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The Silent Triangle Speaks: Hypertrophic Olivary Degeneration Unveiled

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Abstract: ***Background*: Hypertrophic olivary degeneration is an uncommon form of trans synaptic degeneration affecting inferior olivary nucleus. It mostly develops secondary to a destructive lesion involving the Guillain–Mollaret pathway. It presents with symptomatic palatal tremor, less frequently can cause dentatorubral tremor (Holmes tremor). In some cases, these tremors may also involve facial muscles, diaphragm, or muscles of the tongue and larynx.HOD commonly develops secondary to a focal lesion like haemorrhage, infarction, tumours, infection, inflammatory conditions, trauma, or post-surgical. In rare cases, HOD has been documented to be idiopathic. *Case presentation*: We present a case of a 29-year-old male with complaints of diplopia, syncopal attacks. On physical examination, ocular tremors was seen. Patient had no significant past medical history. There was no history of trauma, neurosurgical interventions.***

Keywords: Guillain–Mollaret triangle; hypertrophic olivary degeneration; magnetic resonance imaging

1. Introduction

Hypertrophic olivary degeneration (HOD) is a rare trans-synaptic degeneration that occurs as a result of damage to the neuronal fibres of the dentato-rubral-olivary pathway (DRO)pathway. It is a circuit connecting the dentate nucleus in the cerebellum of one side to the red nucleus and the inferior olivary nucleus in the midbrain and medulla of the contralateral side respectively. In 1931, French physicians Georges Guillain and Pierre Mollaret first found out the trilateral connection between these three nuclei, and it is also known as the triangle of Guillain-Mollaret.

2. Imaging Findings

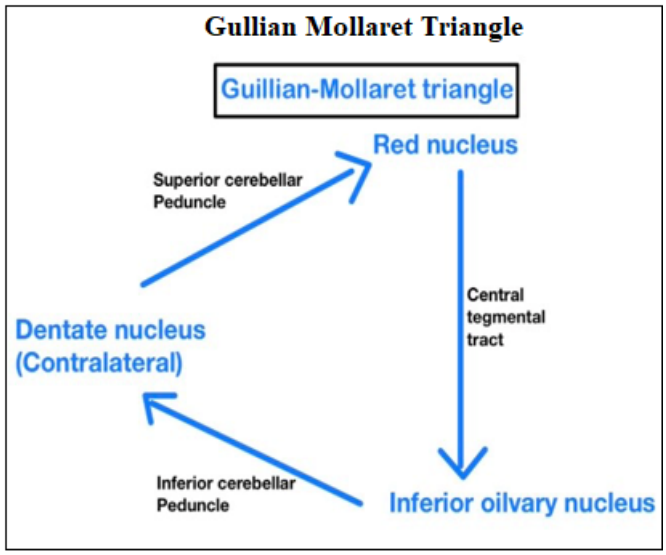
MRI Findings: MRI revealed FLAIR hyperintense non-enhancing lesion in the left medulla showing no diffusion restriction on DWI or blooming on SWI suggestive of Hypertrophic Olivary Degeneration. Linear area of blooming on T2* WI in right superior colliculus and ambient cistern.

