Journal of Clinical Imaging Science

Editor-in-Chief: Vikram S. Dogra, MD
Department of Imaging Sciences, University of Rochester Medical Center, Rochester, USA

OPEN ACCESS
HTML format

For entire Editorial Board visit : www.clinicalimagingscience.org/editorialboard.asp

www.clinicalimagingscience.org

CASE REPORT

Hypertrophic Olivary Degeneration - A Report of Two Cases

Radha Sarawagi, Aravind Murugesan

Department of Radiology, Mahatma Gandhi Medical College and Research Institute, Puducherry, India

Address for correspondence:
Dr. Radha Sarawagi,
HIG C Block, Flat No- 10, Peoples Campus, Bhanpur Road,
Bhopal - 462 037,
Madhya Pradesh, India.
E-mail: radhasanjeevgupta@gmail.com

Received : 24-12-2014
Accepted : 16-01-2015
Published : 30-01-2015

ABSTRACT

Hypertrophic olivary degeneration (HOD) is seen following lesions in the Guillain–Mollaret triangle. This is unique because the inferior olivary nucleus hypertrophies following degeneration unlike the typical atrophy seen in other structures. We report two cases of HOD in two different clinical scenarios.

Key words: Anterolateral medulla, Guillain–Mollaret triangle, hypertrophic olivary degeneration, T2-hyperintensity

INTRODUCTION

Hypertrophic olivary degeneration (HOD) is a transneuronal degeneration of the inferior olivary nucleus in which patients develop hypertrophy of the inferior olivary nucleus following a primary lesion in the dentato-rubro-olivary pathway. This degeneration is unique; in that, it causes hypertrophy of the inferior olivary nucleus rather than atrophy.[1-3] It presents on magnetic resonance (MR) imaging as hyperintensities on T2-weighted images. Familiarity with this entity is important in avoiding mistaking it for one of the other differentials for T2-hyperintensities in the anterolateral medulla, particularly in postoperative patients where it could be mistaken for tumor recurrence.[1] We report two cases of HOD in two different clinical scenarios.

Video Avaiable on www.clinicalimagingscience.org

Access this article online

Quick Response Code:

Website: www.clinicalimagingscience.org

DOI: 10.4103/2156-7514.150454

CASE REPORTS

Case 1

A 42-year-old male who had undergone suboccipital craniotomy and excision of a fourth ventricular epidermoid cyst was subjected to a magnetic resonance (MR) study of the brain for postoperative follow-up 5 months following the surgery. Although the patient had no new neurological deficit, he had continuing dysarthria over the preceding 2 months. MR examination revealed postoperative changes in the posterior fossa with evidence of gliosis around 4th ventricle [Figure 1]. MRI also revealed focal, bilateral, symmetric, ill-defined, T2-hyperintensities involving the olivary region of the medulla with mild hypertrophy [Figure 2]. No evidence of restricted diffusion was seen. No abnormal enhancement was seen on

This article may be cited as:
Sarawagi R, Murugesan A. Hypertrophic Olivary Degeneration - A Report of Two Cases. J Clin Imaging Sci 2015;5:8.
Available FREE in open access from: http://www.clinicalimagingscience.org/text.asp?2015/5/1/8/150454

postcontrast study. On video laryngoscopic examination, the patient had rhythmic, involuntary contractions of the soft palate consistent with palatal myoclonus [video]. A second MR examination of the brain done 19 months following surgery showed a reduction in the hypertrophy and hyperintensity of the olivary region [Figure 3].

Video 1: 42-year-old male with bilateral hypertrophic olivary degeneration following excision of a fourth ventricular epidermoid cyst. Video clip shows palatal myoclunus.

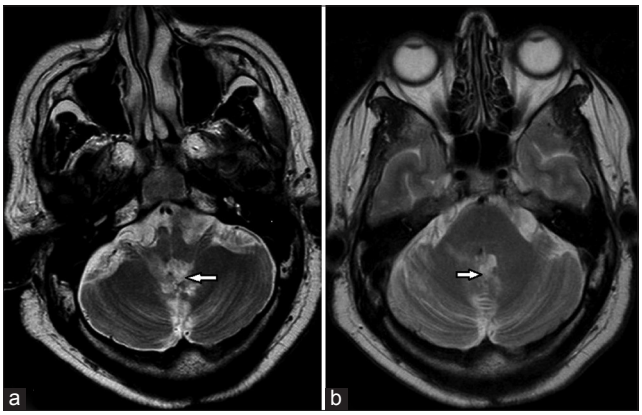

Figure 1: Case 1. 42-year-old male with bilateral hypertrophic olivary degeneration following excision of a fourth ventricular epidermoid cyst Axial. T2-weighted images through fourth ventricle (a and b) show postoperative gliosis (white arrows).

