22 Hypertrophic Olivary Degeneration

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INTRODUCTION

Hypertrophic olivary degeneration (HOD) is the result of an insult to the dentato-rubro-olivary pathway (Fig. 22.1), otherwise known as the Guillain-Mollaret triangle (GMT). After an insult (infarct, hemorrhage, trauma, tumor, surgery) that disrupts the GMT, hypertrophic degeneration of the affected inferior olivary nucleus (ION) develops. The counterintuitive degenerative *hypertrophy* (rather than atrophy) of the ION can lead to confusion. Of note, lesions involving this functional circuit may produce palatal myoclonus, which is one of the few involuntary movements that do not extinguish during sleep. Other classic clinical findings associated with HOD are dentato-rubral tremor and ocular myoclonus.

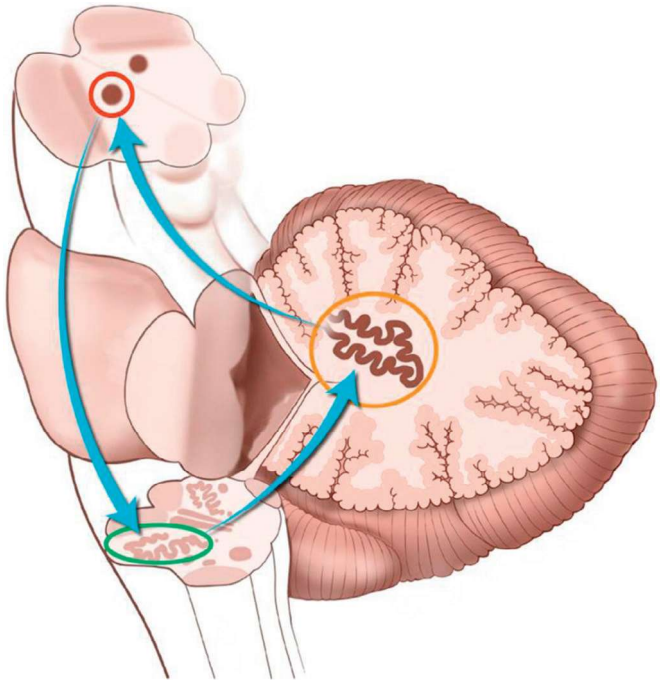

Figure 22.1. Guillain-Mollaret triangle (dentato-rubro-olivary pathway). This functional circuit is responsible for modulating spinal cord motor activity. It is composed of the ipsilateral red nucleus (RN) *(red circle)* of the midbrain, the ipsilateral inferior olivary nucleus (ION) *(green oval)* of the medulla, and the contralateral dentate nucleus (DN) *(orange circle)* of the cerebellum. The RN communicates with the ipsilateral ION via the central tegmental tract (CTT). The ION communicates with the contralateral DN via the inferior cerebellar peduncle. The DN communicates with the contralateral red nucleus via the superior cerebellar peduncle (SCP). The lesions that affect the afferent pathways to the olive result in hypertrophic olivary degeneration (HOD). Therefore lesions resulting in HOD of the *right* ION would involve the *right* RN, the *right* CTT, the *left* DN, or the *left* SCP. Lesions affecting efferent pathways to the olive (inferior cerebellar peduncle lesions) are less likely to cause HOD.

TEMPORAL EVOLUTION: OVERVIEW

The hallmarks of HOD are T2 hyperintensity and enlargement of the ION. The classic teaching for the imaging diagnosis of HOD depends on the identification of a nonenhancing mildly expansile T2 hyperintense olivary lesion in association with a lesion/insult to the contralateral dentate nucleus, contralateral superior cerebellar peduncle, ipsilateral red nucleus, or ipsilateral pontine tegmentum. However, an understanding of the temporal evolution of HOD is also necessary for the accurate interpretation of changing imaging patterns (Fig. 22.2). Three distinct phases are evident on magnetic resonance imaging (MRI): (1) ION T2 hyperintensity without hypertrophy within 6 months. (2) ION T2 hyperintensity with hypertrophy usually resolving by 3 to 4 years. (3) ION atrophy after several years with T2 hyperintensity persisting indefinitely (Fig. 22.3).

HOD is usually unilateral. Pathologic analysis of hypertrophic degeneration of the ION has demonstrated vacuolar degeneration, neuronal enlargement, astrocyte hypertrophy, and gliosis. HOD is ipsilateral to the causative lesion if it involves the brainstem or contralateral if the lesion involves the cerebellum. Rare cases of bilateral HOD have been reported with midline lesions at the level of the superior cerebellar peduncles, which disrupt the bilateral decussating dentate-olivary fibers (Fig. 22.4).