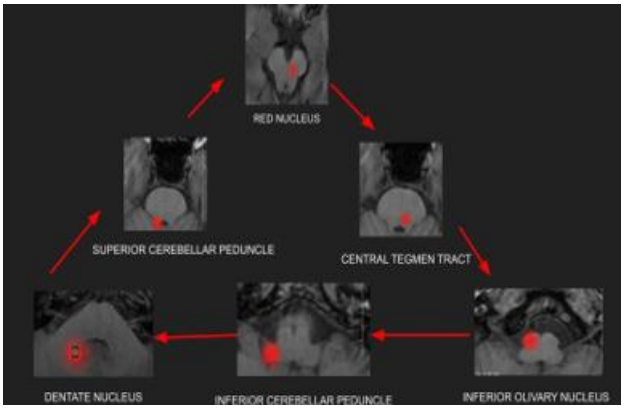
3. Discussion

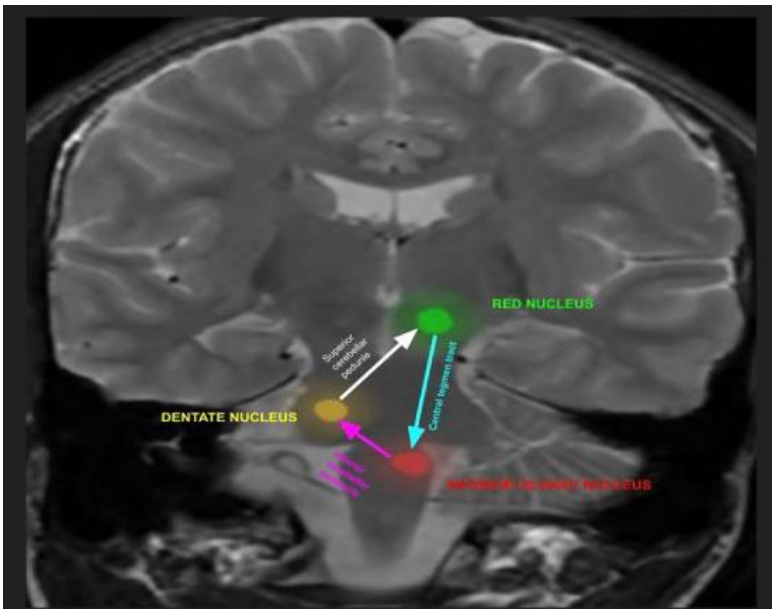
	Name and Duration	MRI Findings
Stage 1	Early acute (0-1 month)	No olivary changes
Stage 2	Acute stage (1-6 month)	Increased T2 FLAIR signal intensity of olivary nucleus
Stage 3	Intermediate (6 months-4 years)	Increased T2 FLAIR signal+ hypertrophy of olivary nucleus
Stage 4	Chronic (3-4years)	Increased signal intensity persists while nucleus size may normalise

Schematic representations of the Guillian-Mollaret triangle. The contralateral dentate nucleus sends efferent fibres to the red nucleus via the superior cerebellar peduncle (denatato-rubral pathway). The red nucleus in turn sends efferent fibres to the inferior olivary nucleus via the central tegmental tract (rubro-olivary pathway). Finally, the inferior olivary nucleus sends efferent fibres the contralateral cerebellar cortex via the inferior cerebellar peduncle (olivo-dentate pathway, which in turn connects to the contralateral dentate nucleus, hence completing the triangle. Three patterns of HOD can be produced depending on the location of the lesions:

MRI is the investigation of choice and typically demonstrates - increasedT2 and FLAIR signal intensity, which may later be accompanied by the enlargement of the inferior olivary nucleus. No diffusion restriction, blooming on T2* and post-contrast enhancement is seen. The evolution of HOD has been described in four stages based on MRI findings.





