



Letter to the Editor

Multifocal dystonia as a manifestation of acute midbrain infarction

Keywords:

Dystonia
Midbrain
Stroke

Dear Sir,

The basal ganglia-motor loop is considered a neural substrate related to dystonia [1]. More recently, the cerebellar-motor loop has been implicated as a key anatomical region associated with dystonia [2]. Case studies have shown secondary dystonia in patients with lesions in the cerebellum or the cerebellar-motor connections [3,4]. It is unclear whether the cerebellum is the origin of dystonia or connects the cerebellar and basal ganglia loops [5]. This study describes a patient with multifocal dystonia combined with ataxia in an acute midbrain infarction.

A 66-year-old man visited the Neurology Department due to gait disturbance and difficulty of writing for 3 weeks. He also had dysphagia and dysarthria. He had a 30 year history of hypertension. He had no family history of neurological disorders, including tremor or dystonia. Twelve years earlier, he experienced an ischemic stroke in the left basal ganglia, for which he underwent carotid angioplasty in the left proximal ICA. Since then, he has been taking an antiplatelet agent, a statin, and an antihypertensive medication regularly. He did not have any neurological deficits after the first stroke, including motor weakness and dystonia. Neurological examination showed dystonia in both hands and feet (Video-Still 1). Dystonia of the right hand was aggravated when writing, accompanied by mirror dystonia of the left hand (Video-Still 2). The tongue exhibited involuntary contractions while the patient was resting. His speech was dysarthric. The score on the Burke-Fahn-Marsden dystonia movement scale was 35 (range, 0–120). Cerebellar function tests showed ataxia of both arms and legs (Video-Still 3). The patient exhibited gait disturbance with wide base, and he swayed in both directions on tandem walking. Diffusion-weighted MRI of the brain revealed an acute infarction in the right midbrain (Fig. 1A). T2-weighted MRI three weeks after symptom onset showed high signal intensity in the same area (Fig. 1B). The lesion was located in the paramedian caudal midbrain, which contains the superior cerebellar peduncle, the decussated superior cerebellar peduncle, and the caudal portion of the ventral tegmental area including the rubrospinal fiber. Old ischemic lesions were observed in the left putamen (Fig. 1C), which were thought to be related to the first clinical stroke, and the right globus pallidus (Fig. 1D). Treatment with clonazepam and trihexyphenidyl did not improve the dystonia of the limbs and tongue.

The patient described here showed multifocal dystonia, involving his arms, legs and tongue, without involvement of the trunk. The patient

also presented with limb ataxia and cerebellar gait disturbance. The lesion observed on brain MRI suggested that the neurological manifestations in this patient were caused by the acute midbrain infarction. Therefore, according to the consensus classification system, dystonia in this patient could be classified as late adult onset, multifocal acquired dystonia of vascular origin [6]. Brain MRI also showed evidence of an old ischemic stroke in the basal ganglia. The basal ganglia lesions can cause dystonia and other movement disorders. However, the temporal relationship between dystonia and MRI findings suggested that dystonia in this patient was caused by the acute midbrain infarction, not by old lesions in the basal ganglia.

Decussated fibers of the superior cerebellar peduncle constitute the bilateral output pathway from the cerebellum to the thalamus. Therefore, caudal paramedian midbrain infarctions usually cause bilateral cerebellar dysfunctions, including dysarthria, bilateral limb ataxia, and gait and truncal ataxia [7]. Although showing typical neurological deficits, this patient also presented with dystonia of both limbs, which is considered highly unusual. Acquired dystonia after midbrain infarction has rarely been reported [3,4]. Moreover, multifocal dystonia involving both arms and legs has not been reported as a manifestation of midbrain infarctions. Most patients with acquired dystonia after midbrain lesion showed hemidystonia contralateral to the lesion side or cervical dystonia [3,4]. Lesions in patients with hemi-dystonia due to midbrain lesions were found to involve dopaminergic dysfunction, as shown by dopamine transporter (DAT) image [8]. The midbrain lesion in our patient did not directly extend to the anatomical region related to the nigrostriatal dopaminergic system. Although the lesion extended into the ventral tegmental area, this likely did not directly cause acute dystonia because this area has been associated functionally with emotion, cognition and reward systems [9].

The patho-mechanism of multifocal limb dystonia in this patient is unclear. One possibility is that the basal ganglia lesions caused by the previous ischemic stroke may have led to the development of dystonia following the midbrain infarction. The basal-ganglia and cerebellar-motor loops have long been considered distinct control circuits and to be entirely separated anatomically and functionally [10]. More recently, however, the cerebellum and basal ganglia have been regarded as communicating with each other and as linked within a functional network [2]. Therefore, they can compensate for each other in patients with structural and functional disorders. In this patient, the cerebellar-motor loop might have compensated for the dysfunction in the basal ganglia motor circuit after the previous basal ganglia infarction. Subsequently, acute destruction of the compensatory mechanism for the basal ganglia motor loop from the cerebellar circuit may have caused acute dystonia after midbrain infarction.

Post-stroke movement disorders usually develop several months to years after the acute stage of stroke [11]. Reorganization of basal-ganglia motor circuits or generation of hyper-excitability areas after the acute stage of insult may cause delayed hyperkinetic movement disorders [11]. However, our patient showed multifocal dystonia as a manifestation of acute stroke, suggesting that dystonia in this patient was not

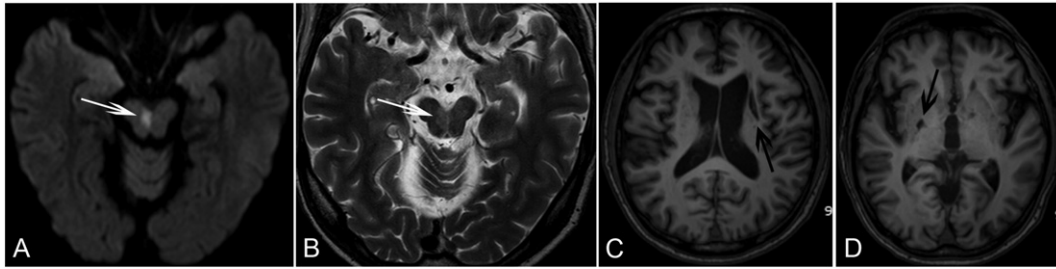


Fig. 1. Initial diffusion-weighted MRI (A) and follow-up T2-weighted MRI (B) showing cerebral infarction in the caudal paramedian midbrain. T1 weighted MRI showed an old ischemic lesion in the left putamen, related to the first clinical stroke (C), and in the right globus pallidus (D).

caused by neuronal reorganization mechanisms. Rather, acute dystonia may have been caused by acute insult to the cerebello-thalamo-cortical tract itself, supporting a close relationship between the cerebellar and basal ganglia motor loops. Therefore, dystonia should be considered not only as a disorder of the basal ganglia, but also as a circuit disorder involving the cortico-ponto-cerebello-thalamo-cortical and cortico-basal ganglia-thalamo-cortical pathways. These linkages were shown directly in animals and indirectly by clinical neuroimaging and neurophysiology [5]. Acute multifocal dystonia as a neurological manifestation of cerebral infarction involving the cerebellar output circuit in this patient may be an important evidence of a functional cerebellar–basal ganglia connection.

Supplementary data to this article can be found online at <http://dx.doi.org/10.1016/j.jns.2015.06.068>.

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16 April 2015