A Case of Abnormal Postures in the Left Extremities after Pontine Hemorrhage: Dystonia or Pseudodystonia?

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Dystonia is a type of involuntary movement characterized by the patterned and repetitive contraction of agonist and antagonist muscles. It can be classified into primary dystonia, dystonia-plus syndromes, heredodegenerative dystonia, and symptomatic dystonia according to its etiology. In particular, focal lesions in areas such as the basal ganglia, thalamus, cerebellum and brainstem have been reported to cause symptomatic dystonia. However, it is difficult to determine the pathoanatomical correlates of dystonia because of its complex pathophysiology, and most cases are associated with basal ganglia lesions. Moreover, it is challenging to distinguish between dystonia and pseudodystonia when abnormal postures are accompanied by other neurological findings in the affected body part. Here, we report a case of abnormal postures with decreased proprioception in the left extremities after right dorsal pontine hemorrhage.

Key Words: Brainstem; Dystonia; Hemorrhage; Pons; Pseudodystonia.
ing movements. When he closed both eyes, the abnormal posture of the left hand was aggravated (Supplementary Video 1 in the online-only Data Supplement). There was no sensory tricks. Brain magnetic resonance imaging showed low signal intensity in the right dorsal pons, extending to the lower midbrain in susceptibility-weighted images. There was also a hyperintense lesion in the left pons on the T2-weighted image, indicating an old ischemic change (Figure 1A). There was no focal lesion in the cortical gray matter, basal ganglia, internal capsule, thalamus, or cerebellum. The electromyography recording showed coactivation of the left flexor carpi radialis and extensor carpi radialis while stretching out his left arm (Figure 1B) as well as coactiva-

![Figure 1](image_url)

**Figure 1.** (A) Axial susceptibility-weighted, T1-weighted, and T2-weighted imaging 1 month later showed a hemorrhagic lesion in the right pons and a chronic ischemic lesion in the left pons. (B) The electromyography (EMG) recording showed coactivation of the left flexor carpi radialis and left extensor carpi radialis while stretching out his left arm. (C) The EMG recording showed coactivation of the left tibialis anterior and left gastrocnemius while stretching out his left leg.
tion of the left tibialis anterior and gastrocnemius while stretching out his left leg (Figure 1C). After the introduction of anticholinergics and clonazepam, his abnormal postures partially improved.

Dystonia has been traditionally regarded as a disorder of the basal ganglia, but recent studies have suggested that it is a network disorder involving the cerebello-thalamo-cortical circuit. In addition, some reports have shown that the brainstem can be associated with the pathogenesis of dystonia; however, structural lesions are not limited to the brainstem in most cases, and hemidystonia has rarely been reported as a consequence of focal brainstem lesions.6-8

To date, there have been three reports on five cases of pontine lesions with associated hemidystonia (Table 1).6-8 Although the exact mechanisms underlying dystonia after pontine lesions remain uncertain, the authors proposed several explanations, including disrupted sensory afferent input to the striatum or thalamus, loss of pallidal inputs to the pedunculopontine fibers, and damaged dentate-rubro-olivary pathways. In this case, some possible structures might have been involved in the pontine hemorrhagic lesion, including the medial lemniscus, central tegmental tract, and pedunculopontine nucleus.

Our patient showed severely decreased proprioception in the left extremities, which was not observed or checked in previous reports.6-8 Abnormal proprioceptive information may not be integrated correctly into the sensorimotor system, which is linked to defective inhibition within the somatosensory cortex, causing abnormal involuntary muscle contraction.9,10 However, it is still a challenging issue that patients with abnormal postures and a loss of proprioception can be diagnosed with true dystonia or pseudodystonia. Pseudodystonia represents abnormal postures or repetitive movements in which clinical, imaging, laboratory, or electrophysiological findings are not compatible with true dystonia.7 The presence of associated neurological findings (e.g., sensory loss) in the affected body part is the key distinguishing feature for making a diagnosis of pseudodystonia. Moreover, in this case, eye closure accentuated the abnormal postures in the left hand, suggesting that this abnormal posture might be compensated by the visual system, unlike what occurs in true dystonia. However, true dystonia may also worsen with eye closure, presumably due to distraction. Moreover, the electromyography recording showed simultaneous contraction of agonist and antagonist muscles in the left arm and leg, which is compatible with dystonia.

In conclusion, this is a rare case of hemidystonia associated with a pontine structural lesion, which might be ascribed to a loss of proprioceptive sensation.

| Table 1. Characteristics of patients with limb dystonia and pontine lesions: literature review |
|---------------------------------|---------------------------------|-------------------------------|-----------------------------|------------------|-----------------|-----------------|-----------------|
| Study                          | Age/sex | Type of lesion                     | Site of lesion | Phenomenology                                                                 | Sensory symptoms                                      | Other clinical symptoms                                 | Literature review |
| Tan et al.6                     | 43/M     | Acute infarction                   | Left paramedian and ventral pons (old infarction in the left caudate nucleus and putamen) | Right hemidystonia, tonic contractions of the right facial musculature | Normal pinprick, temperature, and proprioception             | Normal sensory exam                                      | Pontine lesion    |
| Kim et al.7                    | 75/M     | Spontaneous hemorrhage             | Left pontine tegmentum extended rostrally to the lower midbrain; an enlarged left anterior inferior medulla with hypertrophic olivary degeneration | Left hemidystonia with athetoid movement with rest and postural tremor, cervical dystonia with tremor, right arm bradykinesia | Normal sensory exam                                      | Left-sided hemihypesthesia                               | Pontine lesion    |
| Loher and Krauss               | 31/M     | Spontaneous hemorrhage             | Right lateral and paramedian tegmentum extending from the pontomesencephalic junction up to the area of the red nucleus | Left hemidystonia, cervical dystonia with tremor | Normal sensory exam                                      | Right-sided hemihypesthesia                               | Pontine lesion    |
| 42/F                           |          | Posttraumatic hemorrhage           | Left lateral pontine tegmentum extending from the posterior midbrain to the midpons of the posterior midnucleus  | Right hemidystonia, with tremor, tics, right arm bradykinesia | Normal sensory exam                                      | Right-sided hemihypesthesia                               | Pontine lesion    |

4/M Diffuse axonal injury after head trauma

Left lateral pontomesencephalic tegmentum, extended from the posterior midpons to the midpons of the posterior midnucleus.
Supplementary Video Legend

Video 1. Abnormal postures of the left hand (patterned flexion of the wrist and hyperextension of all five fingers) and left foot (patterned dorsiflexion of the ankle and flexion of the toes).

Supplementary Materials

The online-only Data Supplement is available with this article at https://doi.org/10.14802/jmd.19074.

Conflicts of Interest

The authors have no financial conflicts of interest.

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Ethical Statement

The work has been carried out in accordance with the Code of Ethics of the World Medical Association (Declaration of Helsinki) and its later amendments or comparable ethical standards for experiments involving humans. Informed consent was obtained from all the patients included in the study.

Author Contributions

Conceptualization: Seok Jong Chung and Phil Hyu Lee. Data curation: Chan Wook Park. Formal analysis: Chan Wook Park. Funding acquisition: Seok Jong Chung and Phil Hyu Lee. Investigation: Chan Wook Park. Methodology: Chan Wook Park. Supervision: Young H. Sohn. Validation: Seok Jong Chung. Visualization: Chan Wook Park. Writing—original draft: Chan Wook Park. Writing—review & editing: Seok Jong Chung and Phil Hyu Lee.

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