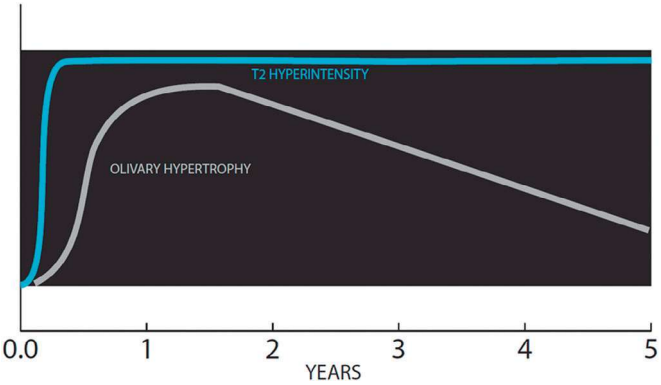

Figure 22.2. Temporal evolution of hypertrophic olivary degeneration (HOD). The evolution of HOD results in three distinct phases on magnetic resonance imaging depending on changing patterns of T2 hyperintensity *(blue line)* as well as olivary hypertrophy *(gray line)*. Histologic and imaging changes (T2 hyperintensity) begin approximately 3 weeks after initial injury in most patients but can first appear on imaging up to 6 months after the initial insult. Olivary hypertrophy follows T2 hyperintensity and initially develops approximately 6 months after the initial insult in most patients. It can persist for years but disappears in most patients by 3 to 4 years. Finally, atrophic changes have been noted after several years (not shown), although T2 hyperintensity persists indefinitely.

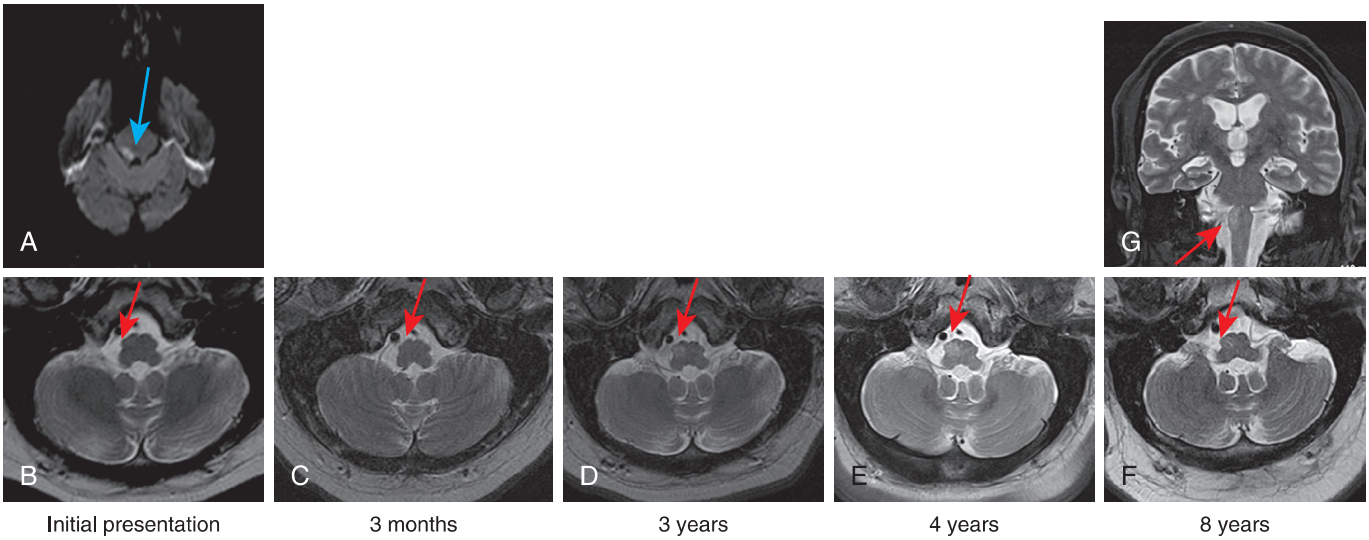

Figure 22.3. Temporal evolution of hypertrophic olivary degeneration (HOD) in a 62-year-old patient with brainstem stroke and subsequent oculopalatal myoclonus. At the time of initial presentation, an axial DWI image (A) through the pons demonstrates an acute infarct *(blue arrow)* involving the right superior cerebellar peduncle. Axial T2 images through the level of the medullary olive at the time of presentation (B) and 3 months later (C) demonstrate normal signal intensity and size of the right olive *(red arrows)*. Axial T2 images at 3 years (D) and 4 years (E) demonstrate interval development of right olivary T2 hyperintensity and hypertrophy consistent with HOD *(red arrows)*. At 8 years, axial (F) and coronal (G) T2 images demonstrate nearly complete resolution of hypertrophy with residual T2 hyperintensity *(red arrows)*.

