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Case Report

Hemidystonia after Pontine Hemorrhage Successfully Treated with Pharmacotherapy and Intensive Rehabilitation: a Case Report

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Correspondence to
Seo Yeon Yoon
Department of Physical Medicine and Rehabilitation, Korea University Guro Hospital, 148 Gurodong-ro, Guro-gu, Seoul 08308, Korea.
E-mail: seoyeon0521@gmail.com

HIGHLIGHTS

- We reported a case of hemidystonia with loss of proprioception by pontine hemorrhage.
- Reports of the effect of rehabilitation in dystonia by pontine hemorrhage were rare.
- Pharmacotherapy with rehabilitation was effective in dystonia by pontine hemorrhage.

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Hemidystonia after Pontine Hemorrhage Successfully Treated with Pharmacotherapy and Intensive Rehabilitation: a Case Report

Gyu Seong Kim [1], **Yeon Gyu Jeong** [1], **Yoon Jeong Jeong** [1], **Seo Yeon Yoon** [2]

[1]Department of Rehabilitation Medicine, Bundang Jesaeng General Hospital, Seongnam, Korea
[2]Department of Physical Medicine and Rehabilitation, Korea University Guro Hospital, Seoul, Korea

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Correspondence to
Seo Yeon Yoon
Department of Physical Medicine and Rehabilitation, Korea University Guro Hospital, 148 Gurodong-ro, Guro-gu, Seoul 08308, Korea.
E-mail: seoyeon0521@gmail.com

ORCID iDs
Gyu Seong Kim
https://orcid.org/0000-0003-2352-8724
Yeon Gyu Jeong
https://orcid.org/0000-0003-0456-245X
Yoon Jeong Jeong
https://orcid.org/0000-0002-6207-6372
Seo Yeon Yoon
https://orcid.org/0000-0002-0365-2923

Conflict of Interest
The authors have no potential conflicts of interest to disclose.

ABSTRACT

Dystonia is a movement disorder characterized by involuntary contraction of muscles resulting in repetitive or twisting movements. Dystonia is generally caused by basal ganglia dysfunction. Recent studies have reported an association between dystonia and brainstem disorders. However, the pathological mechanism is uncertain, and detailed management strategies are limited. Here, we report a case of hemidystonia with abnormal posture and impaired proprioception after pontine hemorrhage that was effectively treated with pharmacotherapy combined with early intensive comprehensive rehabilitation. A 45-year-old man presented with abnormal posture and dystonic movement in the right hand and foot after a pontine hemorrhagic stroke. Pharmacotherapy with clonazepam and benztropine was administered, and comprehensive rehabilitation programs were implemented intensively from the early stages of symptom onset. After 3 months, the patient was able to walk independently, go up and down a few stairs without the use of a handrail, and was able to perform activities of daily living with minimal assistance.

Keywords: Dystonia; Brain Stem; Pons; Rehabilitation; Intracranial Hemorrhages

INTRODUCTION

Dystonia is a movement disorder characterized by repetitive involuntary co-contractions of agonist and antagonist muscles, resulting in abnormal posture [1-3]. Dystonia is generally considered to be caused by basal ganglia dysfunction [3,4]. Recent studies have suggested that dystonia can also be caused by disorders of the thalamus, cerebellum, and brainstem and has the property of network disorders related to the cerebello-thalamo-cortical pathway [4-6]. However, the pathological anatomy and mechanisms are unclear. Although there are a few case reports regarding hemidystonia as a consequence of brainstem lesions, brain lesions were not limited to the brainstem [2-4,7] in most other cases. Several medications, including anticholinergics and benzodiazepines, have been used to manage dystonia, and clonazepam is the most frequent choice for dystonia after stroke in previous studies [2,3,5,8-11]. However, detailed management strategies and rehabilitation approaches are limited. Here, we report a case of hemidystonia with abnormal posture and impaired proprioception in the right upper and lower extremities after pontine hemorrhage that was effectively treated with pharmacotherapy combined with intensive and comprehensive rehabilitation.

