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Case Report

Hypertrophic olivary degeneration secondary to pontine haemorrhage

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ABSTRACT

We report a 58-year-old man who developed hypertrophic olivary degeneration (HOD) after haemorrhage of a cavernous malformation in the pons. Lesions of the triangle of Guillain and Mollaret (the dentatorubro-olivary pathway) may lead to HOD, a secondary transsynaptic degeneration of the inferior olivary nucleus. HOD is considered unique because the degenerating olive initially becomes hypertrophic rather than atrophic. The primary lesion causing pathway interruption is often haemorrhage, either due to hypertension, trauma, surgery or, as in our patient, a vascular malformation such as a cavernoma. Ischaemia and demyelination can also occasionally be the inciting events. The classic clinical presentation of HOD is palatal myoclonus, although not all patients with HOD develop this symptom. The imaging features of HOD evolve through characteristic phases. The clue to the diagnosis of HOD is recognition of the distinct imaging stages and identification of a remote primary lesion in the triangle of Guillain and Mollaret. Familiarity with the classic imaging findings of this rare phenomenon is necessary in order to avoid misdiagnosis and prevent unnecessary intervention.

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1. Introduction

Lesions of the triangle of Guillain and Mollaret (the dentatorubro-olivary pathway) may lead to hypertrophic olivary degeneration (HOD), a secondary transsynaptic degeneration of the inferior olivary nucleus (ION). HOD is considered unique because the degenerating olive initially becomes hypertrophic rather than atrophic. The primary lesion causing pathway interruption is often haemorrhage, either due to hypertension, trauma, surgery or a vascular malformation such as a cavernoma. Ischaemia and demyelination can also occasionally be the inciting events.

2. Case report

A 58-year-old man presented to the emergency department with acute onset of weakness of the right facial muscles and left upper and lower extremities. His past medical history included hypertension, intravenous drug use and alcoholic/hepatitis C cirrhosis (Child–Pugh score A). A non-contrast CT scan performed on admission demonstrated an acute 11 mm right-sided pontine haemorrhage (Fig. 1a). No abnormality was detected on CT angiogram. A routine follow up brain MRI performed 4 months later

revealed a region of heterogeneous T1- and T2-weighted signal in the dorsal right pons with prominent blooming on the susceptibility-weighted sequence but without significant contrast enhancement, most consistent with a cavernoma (Fig. 1b). At that time, the right ION was noted to be expanded and demonstrated high T2 signal characteristic of HOD (Fig. 1c, d).

3. Discussion

HOD is a secondary transsynaptic degeneration of the ION caused by a lesion in the triangle of Guillain and Mollaret (the dentatorubro-olivary pathway) (Fig. 2). The triangle, initially described by Guillain and Mollaret in 1931 [1], consists of the ION, the contralateral dentate nucleus (DN) and the ipsilateral red nucleus (RN). Fibres connecting the DN and RN ascend through the superior cerebellar peduncle (dentatorubral tract) and the central tegmental tract connects the RN to the ION. The triangle is completed by fibres that cross from the ION to the contralateral DN via the inferior cerebellar peduncle (olivodentate fibres).

HOD is caused by lesions in the dentatorubral or central tegmental tracts, as disruption of these pathways leads to functional deafferentation of the ION. Isolated lesions of the inferior cerebellar peduncle do not result in HOD as anatomically there are no direct connections between the ION and the contralateral DN. Rather, inferior fibres in the inferior cerebellar peduncle from

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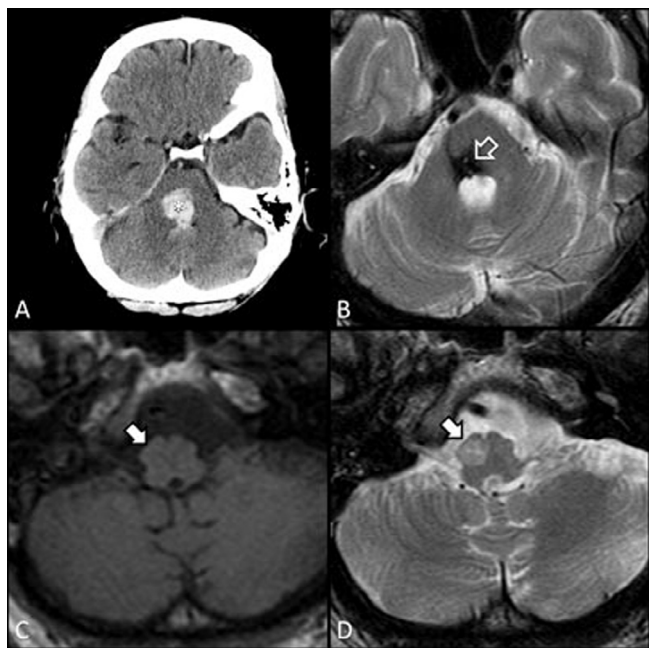


Fig. 1. Non-contrast axial CT scan performed on admission demonstrates an acute right-sided pontine haemorrhage (A). T2-weighted axial MRI performed 4 months later demonstrates a region of low signal intensity within the dorsal right pons at the site of haemorrhage (arrow), most consistent with a cavernoma (B). T1-weighted axial MRI sequence through the medulla demonstrates expansion of the right inferior olivary nucleus (C; arrow). T2-weighted axial MRI sequence of this region demonstrates increased signal intensity of the right inferior olivary nucleus (D; arrow), characteristic of hypertrophic olivary degeneration.

