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Case Report

Diffusion Tensor Imaging in a Case of Pontine Bleeding Showing Hypertrophic Olivary Degeneration and Cerebellar Ataxia

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Keywords
Diffusion tensor imaging · Tractography · Hypertrophic olivary degeneration · Central tegmental tract · Cerebellar ataxia

Abstract
We present diffusion tensor tractography (DTT) findings in a case of hypertrophic olivary degeneration (HOD) and cerebellar ataxia. A 56-year-old man presented with abnormal ataxic gait and dysarthria. MRI 5 months after onset showed chronic pontine hematoma and enlarged bilateral inferior olivary nuclei. DTT showed decreased volume of the bilateral central tegmental tract, in accordance with the conventional hypothesis that HOD is associated with neurologic insult to the Guillain-Mollaret triangle. The patient's cerebellar ataxia was speculated to be due to decreased decussating fibers of the superior cerebellar peduncle, and this was confirmed by DTT.

Introduction

Hypertrophic olivary degeneration (HOD) is a rare condition caused by a unique pattern of trans-synaptic degeneration, thought to be associated with lesion of the Guillain-Mollaret triangle [1]. Pathologically, HOD is characterized by enlargement and vacuolation of neurons,

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astrocytic hyperplasia, and demyelination in the inferior olivary nuclei [2]. According to previous case reports, bilateral HOD is more common than unilateral HOD [3]. Radiologically, it is often seen several months after the original insult and the MRI T2 signal increase lasts for 3–4 years.

The differential diagnosis of causative diseases includes infarction, hemorrhage, tumors, demyelinating lesions, or less frequently, traumatic lesions involving the Guillain-Mollaret triangle. Some neurodegenerative diseases such as olivopontocerebellar atrophy or progressive supranuclear palsy can also cause HOD [4, 5], and we have previously reported a case of spinocerebellar ataxia type 2 with bilateral HOD [6].

Diffusion tensor tractography (DTT), which is performed using data from diffusion tensor imaging, makes it possible to estimate the direction of diffusion of water molecules in the axon and to visualize the fibers running in the white matter. It has been applied extensively to map white matter connectivity in cases of cerebral infarction, demyelinating diseases, brain tumor, and so on. It has also been applied clinically for the presurgical mapping of eloquent white matter tracts before intracranial mass resections.

In this case report, we describe the application of DTT to our patient with bilateral HOD to investigate the white matter connectivity within the Guillain-Mollaret triangle, focusing particularly on the decrease in the volume of the central tegmental tract. We present the characteristic neurological symptoms of our patient, who showed combined medial longitudinal fasciculus (MLF) syndrome and cerebellar ataxia, and the DTT findings of fiber connectivity at the decussation of the superior cerebellar peduncles, which was presumed to be the responsible lesion.

Case Presentation

The patient, a 56-year-old man with a known cavernous angioma at the pons, suddenly noticed difficulty in walking and speech disturbance. A diagnosis of large pontine bleeding was made based on head CT and T2-weighted MRI at an acute care hospital (Fig. 1), and angioma resection was performed 1 month later. His neurological condition gradually improved, and he was transferred to our rehabilitation hospital 2 months after the bleed. Neurological examination at our hospital revealed marked dysarthria, dysphagia, abnormal ocular movements due to MLF syndrome, right dominant facial nerve palsy, and truncal and bilateral limb ataxia. There was no weakness or sensory disturbance of the extremities. Palatal myoclonus was not observed during his course.

Conventional MRI of the brain and DTT were performed 5 months after the onset with a 1.5-T superconducting system (Magnetom AERA; Siemens). Fluid-attenuated inversion recovery (FLAIR) images revealed a regressed hematoma at the left side of the paramedian part of the pontomesencephalic tegmentum. Bilateral enlarged inferior olivary nuclei were clearly observed as high-signal intensity lesions (Fig. 2).

Diffusion tensor imaging was performed by using a monopolar (Stejskal-Tanner) sequence with simultaneous multi-slice single-shot spin-echo echo-planar imaging (TR/TE, 4,500/75 ms; matrix size, 128 × 128; FOV, 22 × 22 cm, section thickness, 3.0 mm, slice acceleration ×2). We obtained diffusion-weighted images along 20 different directions with a *b*-value of 1,000 s/mm^2, as well as an image without diffusion weighting (b = 0 s/mm^2). Brain fiber tracking was performed by using an AZE Virtual Place Arata NT workstation, and fiber tractography maps were generated.

