Focal Dystonia with Primary Medullary Hemorrhage

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We report a patient with hand dystonia related with primary medullary hemorrhage. A 69-year-old man presented with dysarthria, right facial palsy, limb paresthesia, and limb ataxia of acute onset. Neurologic examination revealed abnormal tonic posturing of the right hand and wrist that persisted while at rest. Brain MRI showed acute hemorrhage at the right posteromedial medulla oblongata extending to the upper cervical cord. Ipsilateral limb dystonia might result from the interruption of the reticulospinal tract, sensory or olivocerebellar pathway.

Key Words: Focal dystonia, Medullary hemorrhage, Reticulospinal tract

Dystonia is characterized by the abnormal involuntary sustained muscle contractions that often result in twisted and unnatural postures. It is usually caused by the diseases of basal ganglia or thalamus disrupting the cortical–striatal–thalamic–cortical motor loop. However, a few cases with limb dystonia were reported in the brainstem or spinal cord lesions that might result from the interruption of the descending inhibitory reticulospinal tract. We report an unusual case with focal limb dystonia secondary to medullary hemorrhage.

Case

A 69-year-old man was admitted due to sudden dizziness, speech change and gait disturbance that developed one day before admission. He was a nonsmoker and previously in good health without hypertension, diabetes mellitus, hyperlipidemia and cardiovascular disease. Medical and family history was unremarkable except the history of benign prostate hypertrophy. On the admission, blood pressure was 120/80 mmHg and heart rate was 62/min with a regular rhythm. On the neurologic examination, he was conscious and well oriented. He showed dystarthis. Gag reflex, palatal movement and swallowing were intact. There was a right central type facial palsy with bilateral facial senses being symmetric and intact. Extraocular movement was normal without nystagmus. Proprioceptive and vibration senses were diminished in the right limbs with paresthesia. The severity of sensory impairment was not different between right upper and lower extremities. No abnormalities in touch, pain, and temperature perceptions were detected in four extremities. He showed a right cerebellar dysfunction during a finger–to–nose test and a heel–to–shin test with gait ataxia. Motor power and muscle tone were normal in four extremities. Tendon reflexes were symmetric and nonactive, and Babinski sign was absent. Abnormal sustained tonic posturing of the
right hand and wrist was detected at a fully relaxed state. That was characterized by simultaneous tonic flexion of the wrist, metacarpophalangeal joints, and tonic extension of interphalangeal joints of all fingers with thumb adduction. The degree of metacarpophalangeal flexion was different between lateral (2nd and 3rd) and medial (4th and 5th) fingers, where it was more prominent in medial ones. General contour was similar to obstetric hand associated with hypocalcemia. Dystonic posture was strikingly aggravated by voluntary movements such as writing and holding something. It subsided during sleep. No involuntary movement was detected in other body parts.

Routine laboratory tests including serum electrolyte, thyroid hormones, calcium, and magnesium level were normal. Brain MRI on the 2nd hospital day demonstrated a lesion consistent with acute hemorrhage at the right posteromedial medulla oblongata extending from the middle portion of medulla to the upper cervical cord in the rostrocaudal direction (Fig. 1). MR and conventional cerebral angiography failed to demonstrate any vascular abnormalities causing medullary hemorrhage. During hospitalization, he was treated with gabapentin up to 900 mg/day and amitriptyline 20 mg/day for releasing paresthesias that had no effect on dystonia. After 18 days of admission, dystonia had gradually improved at the time of discharge. Medication was not changed during follow-up. Three months later, the right hand dystonia nearly completely subsided, and the paresthesia in the right limbs gradually improved but still persisted.

Discussion

This patient developed focal limb dystonia of acute onset in association with an ipsilateral medulla oblongata lesion. As far as we know, this case is the first MRI demonstration of a medullary lesion with ipsilateral focal dystonia in Korea. Dystonia in this patient is sustained, different from the paroxysmal type reported in three previous cases related to medullary lesions. It has been proposed that limb dystonia in the brainstem or spinal cord lesions might result from the disruption of the descending inhibitory supraspinal pathways in reticulospinal tracts. The reticulospinal system is involved in the maintenance of posture and in the modulation of muscle tone. Pontine reticulospinal fibers tend to mediate excitatory effects, and medullary reticulospinal fibers usually convey inhibitory effects. In this case, small discrete hematoma in the dorsal part of the paramedian medulla and upper cervical cord disrupted the medullary locomotor center and ipsilaterally descending reticulospinal system that might result in the focal dystonia. Ipsilateral presentation of dystonia supports the notion that direct actions of reticulospinal fibers are much more potent on ipsilateral spinal motor neurons at least in the brainstem level, although reticulospinal tracts are known to act bilaterally through commissural neurons. The study of the medullary reticular formation regarding inhibitory connections with forelimb and hindlimb motor neurons revealed non-specific homogeneously distributed somatotopographic representation in the nucleus reticularis gigantocellularis. So the reason of focal dystonia limited to the hand without involvement of other body parts is uncertain. In this respect, other plausible mechanisms might work to evoke dystonia. Because our patient showed proproceptive sensory disturbances, the impairment of sensorimotor integration might play a role. Abnormal sensory afferent input may interfere with motor program execution in the cortical motor areas, which may cause dystonia. Otherwise the dysfunction or disruption of inferior olivary nucleus or climbing fiber might contribute to the development of dystonia because the lesion was close to the dorsal surface of the right inferior olivary complex, and there is some evidence that the damage of cerebellum or its associated pathways including olivocerebellar and dentate—rubro—thalamic pathways.

Medullary hemorrhage is usually associated with hypertension, small vascular malformations, brain tumor, drugs such as sympathomimetics, anticoagulants or fibrinolytic agents, and vasculitis. Although the cause of hemorrhage could not be defined, the most
probable etiology is a cryptic vascular malformation in this case.

We report the case of ipsilateral focal dystonia resulted from medullary hemorrhage, and the dysfunction of the medullary reticulospinal tract, the sensorimotor integration or olivocerebellar pathway could play a possible pathophysiologic role.

REFERENCES