

Case reports

Thalamic hand dystonia : an MRI anatomoclinical study

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Abstract

Focal dystonia has been attributed to lesions involving the basal ganglia and/or thalamus. Hand dystonia was studied in a patient with a unilateral thalamic infarction documented by MRI. A 18-year-old girl presented with severe isolated dystonia of the right hand as a sequel of perinatal infarction. MRI scan revealed infarction affecting part of the dorsomedian, lateral posterior, ventral lateral, ventral posterior lateral nuclei, and centromedian-parafascicular nucleus of the contralateral thalamus. The unique MRI anatomoclinical presentation of this case, taken together with the literature data, could provide evidence that a lesion affecting one or several thalamic nuclei, including the centromedian nucleus, can induce hand dystonia.

Key words : Dystonia ; hand ; infarction ; magnetic resonance imaging ; thalamus.

Introduction

Dystonia is defined as a syndrome of sustained involuntary contraction of both agonist and antagonist muscles, frequently causing twisting and repetitive movements, or abnormal postures (Fahn, 1988). Dystonia may develop as a result of a variety of cerebral lesions (structural or metabolic) and involve a diversity of structures including the basal ganglia, thalamus or subthalamic nuclei (Marsden *et al.*, 1985). However, the exact neuroanatomical substrate for dystonia remains to be elucidated. One third of patients with movement disorders associated with lesions of the thalamus and subthalamic area present with contralateral dystonia (Lee and Marsden, 1994). The thalamic structures commonly involved were the posterior (Pettigrew *et al.*, 1985), posterolateral (Lee and Marsden, 1994), and paramedian (Bogousslavsky *et al.*, 1988 ; Lee and Marsden, 1994) nuclei, with segmental dystonia (particularly upper limb) or hemidystonias being the most common mode of clinical presentation (Lee and Marsden, 1994). As evidenced by positron emission tomographic (PET) studies (Ceballos-Baumann *et al.*, 1995), increased

thalamocortical drive and consequently inappropriate overactivity of premotor and motor areas probably forms the pathophysiological basis of dystonia.

We report a case of hand dystonia due to a discrete infarction in the contralateral thalamus. The literature of isolated dystonia (dystonia in the absence of any other type of movement disorder e.g., chorea, ballism, tremor) associated with a lesion confined to the thalamus will be reviewed. To our knowledge, only two cases of hand dystonia in association with a contralateral lesion at this location have been reported.

Case report

A 18-year-old girl presented with right hand dystonia as a sequel of perinatal infarction. She developed the symptoms around the age of one. There was no history of any other neurological disease and she had no risk factors for stroke. Neurological examination showed an alert patient with normal language ability and cognitive functions. Neuro-ophthalmologic examination revealed normal visual acuity, fields, and fundi. The pupils were equal and normally reactive to light and near stimuli. There was no eye deviation in primary position. All types of horizontal and vertical eye movements were preserved. Occasional myoclonic movements were observed at the right side of the face but sparing the right upper limb and other parts of the body. At rest, she had severe dystonic posture of the metacarpophalangeal joints with flexed posture of the fingers and thumb clenching. She had to abduct her thumb passively in order to grasp an object. Attempted movements of the right hand or fingers did not induce abnormal postures of the foot or toes. Fig. 1 shows the right hand after voluntary opening. Posture worsened with action and disappeared during sleep but not at rest. Superficial sensory stimuli such as light touch on the palm did not abolish the dystonic posture. Sensation was normal for all modalities. There was no motor deficit, all tendon reflexes were normal and plantar



FIG. 1. — Dystonic posture of the right hand.

responses were flexor. Cerebellar function and gait were normal.

Blood pressure was 160/80 mm Hg. Cardio-pulmonary examination was normal. There were no cervical bruits. The remainder of the general examination was also normal.

