Temporal evolution of Hypertrophic Olivary Degeneration in a pediatric patient: A case report and review of literature

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Background

Hypertrophic Olivary Degeneration (HOD) is a rare type of delayed degeneration associated to interruption of the dentato-rubro-olivary tract or triangle of Guillain-Mollaret(1). This neuronal circuit is also known as the myoclonic triangle. The synaptic circuitry is between the red nucleus, the inferior olivary nucleus (ION) and the contralateral cerebellar dentate nucleus. Damage to this tract has been classically associated to ataxia and palatal myoclonus(2). Current literature presents only few cases in pediatric patients(4). Even fewer descriptions exist in the neurosurgical literature.

Case Description

15 y/o male without prior medical history who presented with progressive headaches and discoordination. Upon evaluation he was found with left-sided dysmetria, fast-phase nystagmus and ataxia. MRI revealed a left cerebellar peduncle contrast-enhancing lesion with an associated cyst. Surgical resection ensued with a sub-occipital craniotomy with teloveral approach to the 4th ventricle. The patient awoke with mild left-sided abducens nerve palsy as well as exacerbated dysmetria and ataxia, which resolved within 72 hours. Follow-up images were obtained at 4 and 11 months post-op. As can be seen from pre-op imaging, prior to tumor resection, no ION hypertrophy was observed. On subsequent imaging 4 months post-op, evident T2 FLAIR hyperintensity and mild hypertrophy can be observed, worse on the 11 months post-op scan. Evident residual tumor with mild interval growth is also noted. However, at the time of this presentation, the patient was asymptomatic.

Pre-op MRI

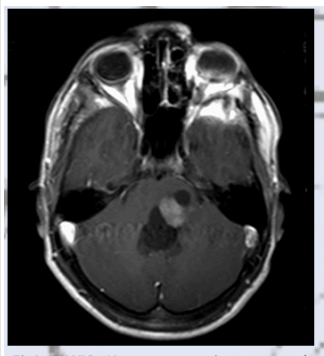

Fig1: T1WIC+ Upon presentation at tumor level

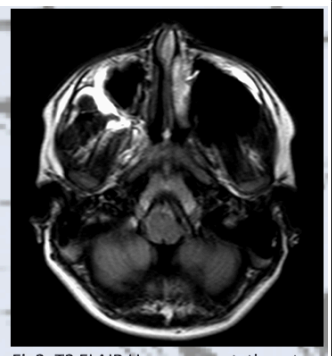

Fig2: T2 FLAIR Upon presentation at ION level

4 months post-op MRI

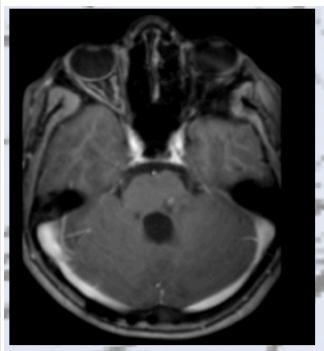

Fig3: T1WIC+ 4 months after resection at tumor level

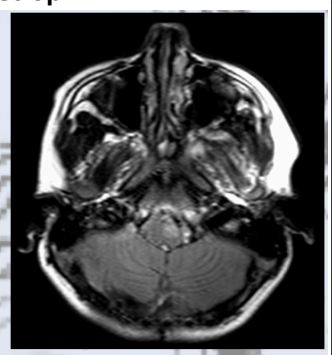

Fig4: T2 FLAIR 4 months after resection at ION level

11 months post-op MRI

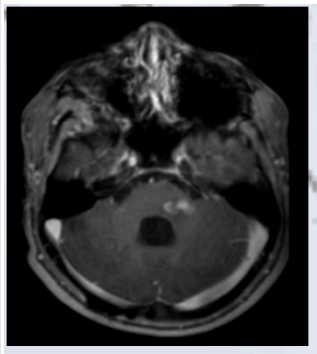

Fig5: T1WIC+ 11 months after resection at tumor level

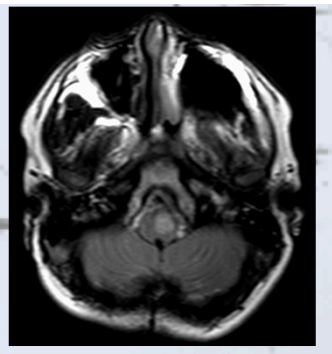

Fig6: T2 FLAIR 11 months after resection at ION level

Conclusions

HOD remains an entity not well discussed in neurosurgical literature. As this case attempts to demonstrate, it's presentation may be early and not accompanied by significant neurologic findings. Particularly in neoplastic cases it may represent a diagnostic challenge and could be easily confused for tumor recurrence. A multidisciplinary approach for this entity as with other pathologies is of particular importance. It's proper recognition will result in the best outcomes for the patient.

Disruption of the triangle of Guillain-Mollaret in our patient

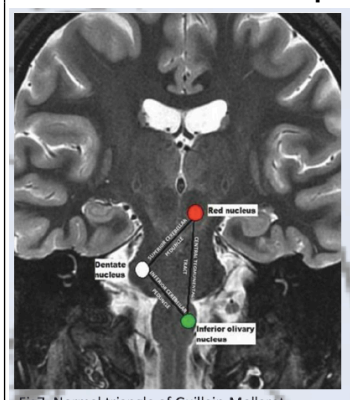

Fig7: Normal triangle of Guillain-Mollaret

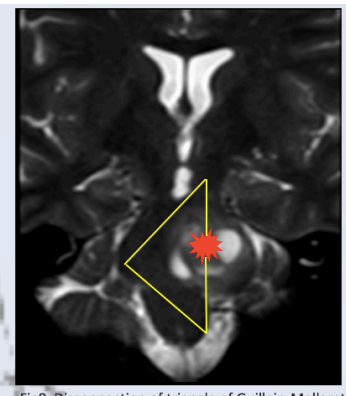

Fig8: Disconnection of triangle of Guillain-Mollaret in our patient

References

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