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Hypertrophic Olivary Degeneration: Review of Literature with emphasis on MRI finding and Differential Diagnosis

Niveditha Naik[1*], Sivasubramaniyan K.M[2], Viyango Pandiyan[3], Rushdina Mithran[4], Yash Bhutada[5]
[1, 4, 5] Post graduate, [2, 3] Assistant Professor
Department of Radiodiagnosis, Sri Lakshmi Narayan Institute of Medical Sciences, Puducherry, India

Corresponding Author:
Niveditha Naik
Post graduate, Department of Radiodiagnosis, Sri Lakshmi Narayan Institute of Medical Sciences, Puducherry, India

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ABSTRACT
Hypertrophic olivary degeneration [HOD] is a degenerative condition involving the Guillain-Mollaret triangle. This triangle is a functional neuronal pathway composed by connections between the red nucleus, the ipsilateral inferior olivary nucleus [ION], and the contralateral dentate nucleus of the cerebellum. This is a unique condition where there is degeneration presents with hypertrophy. This triangle contains dentato-rubro-olivary pathway. Any damage to this pathway leads to hypertrophy of the olivary nucleus. This case is a reminder about the MRI features of hypertrophic olivary degeneration and its differential diagnosis

Keywords: Hypertrophic olivary degeneration, Inferior olivary nucleus, Triangle of Guillain and Mollaret, Fluid-attenuated inversion recovery

INTRODUCTION

Hypertrophic olivary degeneration (HOD) occurs due to insult in the dentate-rubro-olivary pathway , which manifests as occulopalatal myoclonus [1]. The aetiology includes primary haemorrhage, cavernous haemangioma, vascular malformation, infarction, trauma, posterior fossa surgery, and idiopathic cause [2]. HOD is considered an orthodox as there is degeneration with enlargement, rather than atrophy as seen in other systems [3]. HOD is trans-synaptic degeneration where there are neuronal loss and reactive gliosis following an injury to the afferent fibres. We are thus intending to understand the MRI features of HOD and to differentiate from other similar pathologies.

CASE PRESENTATION

This 54-year-old male was diagnosed with cavernoma and was operated for this posterior fossa lesion, due to recurrent bleeding. After the surgery, he came for a follow-up study. Pre-operative images could not be obtained. In his post-operative MR examination performed two months following the surgery, no residual mass was observed, gliotic changes with dark signal areas noted in the right postero-lateral aspect of Pons and cerebral peduncle (at the operative site) suggesting degraded heamoglobin products with no contrast enhancement. T2-weighted and fluid-attenuated inversion recovery (FLAIR) sequences of MRI showed focal hyperintensity and expansion of both medullary olives with no enhancement seen following contrast administration, nor diffusion restriction on diffusion-weighted imaging (Figure No-1). These features were suggestive of bilateral HOD.