All serum laboratory values (including ceruloplasmin) were within the normal range. Somatosensory evoked potentials (SEP) of upper and lower limbs were normal. MRI (2.00 mm thick slices in the axial and coronal planes and 3.00 mm slices in the sagittal plane) revealed a zone of altered signal intensity, suggesting infarction in the left thalamus (Fig. 2). There were no striatopallidal, cortical, cerebellar or brainstem lesions.

She was unsuccessfully treated with several successive oral medications including haloperidol, clonazepam, valproate, levodopa, baclofen, amitriptyline, procyclidine, gabapentin, and local injections of botulinum toxin.

Transcutaneous electric nerve stimulation (TENS) (TPN 300 ; burst mode 150 ms, 20 Hz for 20 min) of the posterior surface of the right thumb facilitated hand opening and abolished thumb clenching.

Discussion

This patient presented with isolated focal dystonia from a contralateral thalamic infarction. Furthermore, there was no sensory or motor deficit and MRI did not show any evidence of involvement of basal ganglia or subthalamic structures. Topographically, the infarct was localized with the use of corresponding anatomical sections as described by Schaltenbrand and Wahren (1977) and by MRI neuroanatomy (Jackson and Duncan, 1996). The nuclei affected corresponded to the rostral part of the centromedian-parafascicular nucleus, and part of the lateral posterior, dorsomedian, ventral lateral and ventral posterior lateral nuclei of the contralateral thalamus (Fig. 3). Although the initial extent of the lesion might be underestimated by shrinkage of the lesion, the clinical history and features were not suggestive for involvement of other thalamic structures.

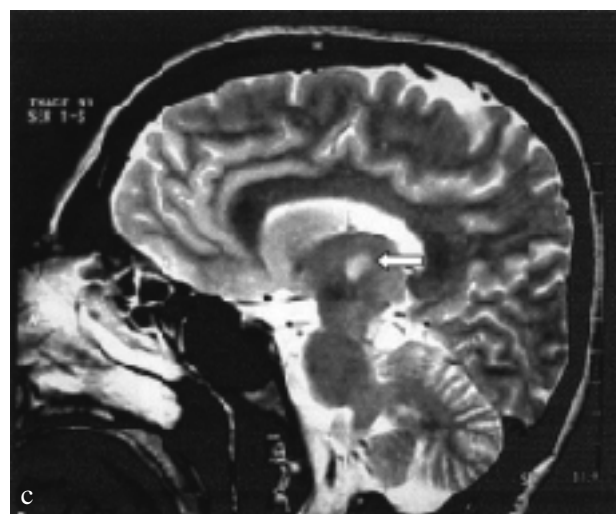
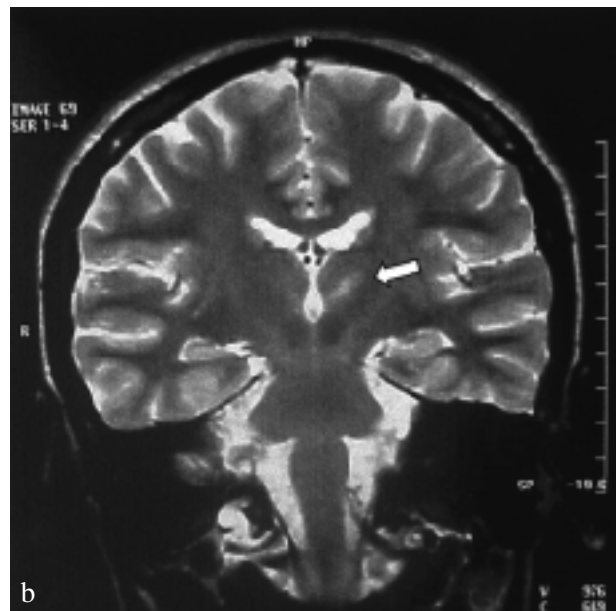
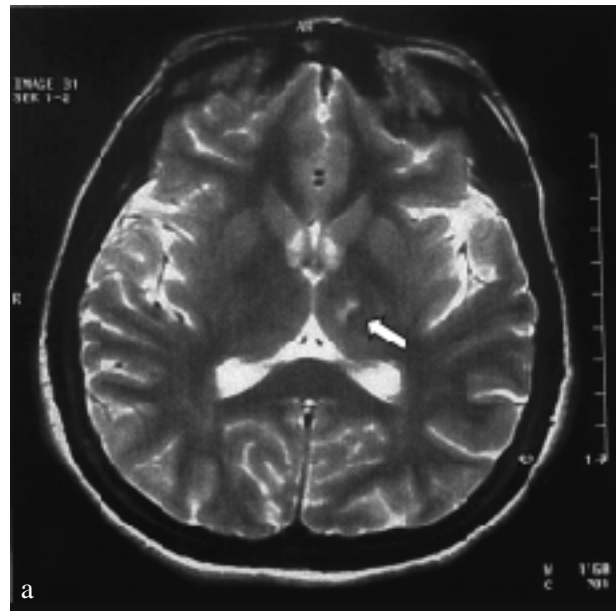


