Wallerian Degeneration of the Bilateral Middle Cerebella Peduncles Secondary to Unilateral Pontine Hemorrhage

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Abstract
The bilateral and symmetrical Wallerian degeneration (WD) of the middle cerebella peduncles (MCPs) is rarely reported in pontine hemorrhage. We described a patient with WD of bilateral MCPs secondary to a strip of unilateral and transversal pontine hemorrhage. Magnetic Resonance Imaging showed hyperintensity on T2-weighted, fluid attenuation inversion recovery and diffusion weighted image, and hypointensity on T1-weighted and apparent diffusion co-efficient map, and no enhancement in bilateral MCPs. These abnormal signals existed over 6 months and subsided gradually. The patients didn’t present with new symptoms when WD happened, the initial clinical manifestations related to pontine hemorrhage persisted for more than one year. In conclusion, a single unilateral pontine stroke along “cross” or “trident” line may cause WD of the bilateral MCPs in the below slice. Although this pathological change brings no additional new symptoms, it is related to the short-term of poor neurological prognosis after pontine stroke. In addition, since this lesion appears restricted diffusion in the imaging, we should avoid misdiagnosing it as new infarction.

INTRODUCTION
Wallerian degeneration (WD) is the process of progressive demyelination and disintegration of the distal axonal segment following the transection of the axon or damage to the neuron. The nerve fibers in the pons are concentrated and complex, particularly in the basis pontis. Some lesions at this special location may lead to bilateral and symmetrical WD of the middle cerebella peduncles (MCPs) [1, 2]. Recently, some cases and small-scale retrospective studies have reported this phenomenon after pontine infarction [3-5]. To our knowledge, this bilateral WD of the MCPs is rarely reported in pontine hemorrhage [6]. In addition, it is not completely clear about the clinical and radiological features of this neuropathology. Here, we described a patient with bilateral WD of the pontocerebellar fibers secondary to unilateral pontine hemorrhage, and characterized the evolutions of the clinical symptoms and brain imaging.

CASE DESCRIPTION
A 51-year-old Chinese female with hypertension, suddenly had vertigo, right sided numbness and weakness, dysarthria and a brief of coma seven months ago. The patient was diagnosed as left pontine hemorrhage by brain CT (Fig.1 A). After she was treated by controlling blood pressure with Telmisartan and rehabilitation, her cerebral hemorrhage disappeared and the right hemiplegia and dysarthria almost recovered. Nevertheless, the patient still suffered from dizzy and unstable on sitting, standing or walking. At last, she was referred to our hospital. Neurological examination revealed slight numbness and weakness of right limbs. Furthermore, her right sided Finger-to-Nose test and Heel-to-Shin test were overshooting, and she was unsteady when standing. Moreover, there were brisker tendon reflex and Babinski’s reflex on right side. The Modified Rankin Scale (mRS) score was 3. Magnetic Resonance Imaging (MRI) revealed a hypointensity lesion in basis pontis on T1- and T2-weighted images, which represented a previous hemorrhage (Fig.1 B, C). At same time, the symmetrical lesions in bilateral MCPs were seen as hyperintensity on T2-weighted, fluid attenuation inversion recovery (FLAIR) and diffusion weighted image (DWI), and hypointensity on T1-weighted and apparent diffusion co-efficient (ADC) map, and no enhancement on contrast MRI (Fig.1 D-H). Magnetic
resonance angiography showed normal in the intracranial arteries (Fig.1 I). Laboratory and cerebral spinal fluid tests showed normal condition. Due to no new stroke and blood pressure steady, the patient was discharged without an additional treatment. After 6 months, the patient was referred to our institution for a reexamination. Her dizzy and unstable feelings became significantly alleviated, whereas neurological examination still revealed the remnant of slight numbness and weakness of right limbs. Meanwhile, the mRS score decreased to 1. In addition, second MRI showed that high signals of bilateral MCPs became smaller and weaker than before (Fig.1 J-L).

DISCUSSION

In the basis pontis, there are fibres that are arranged longitudinally and transversely. The longitudinal fibres are mainly the corticospinal tract (CST) which project through the central portion of the basis pontis. The pontine nuclei surround the longitudinal fibres and give rise to the pontocerebellar fibres. These fibres are oriented transversely, cross the midline as the pontine crossing fibers (PCF) and enter the cerebellum as the MCP. On conventional T2-weighted image of some neurodegenerative and metabolic diseases, in which there is involvement of the PCFs and sparing of the CSTs and tegmentum, the PCFs can be clearly seen as the “hot cross bun sign” or “trident sign”[1]. A unilateral pontine lesion not only damages the ipsilateral pontine nuclei and their axons, which will have to cross the midline to reach the contralateral MCPs, but also injures the axons originating from the contralateral pontine nuclei that have crossed the midline and course through the pons to reach the ipsilateral MCPs. Therefore, a unilateral pontine lesion along “cross” or “trident” line, such as a strip of paramedian pontine infarction or transversal pontine hemorrhage, is enough to cause the WD of bilateral MCPs (Fig.1 B). Moreover, this pontine lesion is commonly located in the slice above the WD of MCPs (Fig.1 B, D).

Our case showed high signals on DWI and low signals on ADC map in bilateral MCPs after pontine hemorrhage, which subsided gradually but existed over 6 months. In addition, our case didn’t present with new symptoms when WD happened. Meanwhile, the clinical manifestations related to pontocerebellar fibers, such as ataxia, dysarthria, and vertigo, occurred immediately after the pontine hemorrhage, and persisted for more than one year. The mRS score also chronically improved for a long time in spite of active rehabilitation. These clinical features may be explained by the following reasons. First of all, it is a consecutively pathophysiological process from initial pontine stroke to secondary WD of the MCPs. Abnormal signals of MCPs may be only the delayed manifestation of WD since some studies reported this MRI change usually happened in the third stage of WD [7]. Therefore, no additional symptoms will happen. Secondly, some study demonstrated that WD in the corticospinal tract or the bilateral MCPs might hinder the process of recovery of neurological function by the application of the diffusion tensor imaging (DTI) [8-10]. A clinical retrospective study suggested that the patients with WD had a worse outcome for disability at 90 days than without WD, which hints the association of WD following pontine stroke with the poor short-term prognosis [3].
Since there were multiple vascular risk factors in this patient, the WD of MCPs must be differentiated from a new infarction in MCPs. However, the infarction could not explain this specific MR finding for the following reasons [11]. Firstly, the high intensity on DWI usually disappears after two weeks and enhancement exists in the acute infarction. Secondly, the bilateral symmetrical nature of the lesions does not reflect a single arterial territory, because the MCPs are supplied by the anterior inferior cerebellar artery and the neighbouring structures within the same vascular territory were not involved in this case. Thirdly, this patient did not report sudden-onset symptoms before MRI scan. Moreover, the symmetrical signal abnormalities of bilateral MCPs can also be seen in other neurological diseases, such as demyelination, neurodegenerative, toxic and metabolic diseases, and neoplasms. These diseases could be excluded because they usually have special symptoms, signs, abnormal laboratory tests and other abnormal imaging [1, 2].

In conclusion, a strip of unilateral pontine infarction or hemorrhage along “cross” or “trident” line may cause WD of the bilateral MCPs in the below slice. Although this pathological change brings no additional new symptoms, it is related to the short-term of poor neurological prognosis after pontine stroke. We should avoid misdiagnosing it as a new infarction.

Informed consent: Informed consent was obtained from patient.

REFERENCES