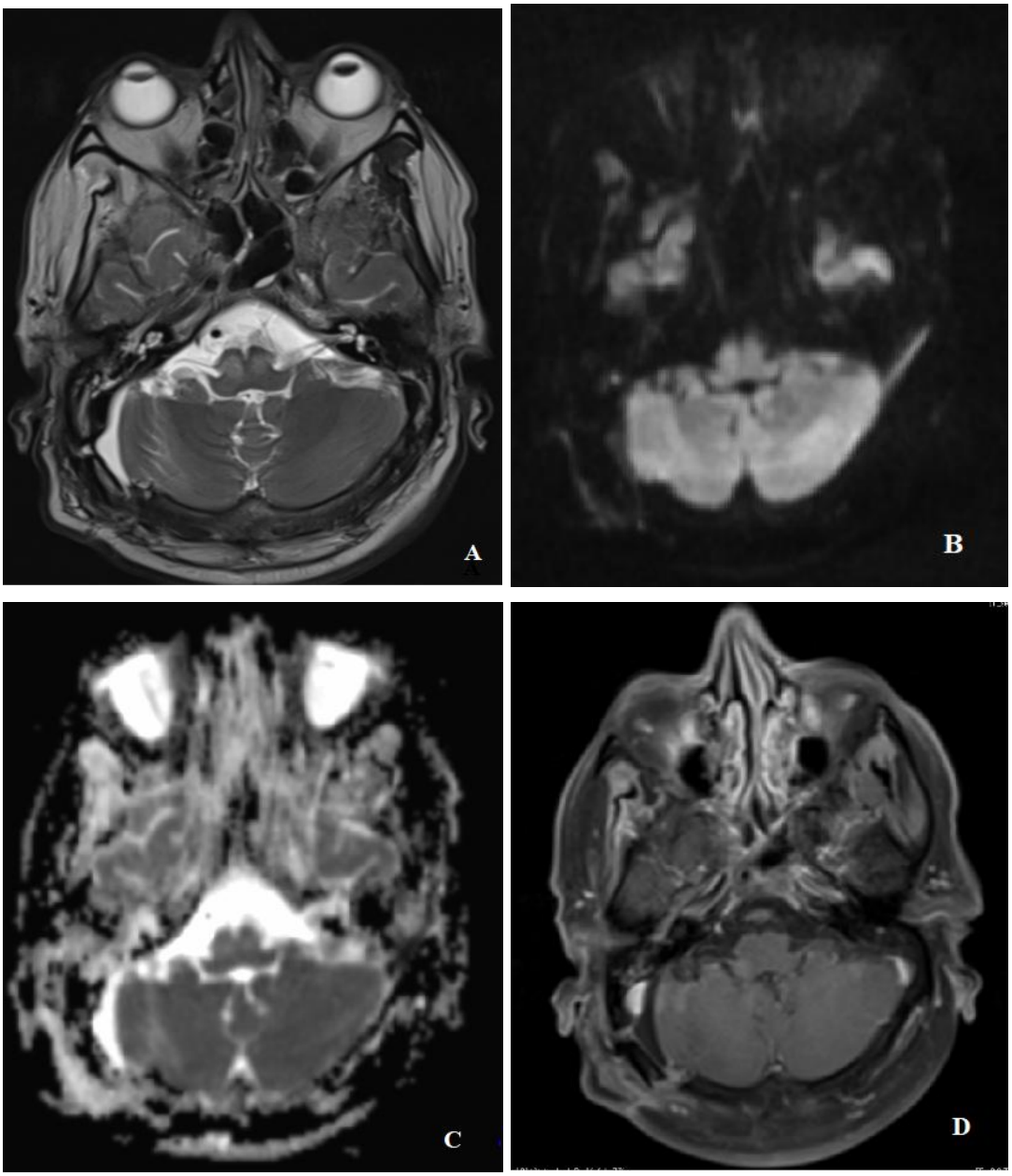

Figure No. 1: A 54 years old male status two months post resection of cavernoma in posterior fossa. MRI brain-Axial sections at the level of internal auditory meatus demonstrates [A] T2W unenhanced sections there is focal nodular hyper intensities in the both olivary nucleus, there is no evidence of restricted diffusion on DWI [B] and ADC [C] sequence. There is no evidence of contrast enhancement [D].

DISCUSSION

Anatomy:

The triangle of Guillain and Mollaret is the area between the ipsilateral red nucleus, inferior olivary nucleus and contralateral dentate nucleus [4]. The red nucleus [midbrain] receives afferent from the contralateral dentate nucleus [cerebellum] via dentatorubral pathway [by decussating at lower pons]. This pathway controls fine voluntary movement. The ION [inferior olivary nucleus] [medulla], which helps in co-ordination receives afferent fibres from the ipsilateral red nucleus via the central tegmental tract. The triangle is completed when the ipsilateral ION sends efferent fibres to contralateral dentate nucleus. In this process, these fibres decussate in cerebellum before reaching the dentate nucleus [4]. Since HOD is due to de-afferentation phenomenon, any injury to the efferent olivocerebellar tract is unlikely to cause palatal myoclonus [5]. Lesions that cause injury to central tegmental tract cause ipsilateral HOD, while the

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involvement of the dentate nucleus and its tract causes bilateral HOD. Our case is a posterior fossa tumour which involves the dentate nucleus as well. Hence there is bilateral HOD [5] (Figure No-2).