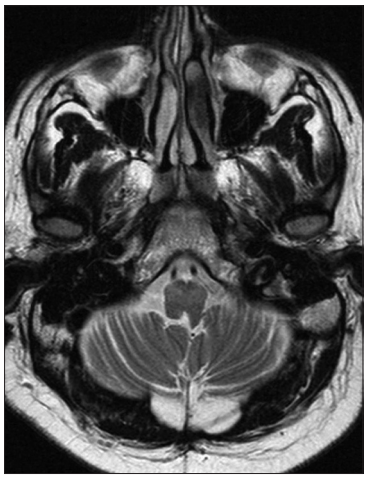

Figure 3: Case 1. 42-year-old male with bilateral hypertrophic olivary degeneration following excision of a fourth ventricular epidermoid cyst. Axial T2-weighted image 14 months later through same level as 2(b) shows reduction in hypertrophy and hyperintensity.

Case 2

A 49-year-old male presented with sudden onset left hemiparesis and slurring of speech. Computed tomographic (CT) examination of the brain revealed an acute hemorrhage involving the pons and midbrain along with old lacunar infarcts and gliotic changes in the heads of both caudate nuclei. Three months later, the patient presented with gradual worsening of dysarthria, which was intermittent, over the preceding 2 weeks. MR examination revealed HOD as a small, ill-defined, nonenhancing T2-hyperintensity involving the olivary region of the medulla on the right side with mild hypertrophy [Figures 4 and 5b]. No evidence of restricted diffusion was seen. Features of the primary lesion were also seen as chronic hemorrhage involving the pons, midbrain [Figure 5], dorsal aspect of the right lentiform nucleus and the adjacent posterior limb of the internal capsule. Although this patient had worsening dysarthria, palatal myoclonus was not demonstrated on video laryngoscopic examination.

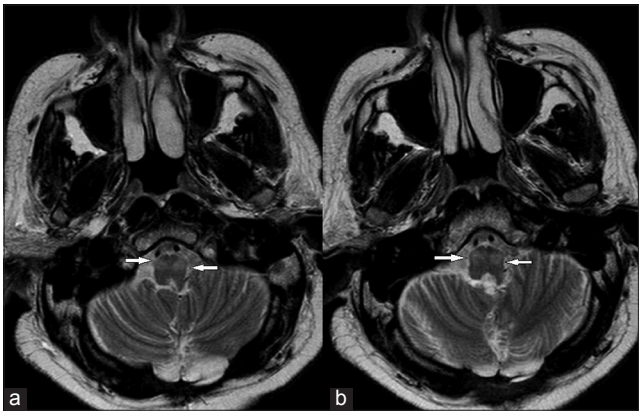

Figure 2: Case 1. 42-year-old male with bilateral hypertrophic olivary degeneration following excision of a fourth ventricular epidermoid cyst Axial. T2-weighted images (a and b) through medulla show bilateral hyperintensities in olivary region with mild hypertrophy (white arrows).

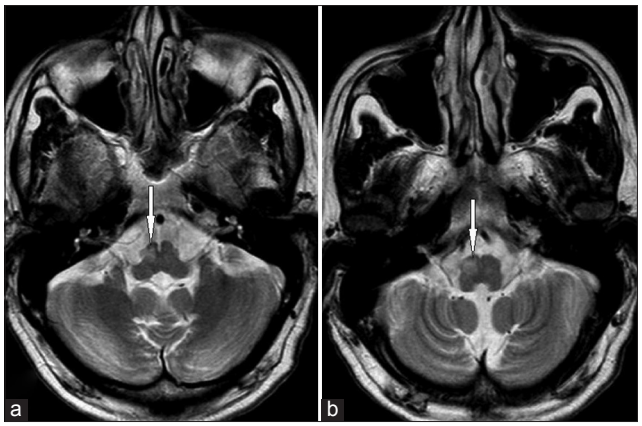

Figure 4: Case 2. 49-year-old male with unilateral HOD following brainstem hemorrhage. (a) and (b) T2-weighted axial images through medulla show ill-defined hyperintensities in olivary region on right side with mild hypertrophy (white arrows).