There was decreased volume of the bilateral central tegmental tract with interruption at the level of the inferior olive. In addition, a decrease in the volume of the decussating fibers at the superior cerebellar peduncle was seen, consistent with the dorso-rostral extension of the hematoma (Fig. 3).

Discussion

HOD is suspected to be produced in response to neurologic insult to the dentato-rubro-olivary pathway. This pathway, known as the Guillain-Mollaret triangle, connects the red nucleus of the midbrain, the inferior olivary nucleus of the medulla, and the contralateral dentate nucleus of the cerebellum. The fibers from the red nucleus descend in the central tegmental tract to the inferior olivary nucleus, and lesions that disrupt the central tegmental tract are likely to result in HOD.

Our patient with large pontine bleeding from cavernous angioma clearly showed signal abnormalities of the bilateral inferior olivary nuclei on FLAIR MRI taken 5 months after the onset. In general, on conventional MRI, three distinct stages of HOD with specific time intervals have been described [2]: (1) T2 hyperintense signal of the inferior olivary nucleus without hypertrophy within a few months after the disruptive event, (2) T2 hyperintense signal and hypertrophy of the inferior olivary nucleus (that is, HOD) up to 3–4 years thereafter, and (3) T2 hyperintense signal with progressive resolution of the hypertrophy. The MR change in our case was consistent with stage 2.

On DTT, we could clearly see the decrease of the central tegmental tract volume, supporting the view that the HOD was caused by disruption of this tract. As conventional MRI could not reveal involvement of the components of the Guillain-Mollaret triangle, there has been no other way to demonstrate them than histopathological techniques on postmortem specimens. However, DTT has made it possible to visualize nerve tracts, and thus neurological assessments can be related to these images.

Interestingly, our patient showed abnormal eye movement due to MLF syndrome and bilateral cerebellar ataxia. The combination of these neurological signs is characteristic and is relatively rare [7–9]. The MLF lies in the paramedian portion of the upper brainstem and the lesion in this fasciculus shows unique abnormal eye movements (so-called MLF syndrome). MRI showed that the hematoma in our patient included the MLF and we speculated that the dorso-rostral extension of the hematoma involved the decussation of the superior cerebellar peduncle, causing truncal and bilateral limb ataxia. In support of this speculation, we observed a reduction of the decussating fibers of the superior cerebellar peduncle on DTT. In addition, our patient did not show palatal myoclonus, the classical presentation of HOD; however, it has been reported that this symptom does not appear in all patients with HOD [10].

In conclusion, DTT in our patient revealed the disruption of pathways associated with HOD in a manner consistent with the known pathologic change. DTT could be a useful tool for understanding complicated neurological symptoms in relation to fiber connectivity.

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Statement of Ethics

The authors have no ethical conflicts to disclose.

Disclosure Statement

The authors declare that there is no conflict of interest regarding the publication of this article.

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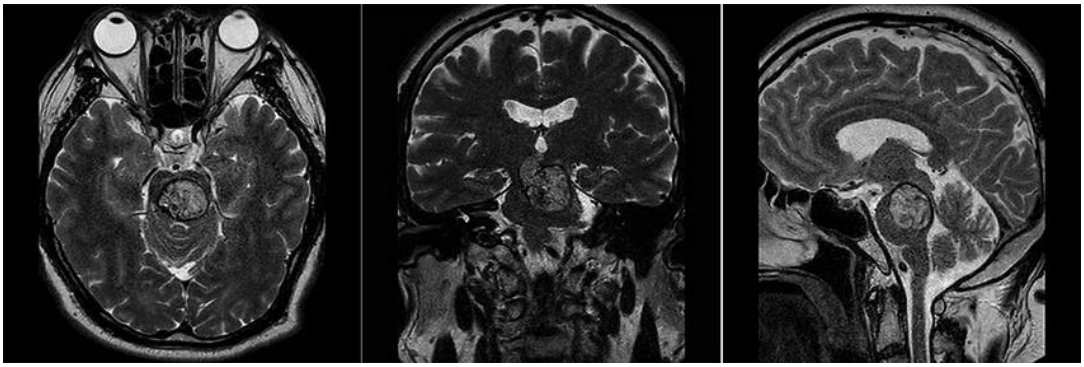

Fig. 1. T2-weighted MRI (TR 5,200 ms, TE 90 ms) just after the onset showed an inhomogeneous large bleeding mass at the paramedian part of the pontomesencephalic tegmentum.