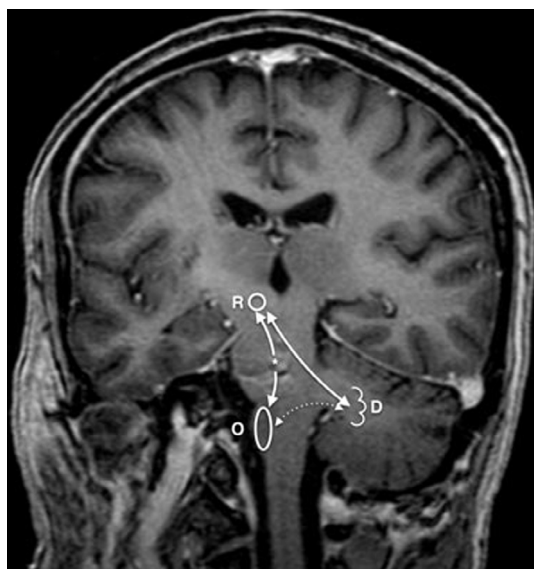


Fig. 2. Coronal T1-weighted sequence demonstrates the Guillain–Mollaret triangle which consists of the inferior olivary nucleus (O), the ipsilateral red nucleus (R) and the contralateral dentate nucleus (D). The dentate nucleus and red nucleus are connected by the dentatorubral tract, the red nucleus and inferior olivary nucleus are connected by the central tegmental tract, and the inferior olivary nucleus and dentate nucleus are connected by the olivodentate fibres.

the ION project first to the contralateral cerebellar cortex (olivocerebellar tracts) and then to the DN [2]. When these fibres are disrupted cerebellar atrophy can occur [3].

The primary lesion causing pathway interruption is often haemorrhage, either due to hypertension, trauma, surgery or, as in our patient, a vascular malformation such as a cavernoma. Ischaemia and demyelination can also occasionally be the inciting events. Lesions of the superior cerebellar peduncle cause contralateral HOD whereas primary lesions of the central tegmental tract cause ipsilateral HOD. Bilateral HOD can occur if the primary lesion involves both of the aforementioned structures.

The classic clinical presentation of HOD is palatal myoclonus which is characterised by rhythmic involuntary movements of the oropharynx due to contractions of the levator veli palatini muscle and can sometimes also involve the larynx, tongue and face. Palatal myoclonus usually develops 10 to 11 months after the primary lesion, although not all patients with HOD develop this symptom [4]. Occasionally, patients with HOD may develop a dentatorubral tremor (Holmes tremor) of the upper limbs [5].

HOD is considered unique because the degenerating olive initially becomes hypertrophic rather than atrophic. Pathologically, olivary enlargement corresponds to vacuolar degeneration of the cytoplasm, glial hypertrophy and proliferation of gemistocytic astrocytes [6]. Over time, the olive undergoes atrophy.

The imaging features of HOD evolve through characteristic phases [3,7]. In the acute stage, the olive appears normal. After 1 month, the ION develops hyperintense T2-weighted signal. Between 6 months and 3 to 4 years, the olive demonstrates hypertrophy and hyperintense T2-weighted signal. The hypertrophy then resolves but the hyperintense T2-weighted signal persists indefinitely. HOD typically does not enhance on contrast-enhanced sequences, although cases of enhancement have been reported [8].

The diagnosis for HOD includes a wide variety of pathological lesions that cause T2-weighted hyperintensity in the anterior medulla. These include infarction, demyelination, tumour (astrocytoma, metastasis, lymphoma) and infectious/inflammatory processes such as tuberculosis, sarcoidosis and rhombencephalitis. The clue to the diagnosis of HOD is the presence of a remote lesion in the contralateral cerebellar DN or superior cerebellar peduncle, or in the ipsilateral RN or pontine tegmentum. Familiarity with the characteristic imaging findings of this rare phenomenon is necessary in order to avoid misdiagnosis and prevent unnecessary intervention.

Conflicts of Interest/Disclosures

The authors declare that they have no financial or other conflicts of interest in relation to this research and its publication.

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