These two cases illustrate two different situations in which HOD can be seen. Both of them had in common, a primary lesion involving the Guillain–Mollaret triangle. Few cases of HOD have been reported following surgery of a posterior fossa epidermoid cyst and few cases of bilateral involvement have been reported.[1,4]

DISCUSSION

The Guillain–Mollaret triangle is an anatomic triangle connecting the dentate nucleus of one side with the contralateral red nucleus and inferior olivary nucleus [Figure 6]. The dentate nucleus is connected with the contralateral red nucleus by the dentato-rubral tract, while it is connected with the contralateral inferior olivary nucleus through the inferior cerebellar peduncle. The central tegmental tract connects the ipsilateral red nucleus and inferior olivary nucleus.[1-5]

Transneuronal degeneration of the inferior olivary nucleus occurs following its deafferentation by interruption of the dentato-rubral or the rubro-olivary pathways [Figure 6b]. This degeneration is unique; in that, it causes hypertrophy of the olivary nucleus rather than atrophy.[1-3] Patients generally develop slurring of speech following HOD due to palatal tremors, although not all patients do.[1,3-5] Other symptoms include dentatorubral tremors related to the cranial nerve nuclei affected in the brainstem. The primary insults include primary hemorrhage, cavernous hemangioma, infarction, and trauma.[4] Most typically, they are hemorrhages following a hypertensive crisis.[2]

Four different patterns are recognized based on the location of the primary lesion: ipsilateral HOD with the primary lesion involving the brainstem, contralateral HOD with the primary lesion involving the cerebellum or the cerebellar peduncle, bilateral HOD with both central tegmental tracts affected by a midline lesion in region of brachium conjuctivum and bilateral HOD with the primary lesion in the unilateral brainstem and cerebellum.[6] Six phases of pathologic changes have been described in the following sequence: (a) No olivary change, (b) olivary amiculum degeneration, (c) olivary hypertrophy, (d) maximum olivary enlargement, (e) olivary pseudohypertrophy, and (f) olivary atrophy.[7]

Imaging findings relate to the changes in the inferior olivary nucleus and the causative primary lesion. Changes of HOD are not typically seen on CT and the findings seen on CT are related to the primary lesion. HOD is primarily diagnosed by MRI and the characteristic finding is a nonenhancing, T2-hyperintense enlargement of the inferior olivary nucleus.

The findings depend on the stage of evolution of the degenerative process.[5,8] On T2-weighted images, they characteristically go through three stages in the sequence of hyperintensity without hypertrophy, hyperintensity with hypertrophy followed by hyperintensity without hypertrophy again.[1,5] The major pathologic changes seen are vacuolar degeneration of the enlarged neurons, hypertrophy of the astrocytes, and gliosis. The T2-hyperintensity is attributed to the increased water content in these abnormalities.[2]

The first stage typically occurs within the first month, the second between 6 and 18 months and the final stage persists indefinitely. Similar changes are seen in T2-weighted Fluid Attenuation Inversion Recovery (FLAIR) images and proton-density weighted images. On T1-weighted images, olivary enlargement that is isointense or hypointense to gray matter is seen.[1,2,4,5]

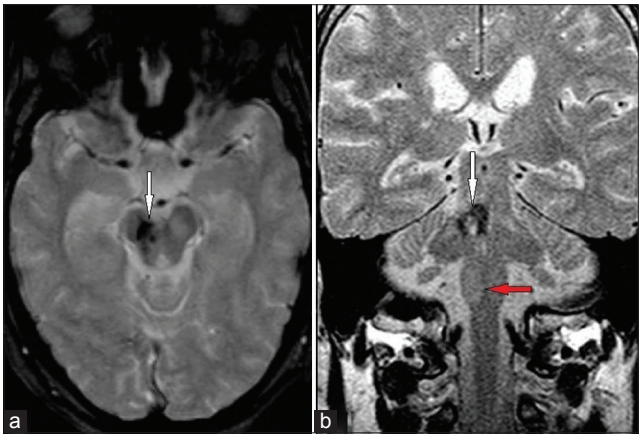

Figure 5: Case 2. 49-year-old male with unilateral hypertrophic olivary degeneration following brainstem hemorrhage. (a) Axial T2-weighted gradient image through midbrain shows susceptibility effect suggestive of chronic hemorrhage on right side involving the region of red nucleus (white arrow). (b) T2-weighted coronal images through medulla shows ill-defined hyperintensity in olivary region on right side (red arrow). Evidence of hemorrhage is noted as heterogeneous hypointensity in right side of pons (white arrow).

