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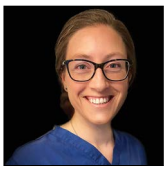
Case Report

Mollaret's triangle: An important neuroanatomical territory for all clinicians

Lindsey Bulleid[1], Tom Hughes[2], Paul Leach[1]

Departments of [1]Neurosurgery, [2]Neurology, University Hospital of Wales, Cardiff, South Wales, United Kingdom.

E-mail: *Lindsey Bulleid - lindsey.bulleid@doctors.org.uk; Tom Hughes - tom.hughes2@wales.nhs.uk; Paul Leach - paul.leach2@wales.nhs.uk



***Corresponding author:**
Lindsey Bulleid,
Department of Neurosurgery,
University Hospital of Wales,
Cardiff, South Wales, United Kingdom.

lindsey.bulleid@doctors.org.uk

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ABSTRACT

Background: Hypertrophic olivary degeneration is a rare condition caused by damage within the triangle of Guillain and Mollaret. We discuss the anatomical, radiological, and clinical history of this rare condition.

Case Description: A 32-year-old lady presented with sub-acute headache, photophobia, and dizziness. She also described facial tingling and itching over her nose, and a thirty-minute episode of slurred speech. Magnetic resonance imaging revealed a 12.1 × 11 × 7.3 mm lesion arising from the floor of the fourth ventricle [Figure 1]. Postoperative imaging confirmed complete resection of the tumor, but changes consistent with hypertrophic olivary degeneration [Figure 2a and b].

Conclusion: An awareness of this complication is of importance to all clinical neuroscience to prevent misdiagnosis with the occurrence of new symptoms.

Keywords: Cervical myoclonus, Fourth ventricular tumor, Hypertrophic olivary degeneration, Oscillopsia, Palatal myoclonus, Triangle of Guillain and Mollaret

INTRODUCTION

Hypertrophic olivary degeneration is a rare condition caused by damage within the triangle of Guillain and Mollaret. We describe a clinical case where this occurred and discuss the anatomical, radiological and clinical history of this rare condition.

CASE SUMMARY

A 32-year-old female presented with sub-acute headache, photophobia, and dizziness. Magnetic resonance imaging revealed a 12.1 × 11 × 7.3 mm lesion arising from the floor of the fourth ventricle [Figure 1]. The tumor – a choroid plexus papilloma – was completely resected through a posterior fossa craniotomy. Initially, she was asymptomatic but then developed occasional palatal myoclonus and oscillopsia. Postoperative imaging demonstrated unilateral hypertrophic olivary degeneration [Figure 2a and b].

DISCUSSION

Hypertrophic olivary degeneration is a rare condition caused by damage within the triangle of Guillain and Mollaret.[4,5] The olivary nucleus receives efferents from the ipsilateral red

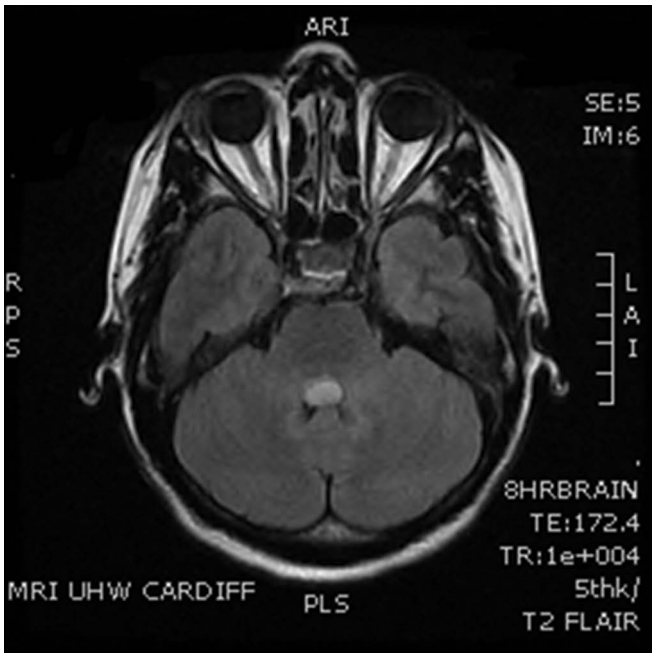

Figure 1: Axial T2 FLAIR magnetic resonance image showing fourth ventricular lesion

