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LETTER TO THE EDITOR

Combined hypertrophic olivary degeneration and Wallerian degeneration of the bilateral middle cerebellar peduncles: a case report

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Dear Editor,

Hypertrophic olivary degeneration (HOD), a rare condition, is characterized by a unique pattern of trans-synaptic degeneration caused by injury to the brainstem or cerebellum within the triangle of Guillain and Mollaret [1]. Bilateral Wallerian degeneration (WD) of the middle cerebellar peduncles is caused by interruption of crossed cortico-ponto-cerebellar pathways and is rarely reported in the literature [2]. The three most common types of main transaxonal degeneration involving cerebellar connections are hypertrophic olivary degeneration, Wallerian degeneration of the middle cerebellar peduncle, and crossed cerebellar diaschisis (CCD). The occurrence of any one of these phenomena is very rare, and they are therefore usually reported separately in the literature. Here, we report a patient who presented with the neuroradiological findings of WD involving the bilateral middle cerebellar peduncles secondary to brainstem hemorrhage, as well as abnormal signaling in the inferior olivary nucleus. HOD combined with bilateral WD of the middle cerebellar peduncles is extremely rare, and to our knowledge, this is the first report on its occurrence.

Case presentation

A 54-year-old man with a known history of long-standing hypertension experienced a brainstem hemorrhage 4 months prior to his presentation. On arrival, he had slurred speech, dysphagia, and right-sided weakness. He complained of daily episodic rotatory dizziness and unsteady gait that had begun 20 days prior. Hence, he was admitted to our neurological department. On admission, his blood pressure was 155/94 mmHg and his heat rate was 95 beats/min and regular. Neurological examination revealed dysarthria, right-sided facial weakness, and decreased muscle strength in the right limbs (Medical Research Council Grade 4). Finger-to-nose and heel-to-shin tests detected marked bilateral motor incoordination and dysmetria. Babinski sign was positive on the right side. He displayed no involuntary movements, such as limb tremor or palatal myoclonus. Overall, he scored a 6 on the National Institutes of Health Stroke Scale (NIHSS). Magnetic resonance imaging (MRI) of the brain revealed hypointense lesions in the pontine tegmentum and left midbrain on both T2-weighted and FLAIR images, which was in

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line with hemosiderosis from the prior hemorrhage (Fig. 1). Diffusion-weighted image (DWI) showed no restricted diffusion. In addition, MRI showed hyperintensity in the bilateral middle cerebellar peduncles and left oblongata medulla on both T2-weighted and FLAIR images and increased volume of the left inferior olivary nucleus (Fig. 1). These findings taken together confirmed a diagnosis of Wallerian degeneration and hypertrophic olivary degeneration.

Discussion

Hypertrophic olivary degeneration (HOD) is a rare neurological condition of trans-synaptic degeneration that occurs secondary to focal lesions in the anatomical functional loop of the Guillain-Mollaret triangle. Possible causes include hemorrhage, infarction, tumor, neurosurgery, metronidazole toxin, progressive multifocal leukoencephalopathy, and Dandy-Walker variants [1]. In this case, the patient developed HOD from his brainstem hemorrhage. HOD is considered to be an unusual degeneration in which hypertrophy is seen instead of atrophy. Pathologically, hypertrophy and vacuolar degeneration of neurons, hypertrophy of astrocytes, and gliosis are described [3]. Enlargement and increased signaling on T2-weighted images of the inferior olivary nucleus (ION) complex is the radiological hallmark of HOD. Patients classically present with rhythmic palatal tremor (myoclonus). Ocular myoclonus and Holmes' tremor are also known to occur. Kalla et al. report on a patient who presented with a rare oculopalatal tremor known as "dancing jaw and dancing eyes" [4]. However, review of the literature reveals that not all HOD patients will develop a palatal tremor [5], and none was observed in our patient.

The Guillain-Mollaret triangle is a feedback circuit involved in voluntary motor control. It is composed of the ipsilateral inferior olivary nucleus, ipsilateral red nucleus (RN), and the contralateral dentate nucleus (DN) (Fig. 2). The superior cerebellar peduncle (SCP), central tegmental tract (CTT), and the inferior cerebellar peduncle (ICP) are white matter tracts that form the arms of the triangle. Lesions of the superior cerebellar peduncle result in