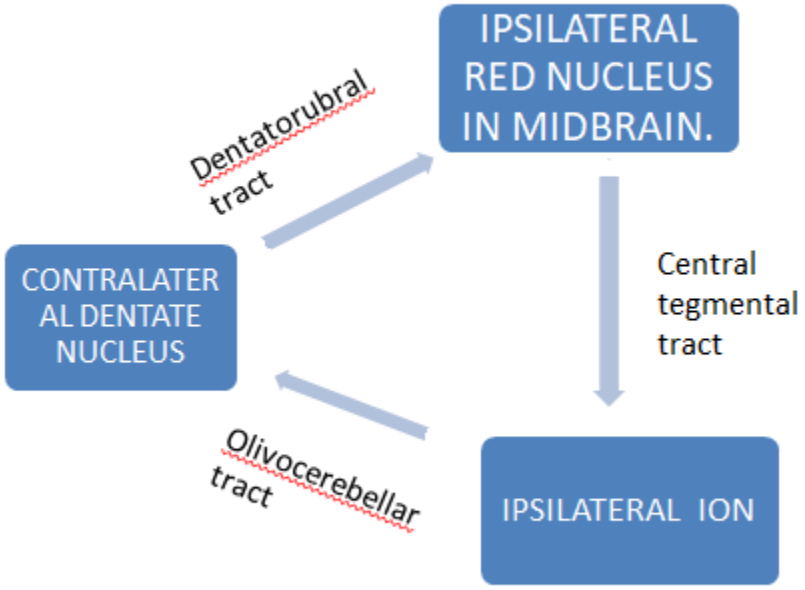

Figure No -2: Triangle of Guillain and Mollaret

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In the available literature, diagnosis of post-operative HOD have been reported rarely [6]. In most cases, degeneration occurs with atrophy, although there are neuronal loss and glial proliferation seen after tissue damage in the CNS yet the degeneration of the ION occurs with hypertrophy [7]. There are few postulates to justify this. Following an injury to neuronal connections in Guillain-Mollaret triangle, there is transsynaptic degeneration, this variety of degeneration is deafferentation syndrome [3,8] There is a decreased synaptophysin immunoreactivity confirming the presynaptic abnormalities linked to deafferentation [3,8]. The symptoms occur because the denervated olivary neurons released from inhibitory inputs enlarge and develop sustained synchronized oscillations. The signal on reaching cerebellum results in abnormal motor output [9]. Hence, clinically the patients present with movement disorder like palatal myoclonus (soft palate), Holmes' tremor and ocular myoclonus [10, 11]. Soft palate movements which can present within 2 to 40 months after the initial insult.

There are six stages pathologic changes in HOD documented by oto et al. which are: [a] no olivary change within 24 hours; [b] Degeneration of the white matter olivary capsule changes are observed by two to seven days; [c] Mild hypertrophy of olivary with no glial reaction may be seen after three weeks; [d] Olivary enlargement occurs with hypertrophy of both neurons and astrocytes are seen after one month; [e] Olivary pseudohypertrophy, due to neuronal dissolution may be seen approximately around nine months. [f] Olivary atrophy is evident after three to four years [12]. In our current study, the period from onset of the disease to MRI of the HOD is two months post-operatively and thus these findings would correspond to the fourth stage. If the hypertrophy progresses to atrophy, the symptoms, however, does not subside, probably due to loss of inhibitory feedback and disruption of rhythmicity [4].

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There are few differential diagnoses of HOD which has similar radiological findings such as infarction and neoplastic disease. HOD is strictly restricted to only one or both inferior olive, without invading surrounding structures. HOD can be differentiated from tumour, infection, and inflammation by the absence of contrast enhancement [4]. Acute infarction can be ruled out by the absence of diffusion restriction [4]. The pilocytic astrocytoma is well defined expansile/exophytic lesion which shows variable enhancement with contrast [4]. Corticospinal tract involvement is seen in Wallerian degeneration involves and hence can be ruled out [4].

The individuals affected with HOD are managed symptomatically. There are promising results with medications like clonazepam, valproic acid, and carbamazepine, they provide symptomatic relief [4]. As the palatal myoclonus is due to contraction of tensore veli palatine, in severe cases botulinum toxin is injected into the muscle [4].

CONCLUSION

Hypertrophic olivary degeneration is a rare condition characterized by T2W/FLAIR-hyperintensity with or without enlargement of the inferior olivary nucleus depending on the stage of the disease. The patients can be asymptomatic and such an incidental imaging finding should not be out looked and radiologists should be aware of its typical imaging features to avoid under diagnosing this condition and to prevent overt treatment.

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