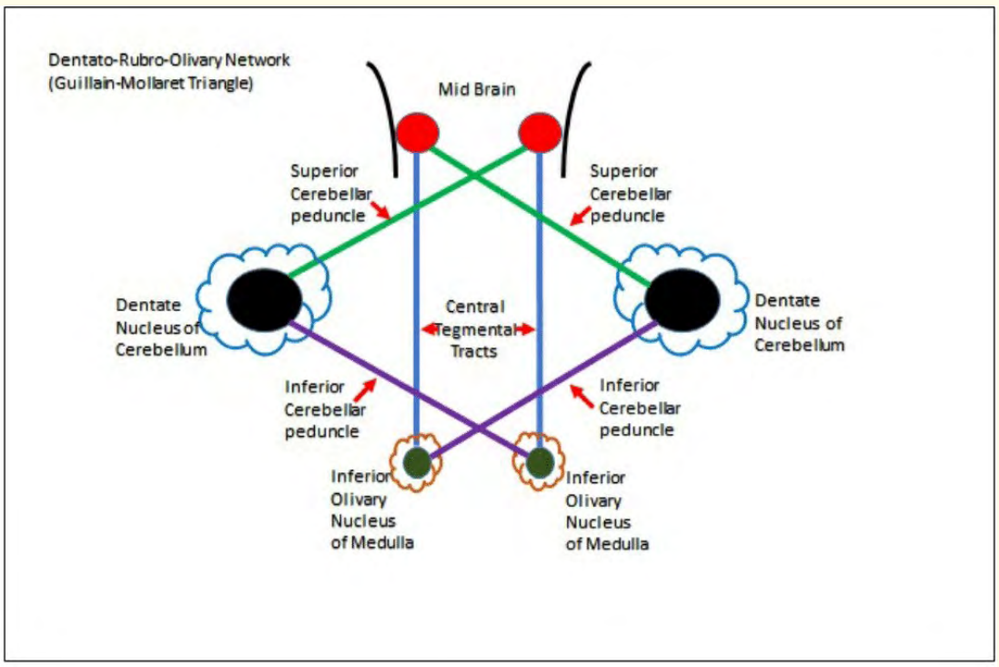

Figure 4: *Schematic diagram of the Dentato-Rubro-Olivary network.*

Hypertrophic Olivary Degeneration is end-result of a variety of lesions that interrupt the Guillain-Mollaret triangle. Ischemia, infarction, tumor, demyelination, hemorrhage, trauma, and surgery are known causes [3]. Knowledge of the anatomical connections of the triangle of Guillain and Mollaret is vital for understanding how lesions can affect the olivary nucleus (Figure 4). HOD is unilateral when the injury involves the red nucleus or central tegmental tract. The contralateral olivary nucleus is engaged when the lesion affects the dentate nucleus or superior cerebellar peduncle. HOD is bilateral when injuries involve the decussation of the superior cerebellar peduncles and seen when the superior cerebellar peduncle and one-sided central tegmental tract are injured [3]. Recent large cohort studies show that in 44% of the patients with HOD, no lesions were found disrupting the triangle of Guillain-Mollaret, thus hypothesizing as idiopathic or injury being outside the triangle or an injury within the triangle far beyond the resolution of MRI [4,5]. Pathologically, HOD is a form of trans-synaptic degeneration of inferior olivary nucleus of the medulla oblongata. It is unique because it results in enlargement of affected structure rather than atrophy. It is due to vacuolar degeneration of the denervated neurons followed by astrocyte hypertrophy, neuronal enlargement, and gliosis [6]. Goto and co-workers [7], have described six stages of HOD based on pathological evolution as described below:

- Stage 1: No change is seen in first 24 hours.
- Stage 2: Degeneration of the white matter of the olivary capsule in 2 to 7 days.
- Stage 3: Mild hypertrophy of the olivary nucleus without glial reaction in three weeks.
- Stage 4: Olivary enlargement is seen with hypertrophy of neurons and astrocytes that occurs between eight to nine months.
- Stage 5: Beyond nine and half months there is olivary pseudo hypertrophy with neuronal dissolution and gemistiocytic astrocytes.
- Stage 6: Is a stage of olivary atrophy seen after 3 to 4 years.

Clinical Findings

Clinically, palatal myoclonus has been described as the most classic sign [3]. This finding can be seen from 2 to 40 months [3]. Other manifestations include abnormal speech, ear clicking, abnormal movements of neck, ataxia, and ocular myoclonus. It is to be noted that HOD is not symptomatic in all cases [3].