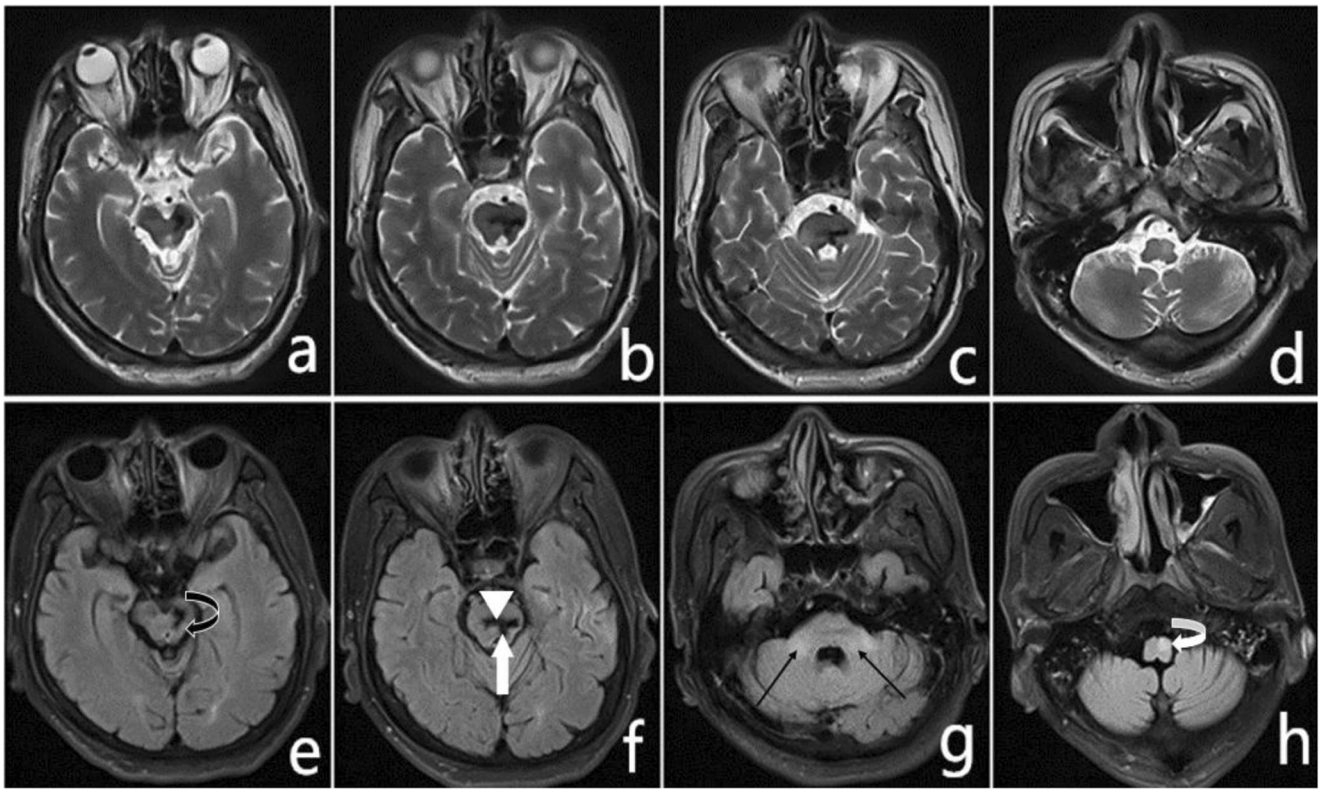

Fig. 1 Axial T2 image (**a**–**c**) and FLAIR (**e**, **f**): Residual lesion in the midbrain (left red nucleus and substantia nigra) and pontine tegmentum, characterized by prominent hypointensity. The lesion corresponds to old hemorrhage, interrupting the red nucleus (black and curved arrow) and afferent fibers of the central tegmentar tract (white arrow) located in the pons to the inferior olivary nucleus. In addition, the old lesion in the pons involving the crossed cortico-pontinecerebellar tracts (arrowhead). Axial T2 image (**d**) and FLAIR (**g**, **h**): Hyperintensity on the left inferior olivary nucleus (white and curved arrow) and both middle cerebellar peduncles (black and thin arrow). In addition, increased volume of the left inferior olivary nucleus can also be seen