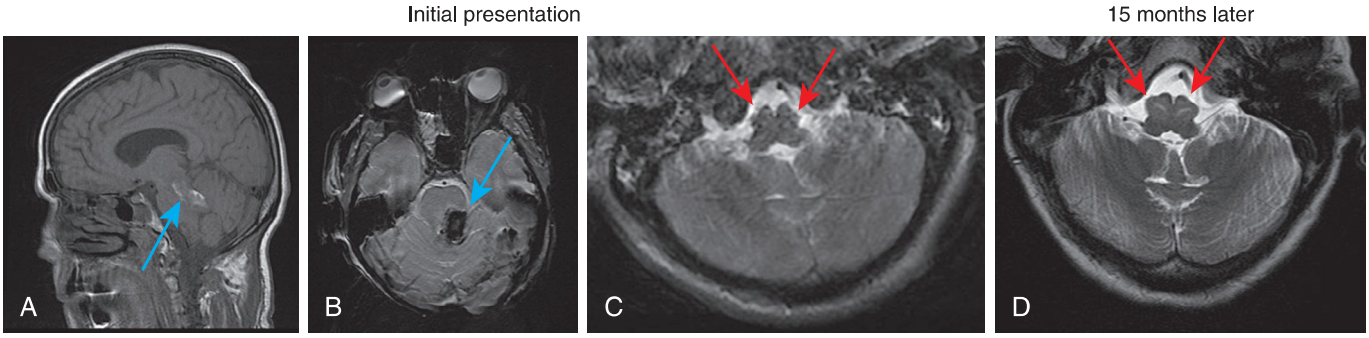

Figure 22.4. Bilateral hypertrophic olivary degeneration (HOD) in a 66-year-old woman with traumatic hemorrhage. Sagittal T1 (A) and axial gradient echo (B) images at the time of presentation demonstrate hemorrhage *(blue arrows)* involving the pons and the superior cerebellar peduncle, disrupting the decussating dento-olivary fibers. Axial T2 (C) image at the time of presentation demonstrates normal appearance of the olives *(red arrows)*. Axial T2 image (D) 15 months later demonstrates bilateral HOD.

MIMICS AND DIFFERENTIAL DIAGNOSIS

Differential diagnosis of olivary nucleus signal abnormality and enlargement includes infarct, neoplasm, demyelination, infection, and cavernous malformation (Fig. 22.5). As stated earlier, the key clue to the diagnosis of HOD is the presence of a causative lesion in the contralateral cerebellum or the ipsilateral brainstem in association with T2 hyperintensity with or without enlargement of the olive. As such, the presence of a lesion extending beyond the expected anatomic margins of the olive as well as the presence of enhancement or calcification should prompt the imaging interpreter to consider other diagnostic entities (Fig. 22.6).