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CASE REPORT

A 45-year-old man with no known medical history was admitted to our hospital with a sudden onset of right-sided weakness on April 18, 2020. An acute intracerebral hemorrhage in the bilateral pons, approximately 2.7 cm, was confirmed by brain computed tomography. The lesion was located mainly in the left pons, closer to the midbrain than to the medulla and involved the left middle cerebellar peduncle (**Fig. 1**). After conservative management, he was transferred from the Department of Neurosurgery to Rehabilitation Medicine on May 6, 2020. On initial evaluation, his range of motion was full in all the bilateral joints, and his affected side was a grade 4 in the manual muscle test. The Korean Mini-Mental State Examination score was 30/30. He showed abnormal posture and dystonic movement in the right hand and foot, which worsened while standing up or grasping objects. His right wrist joint was involuntarily contracting in a flexion pattern, and his right fingers were hyperextended. The affected side was hypertonic, causing cramping pain of numeric rating scale 8 without voluntary relaxation and was difficult for clinicians and physical therapists to release. Finger twisting and involuntary writhing movements were regularly monitored, and he could not utilize his right hand for activities of daily living (ADL). The severity of dystonia in this case was assessed using the Unified Dystonia Rating Scale (UDRS) [12] which includes ratings for 14 body regions and describes the proximal and distal parts of the arms and legs separately. The UDRS score of the affected side was 17.5. The Manual Function Test (MFT) score of the affected side was 18/32, and the Korean Modified Barthel Index (K-MBI) score was 14/100. He showed markedly impaired proprioception in the metacarpophalangeal joints of all fingers of the right hand and metatarsophalangeal joints of all toes of the right foot. He also presented with a slightly impaired light touch sensation, with an intact pain sensation in his right extremities. He showed limited extraocular movement of the medial gaze of the left eye, adducting nystagmus of the right eye, and diplopia. The patient complained of non-whirling dizziness which worsened with eye opening. He also showed intentional tremor, dysmetria, dysdiadochokinesia, and ataxia. Despite nearly intact muscle strength and cognitive function, the patient could not stand independently. Ambulation required maximal assistance or wheelchair locomotion. Transferring from bedside to wheelchair also required maximal assistance, and he was unable to sit on his own. His initial Berg Balance Scale score was 0/54.

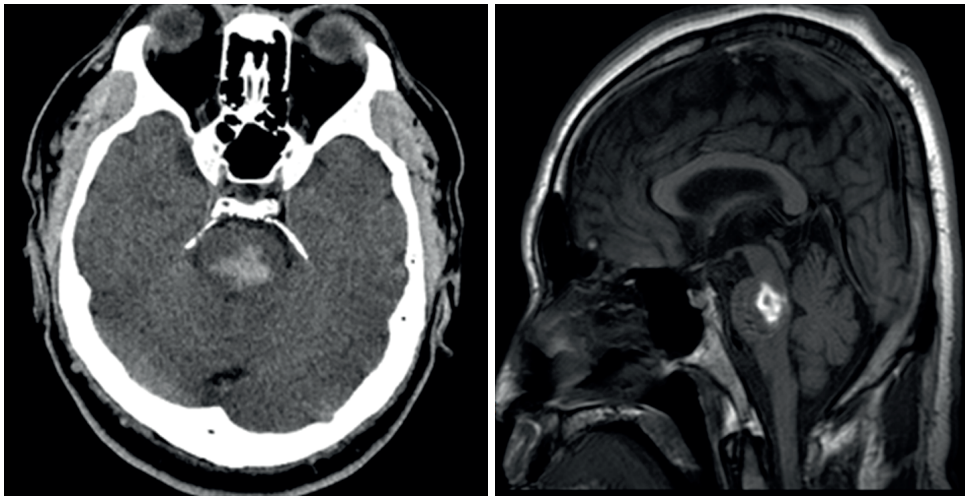

Fig. 1. The brain computed tomography shows acute pontine hemorrhage (left) and the sagittal T1-weighted magnetic resonance imaging shows the location of hemorrhage (right).

