Downbeat Nystagmus Due to a Paramedian Medullary Lesion

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ABSTRACT

Cell groups of the paramedian tract, which are located in the paramedian region of the lower brainstem, are eye-movement-related neurons that project to the cerebellar flocculus. Their inactivation produces downbeat nystagmus, which resembles eye movement disorders resulting from lesions of the cerebellar floccular in an animal experiment. Therefore, paramedian tract cells are assumed to fulfill an important function in ocular movement control, such as gaze-holding and maintaining vestibular balance.

This paper is a report of a case involving a 50-year-old female who manifested downbeat nystagmus due to damage to paramedian tract cells caused by a localized ischemic lesion in the medulla oblongata. The finding that a paramedian medullary lesion induced nystagmus similar to that observed following floccular lesions clearly indicates that a subgroup of paramedian tract cells projecting to the flocculus was impaired. This finding has important implications in considering the presence of a brainstem-cerebellar feedback loop which is involved in vestibulo-oculomotor controls, such as vestibular balance.

Although there have been a few published reports of cases of downbeat nystagmus caused by lesions in the midline region of the lower brainstem, none has been published to date reporting the occurrence of nystagmus due to a strictly localized medullar lesion, such as the one described in this paper.

＜Keywords＞

Downbeat nystagmus; paramedian tract neuron; flocculus; neural integrator; vestibular imbalance; lower brainstem; cerebellum; cerebral infarction

INTRODUCTION

Vertical nystagmus due to small lesions in the lower brainstem, often manifests as upbeat nystagmus. Downbeat nystagmus, however, is often caused by cerebellar floccular lesions (1). Cell groups of the paramedian tract (PMT) occur in and around the medial longitudinal fasciculus in the paramedian region of the lower brainstem. These PMT cells project to the cerebellar flocculus and are involved in ocular movements (2, 3). Although PMT cells have long been suggested to fulfill an important function in the control of eye movements, the mechanism and clinical relevance are poorly understood. In fact, limited evidence is available from animal experiments (3) and clinical studies (4-6) on downbeat nystagmus associated with impaired PMT cells.

This paper is a report of a case of manifested downbeat nystagmus due to damaged PMT cell subgroups caused by a localized lesion in the paramedian region of the medulla tegmentum.

CASE REPORT

The subject, a 50-year-old female, at the age of 42, manifested right-sided lateral medullary syndrome caused by occlusion of the right posterior inferior cerebellar artery. Magnetic resonance imaging (MRI) of her brain showed a small, new ischemic lesion in the right lateral portion of the medulla oblongata.

However, she led a normal life afterward without symptoms of nystagmus. She manifested dizziness and oscillopsia at the age of 47 and visited our clinic with persistent oscillopsia on lateral gaze three months after the onset of symptoms. Her neurological examinations revealed downbeat nystagmus during fixation and mild crossed sensory deficits in the right part of the face, left extremities, and trunk, but she had no ataxia or paralysis. Previous right-sided lateral medullary syndrome was suspected to be responsible for the crossed sensory disturbances. The downbeat nystagmus observed in the present case lasted more than three years from the onset of symptoms. An ischemic lesion in the right dorsal paramedian portion of the medulla oblongata was shown in thin-slice brainstem MRI (Figure 1A, B).

Her eye movements were also recorded with electronystagmography (First, Tokyo, Japan), using a direct current (DC) recording technique.

Downbeat nystagmus was observed in the light and in the dark. Downbeat nystagmus was observed in the absence of fixation (rapid phase: velocity, 61.7°/sec; amplitude, 4.7°; Slow phase: velocity, 14.9°/sec; amplitude, 4.4°; frequency, 2.1/sec) (Figure 2). Downbeat nystagmus was accentuated in the lateral eye position and was not influenced by the upward and downward eye positions of the eyes in the orbit and the position of the head.

DISCUSSION

Because the MRI revealed an ischemic lesion in the paramedian tegmental region of the medulla oblongata after the occurrence of nystagmus, the lesion in the paramedian region, in which cell groups of PMT are located, was suspected to be responsible for the downbeat nystagmus observed.

Cell groups of PMT fulfill an important function in gaze holding and the maintenance of vestibular balance, and impaired PMT cells have been shown to induce downbeat nystagmus in an animal experiment (3). In the present case, the medullary lesion was much lower than the location of PMT cells controlling eye movements and supposed to be at the medullary-pontine junction in the animal (3), as also suggested by a previously reported human lesion (6). However, PMT cell groups spread from the pons to the lower medulla oblongata and project to the cerebellar flocculus (2). Furthermore, the eye movement disorders seen in the present case resembled those observed in the floccular lesions (7). According to previous reports, cerebellar flocculus and paraflocculus fulfill important functions in maintaining gaze holding and vestibular balance, and the impairment of these regions induces downbeat nystagmus (1, 7). In the present case, impairment of PMT cells might induce disorders similar to those resulting from floccular lesions. This finding suggests that feedback from the brainstem might influence the ocular movement control mechanisms in the cerebellum.