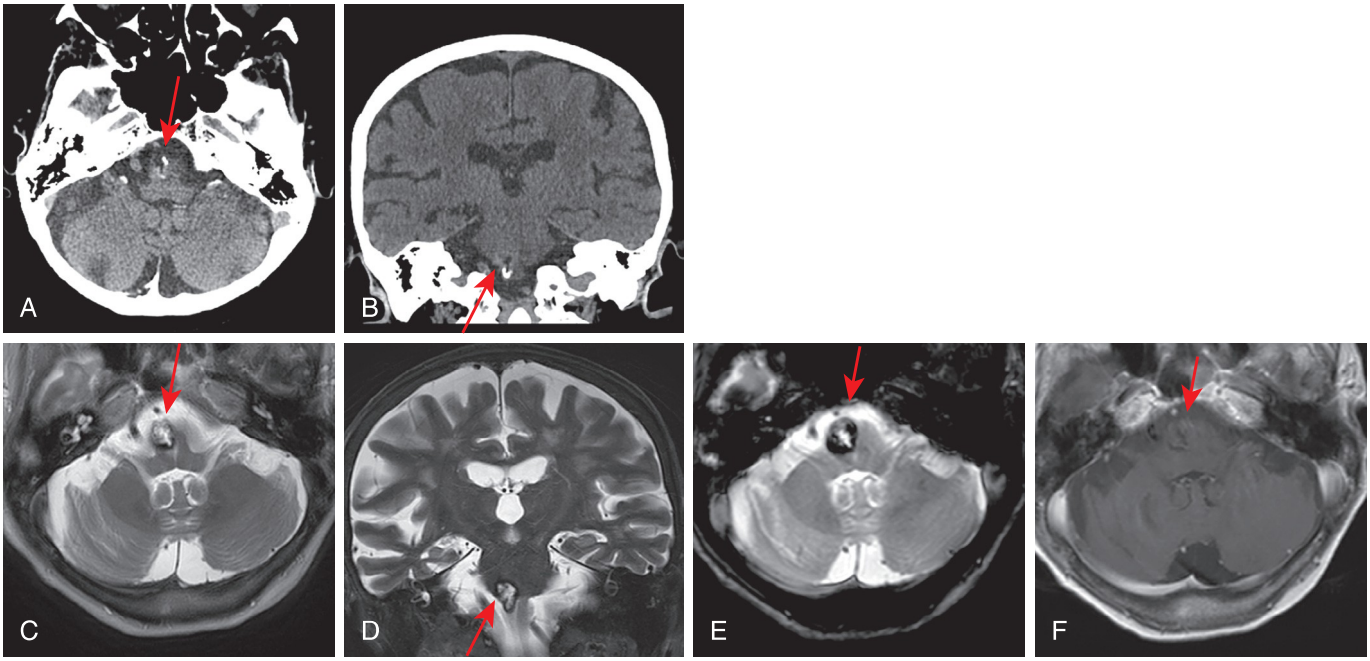

Figure 22.5. Cavernous malformation. An 84-year-old woman presented with altered mental status. Axial (A) and coronal (B) noncontrast head computed tomography images demonstrate an exophytic partially calcified lesion centered in the region of the right medullary olive *(red arrows)*. Axial T2 (C) coronal T2 (D), axial gradient echo (E), and axial T1 postcontrast (F) images confirm the presence of an exophytic right medullary cavernous malformation with a characteristic "popcorn" appearance, complete peripheral rim of T2 hypointensity, susceptibility blooming, and faint enhancement *(arrows)*.

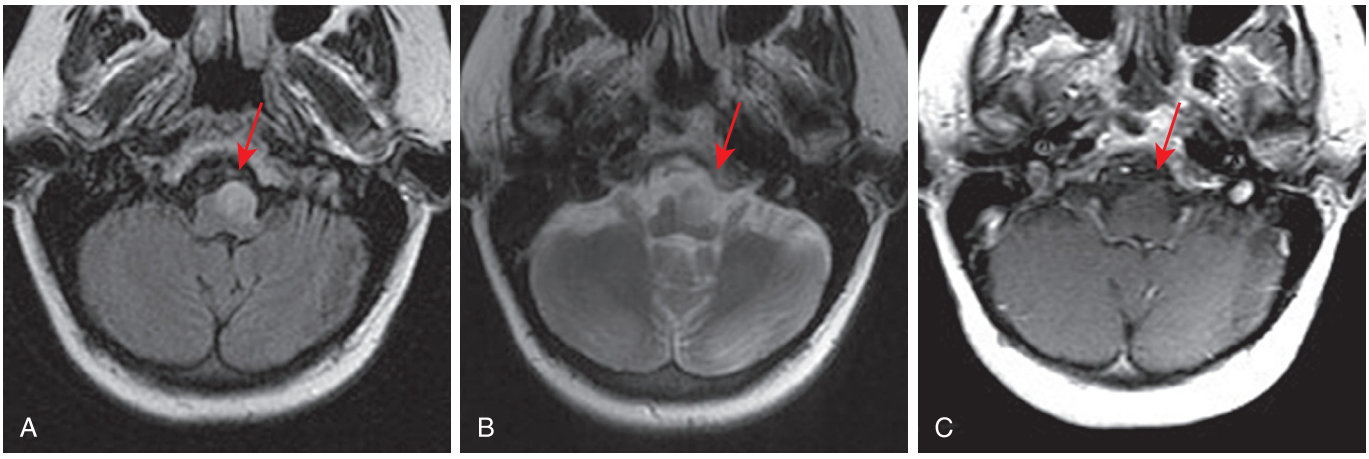

Figure 22.6. A middle-aged woman with known history of brainstem glioma. Axial fluid-attenuated inversion recovery (FLAIR) (A), axial T2 (B), and axial T1 postcontrast images (C) demonstrate a nonenhancing T2/FLAIR hyperintense mass *(arrows)* larger than expected in size for hypertrophic olivary degeneration and extending beyond the expected anatomic margins of the left medullary olive.

SUGGESTED READINGS

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