

Short communication

Hemidystonia precipitated by acute pontine infarct

E.K. Tan^{a,c,d,*}, L.L. Chan^b, A.P. Auchus^{a,c}

^aDepartment of Neurology, Singapore General Hospital, Singapore

^bDepartment of Diagnostic Radiology, Singapore General Hospital, Singapore

^cNational Neuroscience Institute, Singapore

^dSingHealth Research, Singapore

Received 30 December 2004; received in revised form 24 February 2005; accepted 23 March 2005

Available online 2 June 2005

Abstract

Hemidystonia is frequently due to an underlying structural lesion in the basal ganglia and thalamus. It has been suggested that a preserved corticospinal tract may be required for hemidystonia to manifest. We provide the first report of a patient who presented with rapid-onset hemidystonia precipitated by an acute pontine infarct demonstrated on diffusion-weighted magnetic resonance imaging. Acute dysregulation of pallidal efferents to the pedunculopontine and/or pontine afferents to the thalamus may precipitate hemidystonia.

© 2005 Elsevier B.V. All rights reserved.

Keywords: Hemidystonia; Acute; Pontine infarct

1. Introduction

Dystonia is characterized by persistent and prolonged co-contraction of both the agonist and antagonist muscles resulting in abnormal posturing (over flexion, over extension, rotation) at rest and/or duration of action [1]. Both hyperkinetic and hypokinetic movement disorders (such as dystonia, tremor, myoclonus and parkinsonism) are well-recognized consequences of basal ganglia lesions [1,2]. Blepharospasm and other cranial dystonias may be associated with brainstem abnormalities [3]. Hemidystonia is frequently due to an underlying structural lesion in the basal ganglia and thalamus [4–8]. Here, we provide the first report of a patient who presented with hemidystonia precipitated by an acute pontine infarct, and discuss its possible pathophysiologic mechanisms.

2. Case report

The patient, a 43-year-old man with diabetes mellitus and hyperlipidemia presented with sudden onset of weakness and spasms of his right-sided limbs. The patient complained of sudden onset of right-sided weakness, dropping things easily and having difficulty opening the door and holding a cup. He was perfectly well prior to this. Within a few hours, there was pain and spasm in his right-sided limbs. He described this as a cramp-like feeling resulting in abnormal bending of his right elbow and fisting movements of his hand. In addition his right foot would turn inwards and his toes would curl. These involuntary movements were aggravated by actions such as walking and writing, relieved by rest and sleep. His right-sided limbs were weaker due to the pain and spasms. He did not complain of headache, vomiting, or preceding head injury. There was no previous or family history of similar movement disorders, and no exposure to neuroleptic, anti-emetic or anti-depressive medications. Due to the suddenness of his symptoms, he sought help at the hospital.

Neurologic examination revealed normal mentation, and orientation to people, place and time. His pupils were equal

* Corresponding author. Department of Neurology, Singapore General Hospital, Outram Road, Singapore 169608, Singapore. Tel.: +65 6326 5003; fax: +65 6220 3321.

E-mail address: gnrtek@sgh.com.sg (E.K. Tan).

bilaterally and extraocular movements were full. He was dysarthric and had evidence of mild right facial upper motor neuron palsy. In addition, tonic contractions of the right facial muscles were observed. Dystonic posturing of the right-sided limbs was present and characterized by flexion at the elbow, pronation of the forearm, flexion at the wrist with ulnar deviation and flexion deformity of the fingers. Equinovarus deformity of his right foot with clawing of all his toes with extensor spasms of his knees limited his mobility. The resting tone was increased on the right and the power was of grade 4/5 in both the upper and lower limbs and 5/5 on the left. There was no loss of sensation to pin-pick, temperature and proprioception. The reflexes were slightly brisk on the right with extensor plantar response. The right hemidystonia worsened with walking and writing. There was no evidence of dystonic spasms of the neck.