Figure 3 is a summary of the pathogenesis of downbeat nystagmus caused by lesions in cell groups of PMT (3). Lesions in PMT cells attenuate the activity of floccular Purkinje cells, which, in turn, reduce the inhibition of secondary vestibular nucleus neurons that receive inputs from the anterior semicircular canals. However, the neuronal activity of the posterior canal-related secondary vestibular nucleus neurons remains unaffected because the neurons do not receive inhibitory regulation from the flocculus (8). As a result, only the anterior canal-related secondary vestibular nucleus neurons are activated, which induces upward eye movements that constitute the slow phase of nystagmus. This is then followed by compensatory fast-phase beating in a downward direction, resulting in downbeat nystagmus. It is suspected that lesions in PMT cells might be responsible for central vestibular imbalance in the vertical direction, suggesting that a PMT-flocculus-vestibular nucleus pathway might fulfill an important function in maintaining vestibular balance.

This case is unique in that downbeat nystagmus was caused by impairment of the vestibular imbalance as a result of damage caused to PMT cells by small infarctions in the brainstem.

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<Figure Legends>

Figure 1: Brain MRI images

A T1-weighted midsagittal image in Fig. A shows low-intensity lesions in the medulla oblongata.

A T2-weighted axial image in Fig. B shows punctuated, high-intensity lesions in the tegmentum and the right paramedian region of the dorsal medulla oblongata. These lesions, which are probably small infarctions, are believed to be responsible for nystagmus.

Figure 2: Electronystagmography findings (DC recording).

Results for eye movements in the absence of fixation (eyes open in the dark).

The upper section shows the vertical eye position, whereas the lower section shows the corresponding eye movement velocity.

Downbeat nystagmus with fast-phase lowering and slow-phase rising was noted.

Figure 3: Pathogenesis of downbeat nystagmus (3)

This figure shows how vertical eye movements might be generated.

When cell groups of PMT are impaired by lesions in the lower brainstem, the activity of floccular

Purkinje cells is attenuated, leading to reduced inhibition of anterior canal-related secondary vestibular nucleus neurons. This results in the activation of these neurons. However, neuronal activity of the posterior canal-related secondary vestibular nucleus neurons remains unaffected because they do not receive inhibitory regulation from the flocculus (8). Thus, only the anterior canal-related superior vestibular nucleus neurons are activated, resulting in the motoneuron's neuronal activity to produce reciprocal eye movements in the vertical plane. This induces upward eye movements that constitute the slow phase of nystagmus, which is then followed by the compensatory fast-phase beating in a downward direction, resulting in downbeat nystagmus.