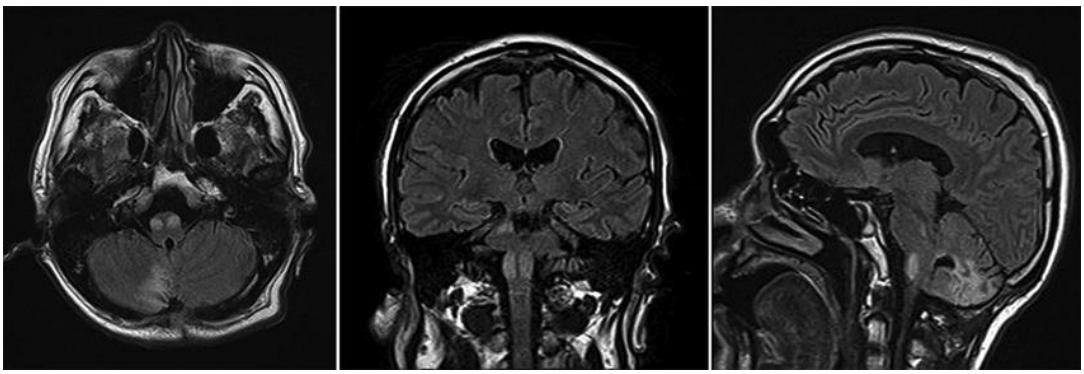

Fig. 2. FLAIR MRI (TR 9,000 ms, TE 108 ms) taken 5 months after the onset showed enlarged symmetric hyperintense inferior olivary nuclei.

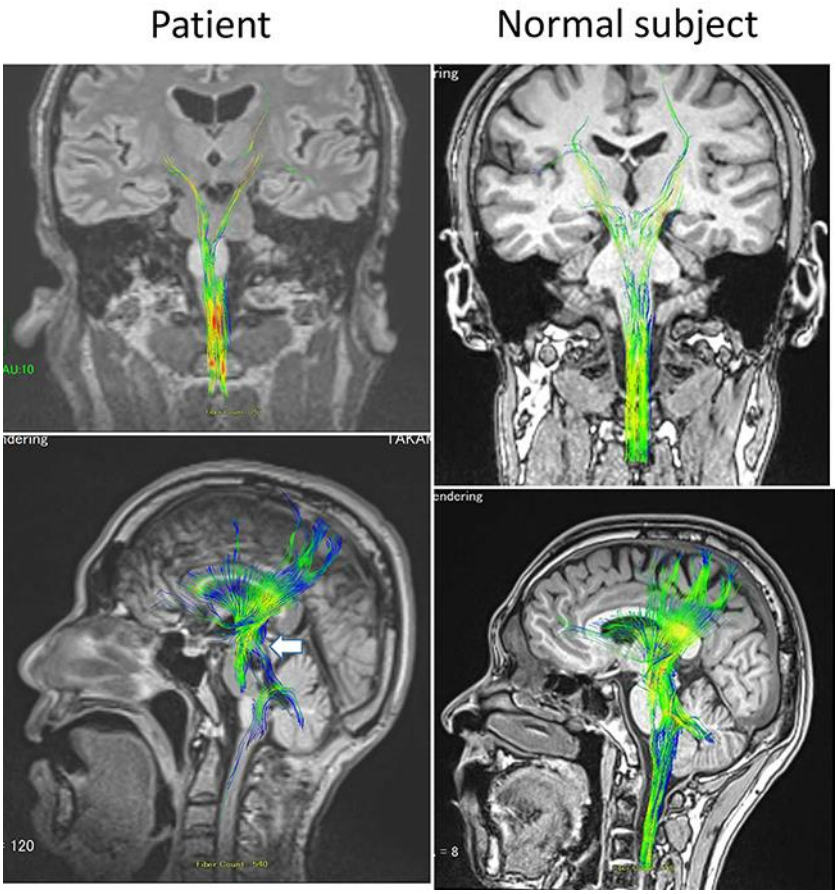

Fig. 3. Compared with DTT of a normal subject (38-year-old male), the patient showed decreased fiber volumes at the central tegmental tract and decussation of the superior cerebellar peduncle (arrow).