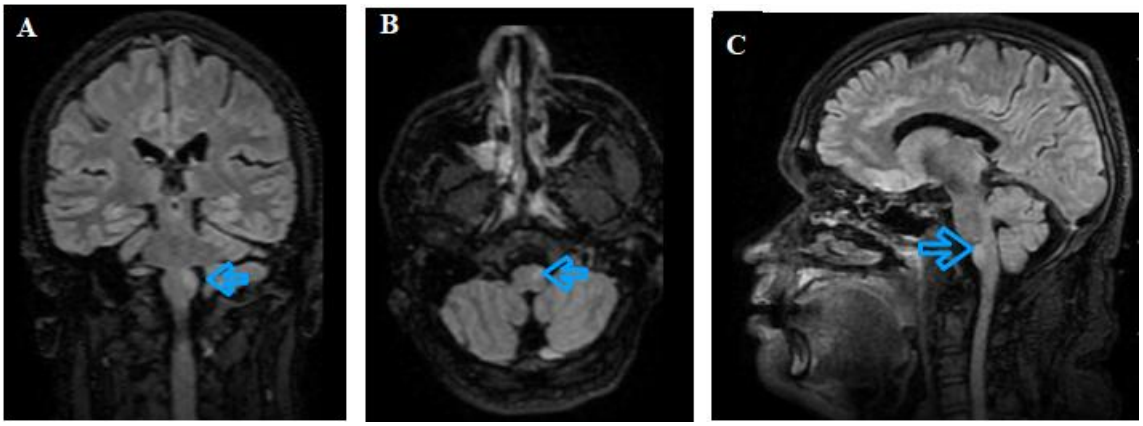


Contralateral HOD: lesions in the dentate nucleus or dentato rubral tract in the superior cerebellar peduncle.

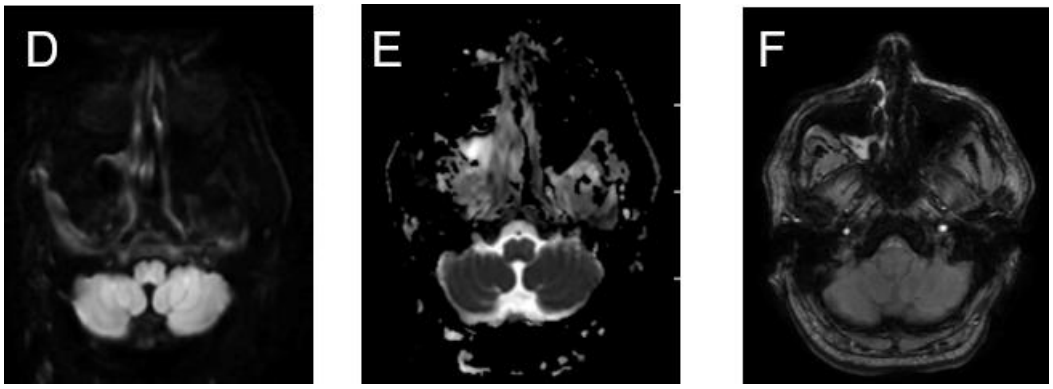
Bilateral HOD: lesions in the midline or Paramedian pons due to simultaneous involvement of superior cerebellar peduncle and central tegmental tract.

Ipsilateral HOD: lesion in brainstem tegmentum disrupts the rubro-olivary pathway in the central tegmental tract.

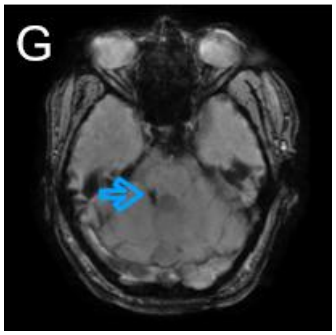
Image Gallery:



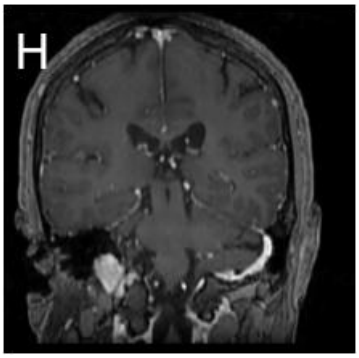
FLAIR hyperintense and swollen left inferior olivary nucleus in coronal (A) Axial (B) Sagittal (C) Sections



DWI (Fig-D) and ADC (Fig-E) -no diffusion restriction No evidence of blooming on SWAN (Fig-G)



SWAN(Fig-G)-linear area of blooming in right ambient cistern



T1 CONTRAST(Fig-H)-no enhancement

4. Conclusion

This case highlights importance of recognising characteristic MRI appearance of hypertrophic olivary degeneration. MRI changes can be noticed as early as 1 month after the initial insult, which can be either secondary to a variety of causes. MRI changes include hyperintensity on T2 and FLAIR with the increased size of the inferior olivary nucleus with no restricted diffusion, post-contrast enhancement or blooming. The distinction between left, right, and bilateral HOD serves as a clue to the location of the primary lesion. An accurate diagnosis is necessary since it mainly has a symptomatic management and does not require special interventions

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