Blood investigations revealed normal full blood counts, electrolytes, renal and liver function. Connective tissue (antinuclear antibody, anti-DS DNA) and hypercoagulability screen (protein C, S and antithrombin III, lupus anticoagulant) were not remarkable. His serum ceruloplasmin level was normal. Diffusion-weighted magnetic resonance imaging demonstrated hyperintensity in the left paramedian and ventral pons (involving the corticospinal tract). There was T2 hyperintensity in the left caudate nucleus and putamen, without diffusion abnormality, consistent with old infarcts (Fig. 1). There was also mild T1-

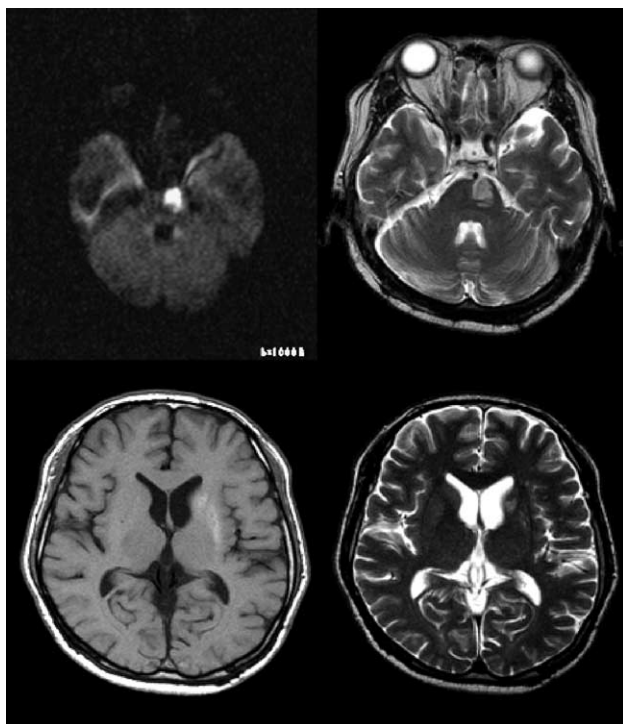


Fig. 1. An acute infarct in the left hemipons demonstrated on diffusion-weighted image (trace, $b=1000$) (left upper row) and T2-weighted image (right upper row). T1-weighted and T2-weighted axial images through the basal ganglia demonstrating chronic lacunar infarcts in the left caudate head and putamen, with mild T1 and T2 hyperintensity (lower row).

weighted hyperintensity in the left caudate and putamen, suggesting underlying dystrophic calcification within the old infarcts. Moderate stenosis at the left M1–M2 junction was observed on MR angiography. His dystonia improved with clonazepam, and an antiplatelet agent was prescribed for his strokes. At 1-year follow-up, he was able to ambulate and independent in his activities of daily living. Mild dystonic posturing with flexion at the elbow and wrist and clawing of toes was still present.

3. Discussion

Lesions in the putamen, globus pallidus, caudate, thalamus and subthalamus predispose to various forms of movement disorders. Cranial nerve palsies, sensorimotor hemiparesis, and rarely cranial dystonias are manifestations of brainstem lesions. Patients with cervical dystonia and blepharospasm secondary to brainstem lesions do not develop widespread dystonia. In a recent review of the literature involving 190 cases of hemidystonia, the most common etiologies were stroke, trauma and perinatal injury, with a mean latency period from insult of about 3 years [8]. The vast majority of the lesions involved the basal ganglia, with the putamen the most common site.

To our knowledge, abrupt onset hemidystonia following an acute pontine infarct observed in our patient is unusual, and has not been highlighted. The suddenness of the patient's right-sided weakness and dystonia, supported by imaging evidence of hyperacute pontine infarct on diffusion-weighted MR strongly suggests that the pontine infarct likely plays some part in unmasking the dystonia in our patient who arguably may already be at-risk of dystonia from the chronic basal ganglia lesion. Furthermore, the patient did not have any previous history or risk factors of dystonia. Hemidystonia frequently progresses in severity in the initial months or years, followed by eventual stabilization. Our patient's symptoms were maximal immediately following the pontine stroke but stabilized and improved after 1 year. Dystonia may sometimes be confused with spasticity. However, the observation of worsening of the limb spasms during specific actions such as writing, holding a cup and walking, and improvement with rest and sleep were suggestive dystonic features.

