Bilateral hypertrophic olivary degeneration

Sameer Vyas, Anuj Prabhakar, Ajay Kumar, Niranjan Khandelwal

Department of Radiodiagnosis and Imaging, Postgraduate Institute of Medical Education and Research, Chandigarh, India

For correspondence:
Dr. Sameer Vyas, Department of Radiodiagnosis and Imaging, Post Graduate Institute of Medical Education and Research, Chandigarh, India. E-mail: sameer574@yahoo.co.in

Ann Indian Acad Neurol 2013;16:408-9

A 40-year-old male patient presented with complaints of headache of 9 months duration. Right facial paresis was detected on examination. Palatal myoclonus was not observed. Magnetic resonance imaging (MRI) brain showed multiple cavernous angioma involving bilateral tegmental tracts, bilateral red nuclei (more on the right side) and in the right thalamus [Figure 1]. Bilateral inferior olivary nuclei are enlarged and hyperintense on T2 Weighted imaging (WI)/fluid attenuation inversion recovery, hypointense on T1WI with effacement of pre- and post-olivary sulci and no enhancement on contrast administration consistent with Bilateral Hypertrophic Olivary degeneration [Figure 2].

Hypertrophic olivary degeneration (HOD) occurs in lesions involving the dentatorubro-olivary system.[1] It has characteristic imaging findings and temporal evolution. The triangle of Guillain and Mollaret is formed by the ipsilateral red nucleus, the inferior olivary nucleus (ION), and the contralateral dentate nucleus. The red nucleus and the inferior olivary nucleus are connected via the central tegmental tract. The superior cerebellar peduncle connects the ipsilateral red and the contralateral dentate nuclei and the inferior cerebellar peduncle links the contralateral dentate and the ipsilateral and inferior olivary nuclei.[2] Hypertrophic olivary degeneration is a degeneration of the inferior olivary nucleus that occurs secondary to a lesion in the dento-rubro-olivary pathway (DROP).[1] Histologically, there is vacuolar cytoplasmic degeneration with hypertrophy of the olivary nucleus due to an increase in the number of astrocytes.[3] HOD has been identified in all age groups and in both sexes. Clinically, the patient may present with palatal tremor, dentorubral tremor,[4] and features of cerebellar/brainstem dysfunction. Focal lesions that may lead to HOD include ischemia, infarct, demyelination, hemorrhage, trauma, and cavernous hemangioma.[5] When

Access this article online

Quick Response Code:

Website: www.annalsofian.org

DOI: 10.4103/0972-2327.116971

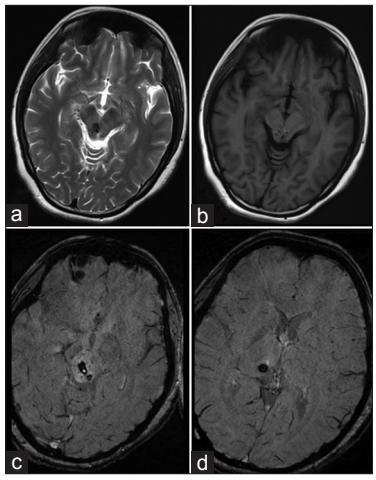

Figure 1: Axial T2 (a) and T1 (b) MR images showing predominantly hypointense and heterogeneous signal intensity lesions in bilateral tegmental tracts, bilateral red nuclei (more on the right side). Susceptibility weighted (c and d) MR images showing blooming in the right thalamus, bilateral tegmental tracts, bilateral red nuclei (more on the right side)

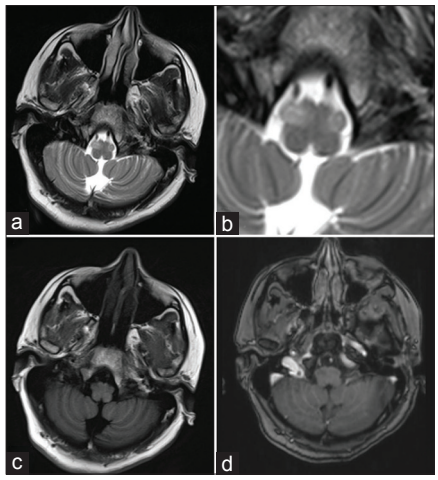

Figure 2: Axial T2 MR (a) and magnified Axial T2 MR (b) images show enlarged and hyperintense bilateral inferior olivary nuclei. Axial T1WMRI (c) shows hypointensity in bilateral inferior olivary nuclei with no enhancement on contrast enhanced MRI (d)

the central tegmental tract in the brainstem is involved in the primary lesion, ipsilateral HOD occurs. On the other hand, if the primary lesion involves the dentate nucleus or the superior cerebellar peduncle, contralateral HOD results. Bilateral HOD is rarer and occurs in midline lesions or lesions in the brachium conjunctivum (superior cerebellar peduncle), that also interrupt decussation of the DROP, can result in bilateral HOD. Three stages of hypertrophic olivary degeneration can be seen on T2 weighted MR WMR images. In the first stage occurring within the first 6 months, only hyperintensity of the inferior olivary nucleus is seen. The second phase lasts from 6 months up till 3 to 4 years, and is characterized by an increase in size and intensity with non-visualization of the pre- and post-olivary sulci. The third stage begins with the resolution of the hypertrophy with persistence of the hyperintense signal in the olivary nucleus, which may last indefinitely. On T1WMR images, iso- to hypo-intense enlargement of the inferior olivary nucleus is seen. There is no enhancement on contrast administration. The primary lesion can also be visualized on MRI. Atrophic changes in the cerebellum and reduction in the size of the contralateral ION may also be seen.

Our cases did not have the classical clinical picture of hypertrophic olivary degeneration. Even though the diagnosis of cavernous hemangioma was not confirmed by biopsy, it remains the most probable imaging cause of HOD in the normotensive patient. The other differential diagnoses for hypertrophic olivary degeneration include tumors, demyelinating lesions, and inflammatory/infective causes. However, the absence of contrast enhancement and the presence of another lesion in the dento-rubro-olivary pathway make other diagnosis unlikely.

References

1. Gatlin JL, Wineman R, Schlakman B, Buciuc R, Khan M. Hypertrophic olivary degeneration after resection of a pontine cavernous malformation: A case report. J Radiol Case Rep 2011;5:24-9.
2. Choh NA, Choh SA, Jehangir M. Hypertrophic olivary degeneration: The forgotten triangle of Guillain and Mollaret. Neurol India 2009;57:507-9.
3. 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.
4. Lim CC, Lim SA. Images in clinical medicine. Pendular nystagmus and palatomyoclonus from hypertrophic olivary degeneration. N Engl J Med 2009;360:e12.
5. 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.

How to cite this article: Vyas S, Prabhakar A, Kumar A, Khandelwal N. Bilateral hypertrophic olivary degeneration. Ann Indian Acad Neurol 2013;16:404-5.
Received: 22-11-12, **Revised:** **Accepted:** 16-12-12
Source of Support: Nil, **Conflict of Interest:** Nil