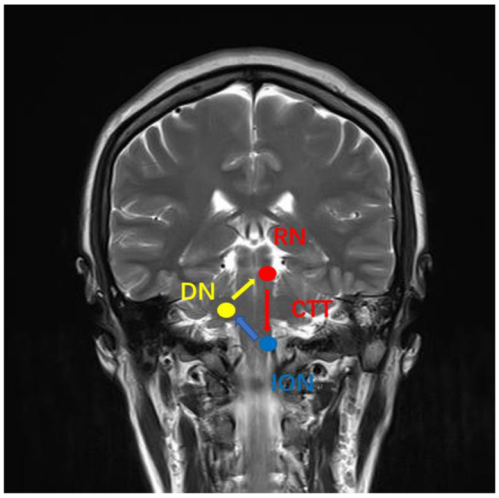

Fig. 2 Schematic drawing illustrating the Guillain-Mollaret triangle

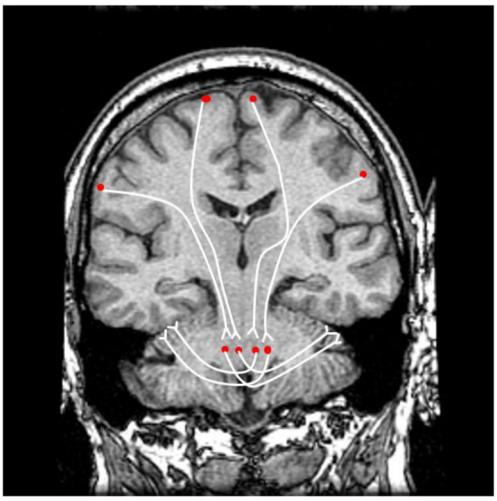

Fig. 3 Schematic drawing illustrating the crossed cortico-ponto-cerebellar pathways

contralateral HOD, while lesions of the central tegmental tract result in ipsilateral HOD. In our patient, the lesion from his prior hemorrhage affected the left red nucleus and central tegmental tract located in the midbrain or pontine tegmentum. Impact to either of these regions can result in HOD. Of note, HOD can be bilateral for cases in which a midline lesion crosses the central tegmental tract at the Wernekink commissure [6]. Lesions of the inferior cerebellar peduncle do not cause HOD.

Wallerian degeneration is the process of progressive demyelination and disintegration caused by injury, most commonly an infarction, to proximal axons or cell bodies [7]. It is most frequently observed in the corticospinal tract. Bilateral WD of the middle cerebellar peduncles is rarely reported and has various etiologies [8]. The WD of middle cerebellar peduncles should be considered in the differential for many diseases including the Wilson's disease, hepatic encephalopathy, extrapontine myelinolysis, multiple system atrophy, and spinocerebellar degeneration, as well as for infarction involving the bilateral anterior inferior cerebellar territory. Ataxia, dysarthria, vertigo, and hearing impairment are potential clinical manifestation of WD of middle cerebellar peduncles. Our patient presented with vertigo and ataxia. The middle cerebellar peduncle contains fibers of the crossed cortico-pontinecerebellar tracts. These fibers originate in the contralateral pontine nuclei and receive afferent information from the cortex through the cortico-pontine pathway (Fig. 3). A unilateral pontine lesion interrupts the ipisilateral cortico-pontine fibers and the crossing contralateral pontinecerebellar fibers. For this reason, both of the middle cerebellar peduncles can be affected by a unilateral injury in paramedian pontine. In our patient, the primary lesion in the pontine crossed over the midline. When WD occurs in this manner, bilateral involvement of the middle cerebellar peduncles is unavoidable.

Conclusion

In this study, we report on a primary lesion to the red nucleus and central tegmental tract that caused unilateral HOD, and the lesion that affected either the crossed or bilateral cortico-pontine fibers resulting in bilateral WD of middle cerebellar peduncles.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

References

1. Konno T, Broderick DF, Tacik P, Caviness JN, Wszolek ZK (2016) Hypertrophic olivary degeneration: a clinico-radiologic study. Parkinsonism Relat Disord 28:36–40. https://doi.org/10.1016/j.parkreldis.2016.04.008

2. Shen Y, Nie H,(2018) Wallerian degeneration of the bilateral middle cerebellar peduncles secondary to pontine infarction. Neurological Sciences 39 (5):961–963
3. Goto N, Kaneko M (1981) Olivary enlargement: chronological and morphometric analyses. Acta Neuropathol 54(4):275–282
4. Kalla R, Meichtry J, Schumacher R, Cazzoli D, Wiest R, Seifert E, Müri R (2016) Dancing jaw and dancing eyes. JAMA Neurol 73(1): 122. https://doi.org/10.1001/jamaneurol.2015.2299
5. Matsuo F, Ajax ET (1979) Palatal myoclonus and denervation supersensitivity in the central nervous system. Ann Neurol 5(1): 72–78. https://doi.org/10.1002/ana.410050111
6. Zhou C, He Y, Chao Z, Zhu Y, Wang P, Wang X, Liu S, Han W, Wang J (2017) Wernekink commissure syndrome secondary to bilateral caudal paramedian midbrain infarction presenting with a unique "heart or V" appearance sign: case report and review of the literature. Front Neurol 8:376. https://doi.org/10.3389/fneur.2017.00376
7. De Simone T, Regna-Gladin C, Carriero MR, Farina L et al (2005) Wallerian degeneration of the pontocerebellar fibers. AJNR 26: 1062–1065
8. Nabavizadeh SA, Mowla A, Mamourian AC (2015) Wallerian degeneration of the bilateral middle cerebellar peduncles.[J]. J Neurol Sci 349:256–257