nucleus, which, in turn, receives efferents from the contralateral dentate nucleus through the superior cerebellar peduncle.[1,3] The decussating fibers from the inferior olivary nucleus terminate at the original dentate nucleus [Figure 3a and b]. The inferior olive gives rise to all the "climbing fibers" innervating the purkinje cells. Damage to the connections between the red nucleus and the contralateral dentate nucleus or between the red nucleus and the ipsilateral inferior olivary nucleus cause trans synaptic degeneration and interruption of the afferent input to the olivary nucleus.[1,2,5] The resulting loss of inhibition within this bidirectional and coupled "feedback loop" leads to hypertrophy of the nucleus before atrophy.[1] Damage to the olivodentate tracts does not produce the same hypertrophy of the olivary nucleus, rather this results in cerebellar atrophy.[2,3]

Olivary hypertrophy develops within 6 months of the initial event and resolves within 3–4 years.[2,3] It is most commonly a unilateral phenomenon ipsilateral to the side of the original insult when this is within the brainstem, contralateral if the original insult is cerebellar. Rarely, bilateral hypertrophic Olivary degeneration can occur where decussating fibers from both olivary nuclei are involved.[3]

The most common clinical presentation is of palatal or oculopalatal myoclonus, weeks to months after the initial insult.[1-5] For those patients who have had surgery, the occurrence of new symptoms may raise concerns about a recurrence of the original pathology. For those patients who have had brain stem strokes, the occurrence of new symptoms may lead to rapid repeat assessment with a view

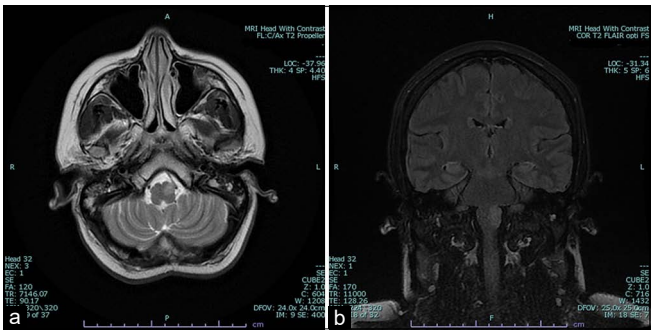

Figure 2: (a) Axial T2-weighted magnetic resonance image showing signal increase in the anterior medulla oblongata (more pronounced on the left) consistent with hypertrophic olivary degeneration. (b) Coronal T2 FLAIR magnetic resonance image showing hyperintensity over the region of the inferior olivary nucleus.

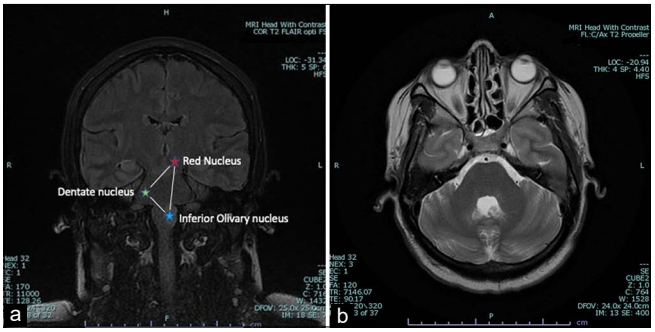

Figure 3: (a) Coronal T2 FLAIR magnetic resonance image showing schematic of triangle of Guillain-Mollaret. (b) Axial T2 magnetic resonance image at the level of the dentate nucleus and inferior/middle cerebellar peduncles.

to thrombolysis or thrombectomy. In both instances, detailed brainstem imaging is required to confirm the presence of the hypertrophic olivary nucleus and the pathological process involved. Presentation of this condition differs from Multi system atrophy diseases and olivopontocerebellar atrophy, where a more global ataxia is characteristically part of the presenting signs and symptoms. An awareness of this complication of past insults to the brain stem is of importance to all clinical neuroscience to prevent misdiagnosis with the occurrence of new symptoms.

CONCLUSION

This case provides an opportunity to review clinical anatomy, radiological change and neurology, and is an interesting illustration of a rare complication of fourth ventricular surgery. It demonstrates the intricacy of brainstem function and the variable relationship between radiological hypertrophy of the olivary nucleus and clinical symptoms, providing an important reminder of key functional anatomy and pathology.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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