Hypertrophic Olivary Degeneration Following Clivus Meningioma Surgery

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CASE PRESENTATION

A 60-year-old man had a symptomatic clivus meningioma removed in May 2017. Postoperatively, he had skew diplopia, numbness on the left side of his face, drooling, dysphagia, and gait ataxia. He gradually improved over the next several months and reported being able to walk one mile in 14 minutes without the need for a supportive device. He had left eye muscle surgery to reduce his skew diplopia, without success.

Approximately 11 months after surgery, having been slowly improving, he developed progressive weakness in his legs, imbalance, dysarthria, a dull headache on the left side, increased drooling, and worsened gait. It then took him 45 minutes to cover just one mile. An MRI in May 2018 showed signs of hypertrophic olivary degeneration (HOD), characterized by T2-hyperintensity and mild enlargement of both inferior olivary nuclei (See **Figures 1A,B,C**), a new finding compared to the MRI performed four months earlier.

At his first examination in our clinic in June 2018, he had gait ataxia, mild bilateral proximal leg weakness, left facial numbness. The left eye had limited movement in all directions, presumed post-surgical, while the right eye had full movement. When looking laterally to either side however, the right eye would jerk down then laterally and then up.

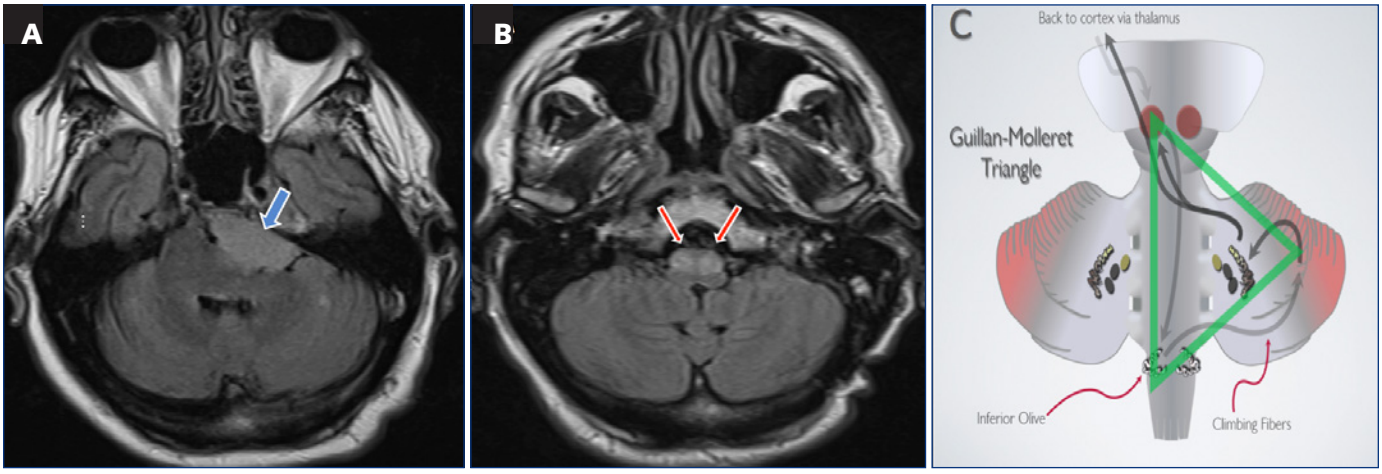
Over the following three years, ataxia, dysphagia, drooling, and dysarthria worsened, causing increased falls until 2021, when his symptoms stabilized, with no signs or symptoms of further deterioration. The MRI findings have remained unchanged however, as of May 2023.

DISCUSSION

Hypertrophic olivary degeneration (HOD) is a rare disorder, characterized by degenerative hypertrophy of the inferior olivary nucleus (ION) following a lesion of any type, anywhere in the Guillain-Mollaret triangle (GMT). The GMT consists of the dentate nucleus in the cerebellum, contralateral ION in the medulla oblongata, and the contralateral red nucleus in the midbrain. When the afferent fibers of the GMT are disrupted, HOD may occur.[1]

HOD presents clinically with various symptoms, including palatal tremor, ocular myoclonus, cerebellar signs, or Holmes' tremor,[2] which develop insidiously after a latent period that varies between 15 days and 18 months and not acutely after the insult.[3] The pathophysiology is hypothesized to be caused by the loss of inhibitory input to the ION, leading to hyperactivity of the olivary neurons,[1] but there is no evidence to support this.

Figure 1. Hypertrophic olivary degeneration (HOD) attributed to pontine compression by petroclival meningioma. [**A**] T2-FLAIR pre-surgery image at level of mid-pons shows extra-axial mass consistent with meningioma (blue arrow). [**B**] T2-FLAIR MR image 12 months after surgery at level of upper medulla shows hyperintense signal in both inferior olivary nuclei (red arrows) consistent with HOD. [**C**] Guillan-Molleret Triangle.



The pathology is unique and is postulated to be a variant of Wallerian degeneration characterized by neuronal and astrocytic swelling, resulting in enlargement, followed by degeneration and eventual atrophy of the ION.[4] Although early pathological changes are thought to develop within days of damage to the GMT, imaging and clinical changes develop months later.[5] HOD can also be an occasional imaging finding in some asymptomatic patients, for which the underlying mechanism remains unknown.[6] The condition can occur unilaterally or bilaterally, with bilateral cases being more common. Bilateral HOD often lacks an obvious cause in over half of the cases.[7]

The diagnosis of HOD rests on the characteristic MRI findings. The MRI changes occur in three stages: increased signal on T2-weighted MRI involving ION without hypertrophy (early stage); persistent T2-enhancement with associated hypertrophy (intermediate stage); then atrophy of the ION with persistent T2-enhancement (late stage).[1] Differential diagnosis includes infarction, inflammation, demyelination, or tumor.[2] There is no treatment for HOD, or most of the symptoms it causes.

References

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