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For the management of dystonia, clonazepam 0.25 mg once daily (in the evening) was prescribed on the 24th day after onset and after 3 days, the dose was increased to 0.25 mg twice per day (in the morning and evening). There was a significant improvement in dystonia after administration of clonazepam. For the remaining dystonic feature, benztropine 0.5 mg once daily (in the morning) was added to the regimen after 8 days. Comprehensive rehabilitation programs consisted of 2 sessions of physical therapy and 2 sessions of occupational therapy per day. Each session lasted 30 minutes and was conducted 5 days a week for 12 weeks. For physical therapy, proprioceptive motor control training (using visuospatial compensation) and standing balance training were both performed for 30 minutes in the morning, while gait training was performed for 30 minutes in the afternoon. For occupational therapy, sensory and vibration stimulation training and fine motor training of the right hand were performed for 30 minutes in the morning, while bimanual ADL training was practiced for 30 minutes in the afternoon. Over the course of the treatment period, his symptoms gradually improved, especially in dystonia. His abnormal dystonic movements lessened in severity, and his UDRS score decreased from 17.5 to 7 points. Hypertonicity of his right hand and fingers improved, thus allowing voluntary control. On the one-month follow-up evaluation, the MFT score improved from 18 to 24/32, and the K-MBI score improved from 14 to 51/100. Other symptoms, such as ophthalmic problems, non-whirling dizziness, and cerebellar symptoms, also showed remarkable improvement. He could stand and walk independently for about 10 meters indoors, and his Berg Balance Scale (BBS) improved from 0 to 31/54. The medication and comprehensive rehabilitation programs were continued thereafter. At the 3-month follow-up evaluation, he could walk independently and go up and down several steps. The UDRS score decreased from 7 points to 3 points. BBS improved to 54/54, K-MBI scores improved to 64/100, and the MFT score improved to 25/32 (**Table 1**).

DISCUSSION

Hemidystonia is a manifestation of secondary dystonia associated with central nervous system lesions. Dystonia is mainly caused by lesions involving the basal ganglia, most commonly the caudate and the putamen [3,4]. Additionally, the thalamus, internal capsule, and cerebral cortex are pathoanatomically associated sites [4]. To date, only a few cases have reported pontine lesions associated with hemidystonia. Although the exact pathomechanism of dystonia after pontine lesions remains uncertain, the suggested mechanisms are as follows: disrupted sensory afferent input to the striatum or thalamus [3]; loss of pallidal inputs to the pedunculopontine fibers [3,4]; damaged dentate-rubro-olivary pathways [4,7]; and pontine structural lesions including those located in the medial lemniscus, central tegmental tract, and pedunculopontine nucleus [2].

In 2020, Park et al. [2] reported a case of abnormal posture with decreased proprioception in the left extremities after right pontine hemorrhage. The lesion was located in the right dorsal pontine, extending to the lower midbrain and fourth ventricle. The author also proposed 5 previous cases of hemidystonia after pontine lesions. In the presented case, the locations of the lesions extended to the lower midbrain, pontomedullary junction, or red nucleus and involved ophthalmic problems such as restriction of upward and horizontal conjugate gaze as a result of 6th and 7th cranial nerve palsy [3,4]. Brain magnetic resonance imaging showed cerebral hemorrhage in the bilateral pons with left middle cerebellar peduncle involvement, which was more rostral compared with the previous case reports. The involvement of the left medial longitudinal fasciculus resulted in ipsilateral medial gaze impairment that was different from the ophthalmic symptoms of the previous cases.

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Table 1. Changes in physical examinations, evaluations, symptoms and functional levels during 3 months of treatment

Characteristics	2020.05.07	2020.06.07	2020.08.05
Physical examinations			
ROM (degree)	Full range	Full range	Full range
MMT (grade)	Grade 4	Grade 5	Grade 5
Light-touch sensation	Slightly impaired	Slightly impaired	Nearly intact
Proprioception	Remarkably impaired	Impaired	Slightly impaired
Evaluations (score)			
K-MMSE	30/30	30/30	30/30
K-MBI	14/100	51/100	64/100
BBS	0/54	31/54	54/54
MFT*	18/32	24/32	25/32
UDRS*			
Shoulder and proximal arm	3.5	1.5	0
Distal arm and hand including elbow	6	3	1.5
Pelvis and proximal leg	2.5	0	0
Distal leg and foot including knee	5.5	2.5	1.5
Total	17.5	7	3
Symptoms			
NRS of cramping pain (score)	8/10	6/10	3/10
Non-whirling dizziness	Severe	Improved	Much improved
Opthalmologic symptoms	Prominent	Improved	Much improved
Cerebellar symptoms	Prominent	Improved	Much improved
Functional levels			
Ambulation	Wheel chair dependent	Independent gait about 10 m	Independent gait and stair up/down
ADLs	Maximal assist	Moderate assist	Minimal assist

ROM, range of motion; MMT, manual muscle test; K-MMSE, Korean Mini-Mental State Examination; K-MBI, Korean Modified Barthel Index; BBS, Berg Balance Scale; MFT, Manual Function Test; UDRS, Unified Dystonia Rating Scale; NRS, numeric rating scale; ADL, activities of daily living.
*The patient was tested in affected side of the extremity.