Imaging Findings

MRI is the investigation of choice. It is interesting to note that the MRI signal changes correspond to pathological evolution [8]. There are three phases described on MRI. The first phase shows signal change without hypertrophy of the olivary nucleus. It occurs after one month of injury and can be seen on fluid-sensitive sequences like T2W images and FLAIR images. The second phase shows hypertrophy of the nucleus with hyperintensity of the Olives on FLAIR / T2W images. Hypertrophy is seen after six months of injury and persists for 3-4 years. In the third phase the Olivary nucleus show atrophy or return to average size, but signal change continues. In all stages, the lesions show neither contrast enhancement nor restricted diffusion. Advanced imaging with diffusion tensor imaging and tractography may reveal the injury to the connections of a Guillain-Mollaret triangle [9]. Although signal changes may resolve over time, the symptoms may persist [10]. Medical management is the mainstay of treatment although the symptoms can be resistant [11].

Differential diagnosis

Various conditions can cause signal changes and swelling of the olivary nucleus. Hence a broad differential diagnosis can be offered [12]. Infarction, demyelination, primary tumour, metastasis, and infection are the most common differentials. Temporal (evolutional) changes within days and weeks should prompt diagnosis of demyelination, ischemia, or inflammatory lesions. Unlike HOD, Tumour, infection, and inflammatory lesions show post contrast enhancement. The absence of diffusion restriction distinguishes an acute infarction or acute cerebritis. Other clues like exclusive localization of the signal to the olivary nucleus and sparing surrounding tissue should also help.

Conclusion

Although rare, HOD is potentially diagnosable on MRI, provided the differential diagnoses are carefully ruled out. The Radiologist should do an active search for an inciting lesion in the triangle of Guillain-Mollaret. Familiarity with the clinical and radiological findings will lead to a proper diagnosis and prevents many unnecessary interventions and diagnostic examinations.

Conflict of Interest

There are no conflicts of interest.

Bibliography

1. Kitajima M., *et al*. "Hypertrophic Olivary Degeneration: MR Imaging and Pathologic Findings". *Radiology* 192.2 (1994): 539-543.
2. Smets Gitte., *et al*. "The Dentato-Rubro-Olivary Pathway Revisited: New MR Imaging Observations Regarding Hypertrophic Olivary Degeneration". *Clinical Anatomy* 30.4 (2017): 543-549.
3. Eetvelde Ruth Van., *et al*. "Imaging Features of Hypertrophic Olivary Degeneration". *Journal of the Belgian Society of Radiology* 100.1 (2016): 71.
4. Gu Chris N., *et al*. "MRI Findings in Nonlesional Hypertrophic Olivary Degeneration". *Journal of Neuroimaging* 25.5 (2015): 813-817.
5. Carr Carrie M., *et al*. "Frequency of Bilateral Hypertrophic Olivary Degeneration in a Large Retrospective Cohort". *Journal of Neuroimaging* 25.2 (2014): 289-295.
6. Goyal Mayank., *et al*. "Hypertrophic Olivary Degeneration: Metaanalysis of the Temporal Evolution of MR Findings". American Journal of Neuroradiology 21.6 (2000): 1073-1077.
7. Goto N and M Kaneko. "Olivary Enlargement: Chronological and Morphometric Analyses". *Acta Neuropathologica* 54.4 (1981): 275-282.
8. Tilikete Caroline and Virginie Desestret. "Hypertrophic Olivary Degeneration and Palatal or Oculopalatal Tremor". *Frontiers in Neurology* 8 (2017): 302.
9. Hirano Miki., *et al*. "Hypertrophic Olivary Degeneration Resulting from Posterior Fossa Masses and Their Treatments". *Clinical Imaging* 39.5 (2015): 787-790.
10. Krings T. "Hypertrophic Olivary Degeneration Following Pontine Haemorrhage: Hypertensive Crisis or Cavernous Haemangioma Bleeding?" *Journal of Neurology, Neurosurgery and Psychiatry* 74.6 (2003): 797-799.
11. Bouz Peter., *et al*. "The Pathophysiological Basis for Hypertrophic Olivary Degeneration (HOD) Following Brainstem Insult". *JSM Neurosurgery and Spine* 1.1 (2013): 1004.
12. Elnekiedy Abdelaziz., *et al*. "MRI and Neurological Presentation of Hypertrophic Olivary Degeneration". *The Egyptian Journal of Radiology and Nuclear Medicine* 47.3 (2016): 1019-1029.

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