P: Purkinje cells; G: cerebellar granule cells

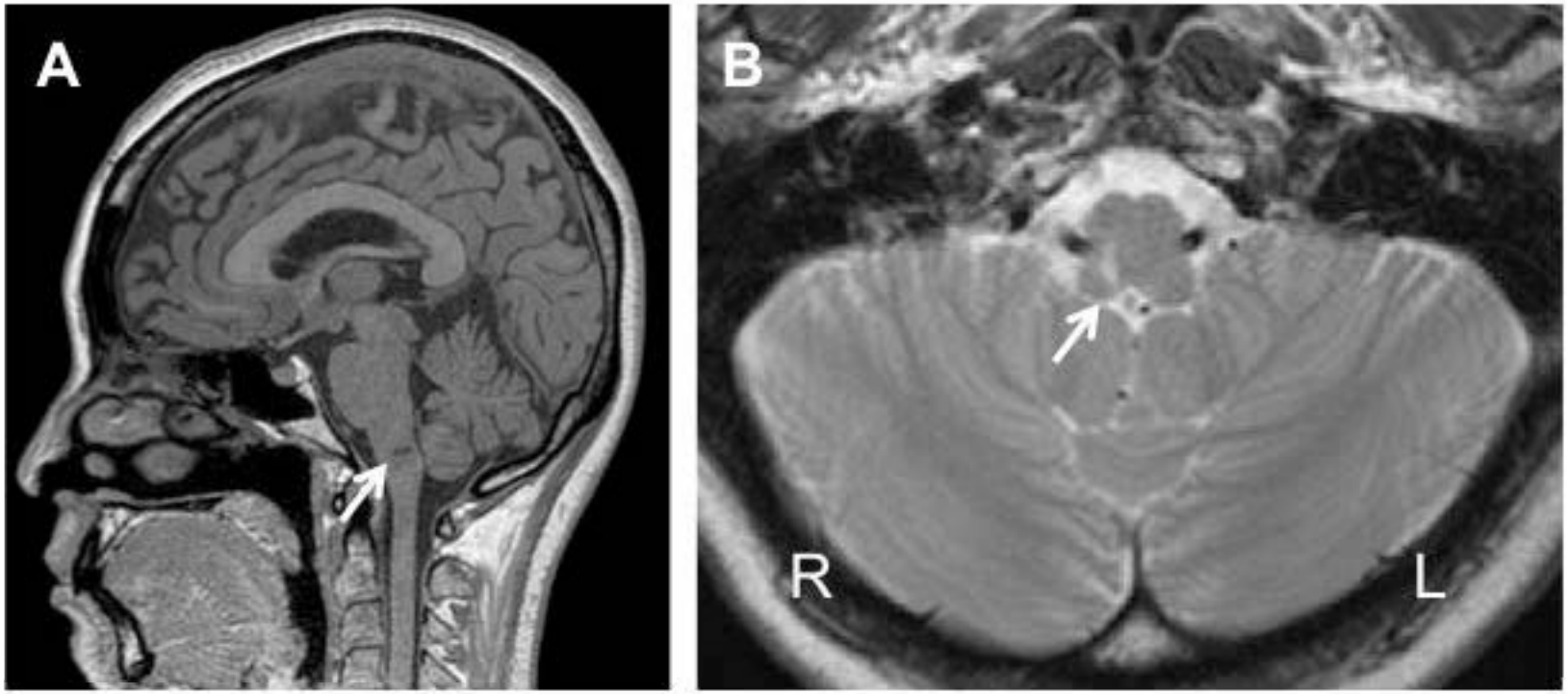

Fig. 1

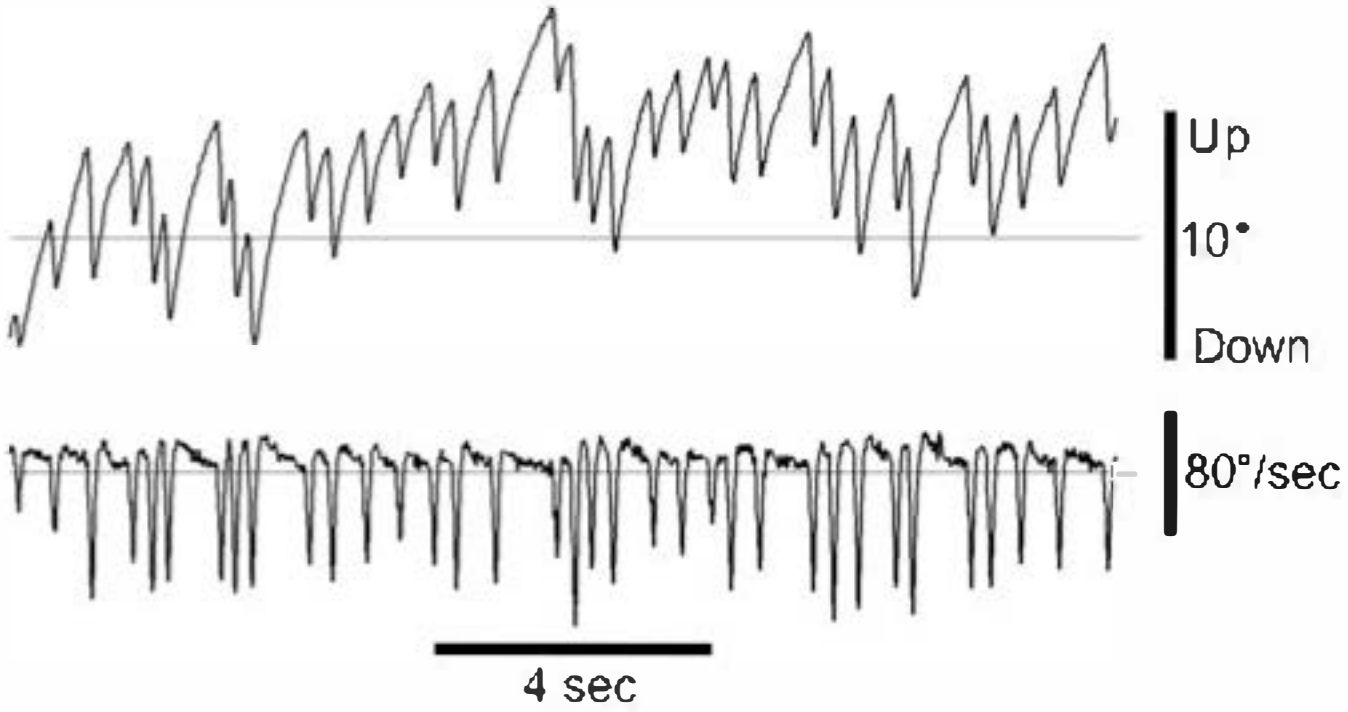

Fig. 2

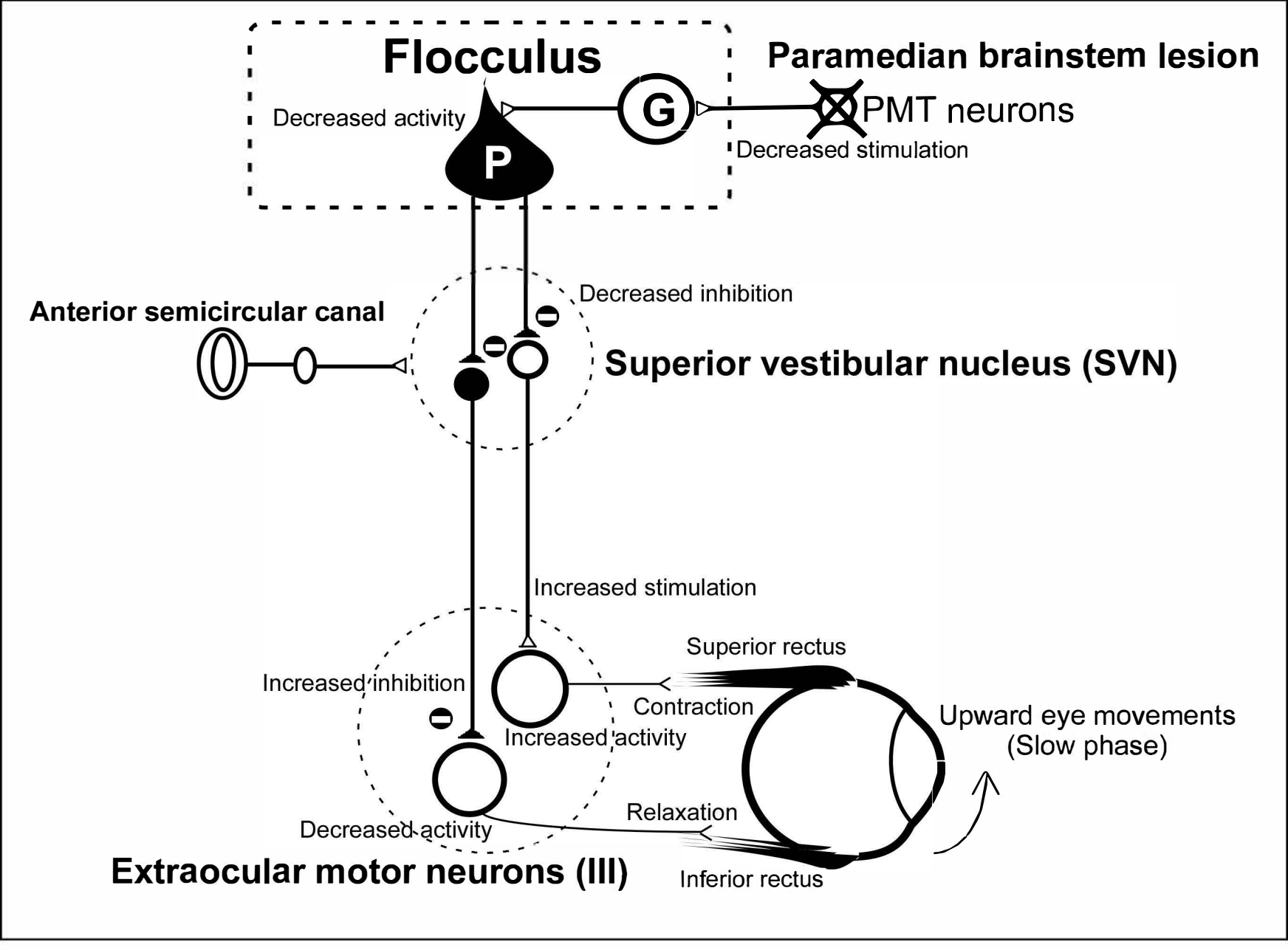
Flocculus
Paramedian brainstem lesion
Decreased activity
G
PMT neurons
P
Decreased stimulation
Anterior semicircular canal
Decreased inhibition
Superior vestibular nucleus (SVN)
Increased stimulation
Superior rectus
Increased inhibition
Contraction
Upward eye movements
(Slow phase)
Increased activity
Relaxation
Decreased activity
Extraocular motor neurons (III)
Inferior rectus

Fig. 3