The timing of the onset of hemidystonia after pontine stroke was also different from that in previous reports. In previous cases, dystonia after pontine stroke did not occur abruptly. In a previous study of 4 patients with dystonia associated with pontomesencephalic lesions, the latency of dystonia onset was 1, 3, 6, and 14 months, respectively [4]. Similarly, in a case recently reported in 2020, hemidystonia occurred on the 11th day of onset [2]. In this case report, dystonia was observed within a few hours after stroke onset. To our knowledge, there has been only one case report in which involuntary movements appeared within a few hours after pontine infarction [3].

Treatment options for dystonia include physical and occupational therapy, oral medications, intramuscular injection of botulinum toxins, non-invasive brain stimulation such as repetitive transcranial magnetic stimulation and transcranial direct current stimulation, and neurosurgical interventions [10,11,13-16]. Among these options, we used the oral medications clonazepam and benztropine and performed intensive comprehensive rehabilitation. Clonazepam is a highly potent benzodiazepine that exerts pronounced antidystonic effects [10]. The potential link between alterations in γ-aminobutyric acid type A (GABA A) receptor subunits and GABAergic disinhibition may contribute to the abnormal synaptic plasticity underlying dystonia [17]. Clonazepam acts as a GABA A receptor agonist in a highly potent, long-acting manner, which makes the GABA receptor more responsive and sensitive to the amount of GABA present [18]. Although it is not as common as clonazepam, benztropine is a treatment option for dystonia. The anticholinergic effect of benztropine is believed to be mediated through blockage of muscarinic acetylcholine receptors, as well as inhibition of dopamine reuptake, thus restoring the imbalance between acetylcholine and dopamine, which are both involved in the pathophysiology of dystonia [9,10,19].

Meanwhile, there have been few reports on the effectiveness of intensive rehabilitation treatment in patients with dystonia. Our patient underwent intensive comprehensive rehabilitation treatment including sensory stimulation, proprioceptive control, balance training, task-specific fine motor training, and ADL training, while simultaneously receiving medical treatment. Similarly, Umar et al. [13] reported that task-specific training improved upper limb function in patients with post-stroke focal dystonia. Specialized rehabilitation techniques such as sensorimotor retraining might potentially target pathophysiological processes causing dystonia, and these techniques have been reported to be beneficial [20]. Attempts to normalize muscle activity to restore voluntary control using biofeedback, vibration, or electrical stimulation have also been presented [20]. During the rehabilitation programs concomitant with medication administration, the severity of dystonia and functional levels improved gradually. After 3 months, the patient was able to walk independently, go up and down several steps, and perform ADLs with minimal assistance. This clinical course is different from previous cases in which hemidystonia frequently progressed to severity in the initial months or years [3]. Early intensive comprehensive rehabilitation programs combined with proper medication administration would yield a gradual recovery. More long-term follow-up observations are necessary to check the extent of the patient's recovery, and further studies are needed to establish therapeutic strategies for dystonia after pontine lesions.

DISCUSSION

This is a rare case of contralateral hemidystonia accompanied by loss of proprioception as a result of pontine hemorrhage. Administration of clonazepam and benztropine combined with intensive rehabilitation programs showed beneficial effects on dystonia and functional improvement. Recovery was gradual during the rehabilitation period. More long-term follow-up observations are needed, and future studies to establish therapeutic strategies for hemidystonia after pontine lesions are warranted.