The thalamus receives afferent tracts from the striatum and sensory afferents from the trigeminothalamic tract. Efferents from the thalamus branch out to the motor cortex. In dystonia, there is an increased thalamocortical excitation as a consequence of decreased output from the globus pallidus internus and subthalamic nucleus [2,4]. The altered thalamic output influences the cortical motor planning and executive areas, and there is abnormal regulation of brainstem and spinal cord inhibitory interneuronal mechanisms [9]. In our patient, the left pontine infarct could have precipitated the hemidystonia by disrupting the sensory afferent input to the striatum or thalamus, and/or loss of

pallidal inputs to the pedunculopontine fibres, leading to cortical disinhibition in an already “unbalanced” system from the chronic basal ganglia abnormality.

Dooling has suggested that a preserved corticospinal tract may be required for hemidystonia to manifest [10]. In their series of hemidystonia patients, pathologic involvement was observed in the striatum, striatopallidal fibres, the ventral and lateral thalamic nuclei, but the descending cortical motor pathways were relatively preserved [9]. Marsden and colleagues found no cases of symptomatic hemidystonia due to an isolated brainstem abnormality or in combination with basal ganglia lesions [4]. Our case highlights that corticospinal tract damage from an infarction in the presence of chronic striatal abnormality can result in hemidystonia. The relative role of residual corticospinal function in modulating this manifestation is not clear. It is interesting to note that dystonia has been associated with pontine and extrapontine myelinolysis [11–16]. However, the pathophysiology of progressive and delayed dystonia from an osmotic demyelinating process with ineffective reorganization of neuronal structures is not entirely comparable to those associated with a structural lesion. In conclusion, we illustrate a unique case of rapid-onset hemidystonia associated with an acute pontine infarct, adding a piece of clinical observation that may be useful for future pathophysiologic studies of dystonia.

References

- [1] Jankovic J. Dystonia: medical therapy and botulinum toxin. *Adv Neurol* 2004;94:275–86.
- [2] Sanghera MK, Grossman RG, Kalhorn CG, Hamilton WJ, Ondo WG, Jankovic J. Basal ganglia neuronal discharge in primary and secondary dystonia. *Adv Neurol* 2004;94:29–36.
- [3] Dietrichs E, Heier MS, Nakstad PH. Jaw-opening dystonia presumably caused by a pontine lesion. *Mov Disord* 2000;15(5):1026–8.
- [4] Marsden CD, Obeso JA, Zarranz JJ, Lang AE. The anatomic basis of symptomatic hemidystonia. *Brain* 1985;108:463–83.
- [5] Pettigrew LC, Jankovic J. Hemidystonia: a report of 22 patients and a review of the literature. *J Neurol Neurosurg Psychiatry* 1985;48:650–7.
- [6] Krauss JK, Jankovic J. Hemidystonia secondary to carotid artery gunshot injury. *Child’s Nerv Syst* 1997;13:285–8.
- [7] Friedman DI, Jankovic J, Rolak LA. Arteriovenous malformation presenting as hemidystonia. *Neurology* 1986;36:1590–3.
- [8] 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.
- [9] Berardelli A, Rothwell JC, Hallett M, Thompson PD, Manfredi M, Marsden CD. The pathophysiology of primary dystonia. *Brain* 1998;121(Pt. 7):1195–2212.
- [10] Dooling EC, Adams RD. The pathological anatomy of posthemiplegic athetosis. *Brain* 1975;98:29–48.
- [11] Yoshida Y, Akanuma J, Tochikubo S, Hoshi A, Matsuura Y, Homma M, et al. Slowly progressive dystonia following pontine and extrapontine myelinolysis. *Intern Med* 2000;39:956–60.
- [12] Defebvre L, Rogelet P, Destee A, Verier A. Regressive dystonia and cerebellar ataxia: two unusual symptoms in central pontine myelinolysis. *J Neurol* 1995;242:450–4.
- [13] Wu CL, Lu CS. Delayed-onset dystonia following recovery from central pontine myelinolysis. *J Formos Med Assoc* 1992;91:1013–6.
- [14] Grafton ST, Bahls FH, Bell KR. Acquired focal dystonia following recovery from central pontine myelinolysis. *J Neurol Neurosurg Psychiatry* 1988;51:1354–5.
- [15] Gibb WR, Lees AJ, Marsden CD. Pathological report of four patients presenting with cranial dystonias. *Mov Disord* 1988;3:211–21.
- [16] Seah AB, Chan LL, Wong MC, Tan EK. Evolving spectrum of movement disorders in extrapontine and central pontine myelinolysis. *Parkinsonism Relat Disord* 2002;9:117–9.