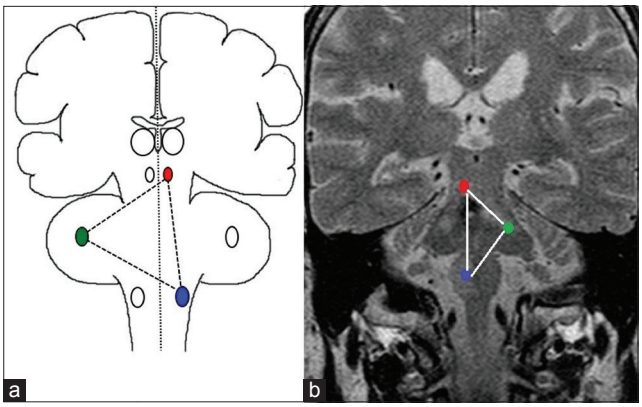

Figure 6: (a) Illustrative sketch of the Guillain–Mollaret triangle shows the inferior olivary nucleus (in blue), contralateral dentate nucleus of the cerebellum (in green) and the ipsilateral red nucleus (in red). (b) Schematic diagram of Guillain–Mollaret triangle, superimposed on T2W coronal image.

Diffusion tensor imaging and MR fiber tractography have also been shown to be capable of demonstrating disruption of the pathways in the Guillain–Mollaret triangle when changes on conventional MR imaging are equivocal.[6,9,10]

CONCLUSION

The significance of recognizing HOD lies in avoiding misdiagnosis of T2-hyperintensities in the anterolateral medulla in the appropriate clinical setting. The differential diagnosis for this includes tumors, infarction, infections, demyelinating lesions, and inflammatory processes among others.[2,4] However, the lack of contrast enhancement is significant as it differentiates HOD from inflammation and malignant tumors. The enlargement of the olives is not expected in diseases like multiple sclerosis and chronic stages of infarction.[2]

ACKNOWLEDGMENT

The authors would like to acknowledge the Department of Otorhinolaryngology of Mahatma Gandhi Medical College and Research Institute for providing the video clip.

REFERENCES

1. Sanverdi SE, Oguz KK, Haliloglu G. Hypertrophic olivary degeneration in children: Four new cases and a review of the literature with an emphasis on the MRI findings. Br J Radiol 2012;85:511-6.
2. Krings T, Foltys H, Meister IG, Reul J. Hypertrophic olivary degeneration following pontine haemorrhage: Hypertensive crisis or cavernous haemangioma bleeding? J Neurol Neurosurg Psychiatry 2003;74:797-9.
3. Kitajima M, Korogi Y, Shimomura O, Sakamoto Y, Hirai T, Miyayama H, et al. Hypertrophic olivary degeneration: MR imaging and pathologic findings. Radiology 1994;192:539-43.
4. Vaidhyanath R, Thomas A, Messios N. Bilateral hypertrophic olivary degeneration following surgical resection of a posterior fossa epidermoid cyst. Br J Radiol 2010;83:e211-5.
5. Goyal M, Versnick E, Tuite P, Cyr JS, Kucharczyk W, Montanera W, et al. Hypertrophic olivary degeneration: Metaanalysis of the temporal evolution of MR findings. AJNR Am J Neuroradiol 2000;21:1073-7.
6. Sen D, Gulati YS, Malik V, Mohimen A, Sibi E, Reddy DC. MRI and MR tractography in bilateral hypertrophic olivary degeneration. Indian J Radiol Imaging 2014;24:401-5.
7. Goto N, Kaneko M. Olivary enlargement: Chronological and morphometric analyses. Acta Neuropathol 1981;54:275-82.
8. Birbamer G, Buchberger W, Felber S, Aichner F. MR appearance of hypertrophic olivary degeneration: Temporal relationships. AJNR Am J Neuroradiol 1992;13:1501-3.
9. Shah R, Markert J, Bag AK, Curé JK. Diffusion tensor imaging in hypertrophic olivary degeneration. AJNR Am J Neuroradiol 2010;31:1729-31.
10. Dinçer A, Özyurt O, Kaya D, Koşak E, Öztürk C, Erzen C, et al. Diffusion tensor imaging of Guillain-Mollaret triangle in patients with hypertrophic olivary degeneration. J Neuroimaging 2011;21:145-51.

Source of Support: Nil, **Conflict of Interest:** None declared.