REFERENCES

1. Albanese A, Bhatia K, Bressman SB, Delong MR, Fahn S, Fung VS, Hallett M, Jankovic J, Jinnah HA, Klein C, Lang AE, Mink JW, Teller JK. Phenomenology and classification of dystonia: a consensus update. Mov Disord 2013;28:863-873.
PUBMED | CROSSREF
2. Park CW, Chung SJ, Sohn YH, Lee PH. A case of abnormal postures in the left extremities after pontine hemorrhage: dystonia or pseudodystonia? J Mov Disord 2020;13:62-65.
PUBMED | CROSSREF
3. Tan EK, Chan LL, Auchus AP. Hemidystonia precipitated by acute pontine infarct. J Neurol Sci 2005;234:109-111.
PUBMED | CROSSREF
4. Loher TJ, Krauss JK. Dystonia associated with pontomesencephalic lesions. Mov Disord 2009;24:157-167.
PUBMED | CROSSREF
5. Balint B, Mencacci NE, Valente EM, Pisani A, Rothwell J, Jankovic J, Vidailhet M, Bhatia KP. Dystonia. Nat Rev Dis Primers 2018;4:25.
PUBMED | CROSSREF
6. Jinnah HA, Neychev V, Hess EJ. The anatomical basis for dystonia: the motor network model. Tremor Other Hyperkinet Mov (N Y) 2017;7:506.
PUBMED | CROSSREF

7. Kim HJ, Cho YJ, Cho JY, Hong KS, Jeon BS. Choreodystonia in a patient with hypertrophic olivary degeneration after pontine tegmental hemorrhage. Mov Disord 2008;23:920-922.
PUBMED | CROSSREF
8. Salerno SM, Kurlan R, Joy SE, Shoulson I. Dystonia in central pontine myelinolysis without evidence of extrapontine myelinolysis. J Neurol Neurosurg Psychiatry 1993;56:1221-1223.
PUBMED | CROSSREF
9. Goff DC, Arana GW, Greenblatt DJ, Dupont R, Ornsteen M, Harmatz JS, Shader RI. The effect of benztropine on haloperidol-induced dystonia, clinical efficacy and pharmacokinetics: a prospective, double-blind trial. J Clin Psychopharmacol 1991;11:106-112.
PUBMED
10. Cloud LJ, Jinnah HA. Treatment strategies for dystonia. Expert Opin Pharmacother 2010;11:5-15.
PUBMED | CROSSREF
11. Chuang C, Fahn S, Frucht SJ. The natural history and treatment of acquired hemidystonia: report of 33 cases and review of the literature. J Neurol Neurosurg Psychiatry 2002;72:59-67.
PUBMED | CROSSREF
12. Comella CL, Leurgans S, Wuu J, Stebbins GT, Chmura TThe Dystonia Study Group. Rating scales for dystonia: a multicenter assessment. Mov Disord 2003;18:303-312.
PUBMED | CROSSREF
13. Umar M, Masood T, Badshah M. Effect of botulinum toxin A & task-specific training on upper limb function in post-stroke focal dystonia. J Pak Med Assoc 2018;68:526-531.
PUBMED
14. Borich M, Arora S, Kimberley TJ. Lasting effects of repeated rTMS application in focal hand dystonia. Restor Neurol Neurosci 2009;27:55-65.
PUBMED | CROSSREF
15. Bonouvrié LA, Becher JG, Vles JS, Boeschoten K, Soudant D, de Groot V, van Ouwerkerk WJ, Strijers R, Foncke E, Geytenbeek J, van de Ven PM, Teernstra O, Vermeulen RJ. Intrathecal baclofen treatment in dystonic cerebral palsy: a randomized clinical trial: the IDYS trial. BMC Pediatr 2013;13:175.
PUBMED | CROSSREF
16. Tassorelli C, Mancini F, Balloni L, Pacchetti C, Sandrini G, Nappi G, Martignoni E. Botulinum toxin and neuromotor rehabilitation: an integrated approach to idiopathic cervical dystonia. Mov Disord 2006;21:2240-2243.
PUBMED | CROSSREF
17. Berman BD, Pollard RT, Shelton E, Karki R, Smith-Jones PM, Miao Y. $GABA_A$ receptor availability changes underlie symptoms in isolated cervical dystonia. Front Neurol 2018;9:188.
PUBMED | CROSSREF
18. Griffin CE 3rd, Kaye AM, Bueno FR, Kaye AD. Benzodiazepine pharmacology and central nervous system-mediated effects. Ochsner J 2013;13:214-223.
PUBMED
19. Pisani A, Bernardi G, Ding J, Surmeier DJ. Re-emergence of striatal cholinergic interneurons in movement disorders. Trends Neurosci 2007;30:545-553.
PUBMED | CROSSREF
20. Lizarraga KJ, Al-Shorafat D, Fox S. Update on current and emerging therapies for dystonia. Neurodegener Dis Manag 2019;9:135-147.
PUBMED | CROSSREF