FIG. 2. — T₂-weighted MRI scan : (A) axial, (B) coronal and (C) sagittal views showing infarction (arrow) in the left thalamus.

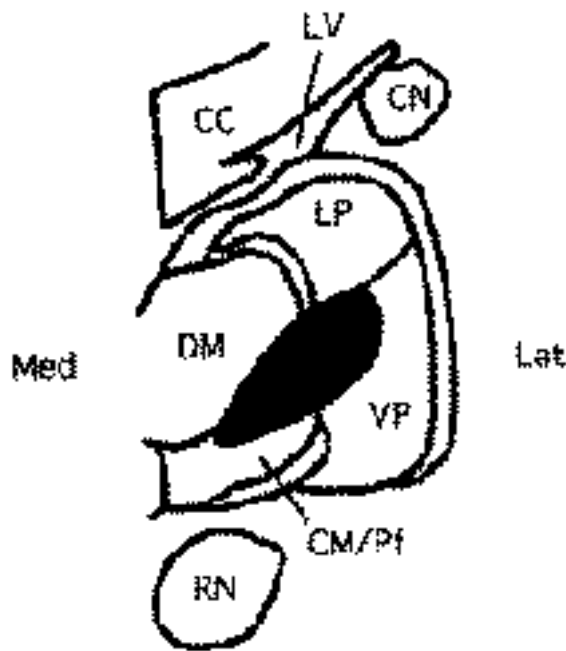


FIG. 3. — Schematic representation (coronal section) of the ischemic lesion (black shaded area) at its most posterior level in the left thalamus. CC, corpus callosum ; CM/Pf, centromedian-parafascicular nucleus ; CN, caudate nucleus ; DM, dorsomedian nucleus ; Lat, lateral ; LV, lateral ventricle ; LP, lateral posterior nucleus ; Med, medial ; RN, red nucleus ; and VP, ventral posterior nuclei.

In their literature review, Lee and Marsden (1994) analyzed the reports of 62 cases with movement disorders associated with a focal thalamic and/or subthalamic lesion. Out of 33 patients with a lesion confined to the thalamus, 16 patients presented with dystonia (10 with hemidystonia, 5 with hand dystonia, and 1 with segmental dystonia), topographically corresponding (when recorded) with a lesion in either the posterior, posterolateral or paramedian part of the contralateral thalamus. Karşıdağ *et al.* (1998) described 9 patients with poststroke focal dystonia. In their series, all patients presented with hand dystonia in association with either hemiparesis, hemihyperesthesia or other movement disorders including hemichorea, action tremor, parkinsonism or hemiballism. This probably confirmed that these patients had in addition to contralateral thalamic damage also lesions in other brain structures, which were probably not detected by CT scan. In their study, the core of thalamic lesions was located in the centromedian nucleus, and ventral posterior lateral and ventral posterior medial nuclei. Lehericy *et al.* (1996) performed MRI anatomoclinical correlations in patients with unilateral myoclonic dystonia resulting from localized cerebral infarction. Four patients had an isolated thalamic lesion and shared the same clinical and neuroanatomical features: myoclonic dystonia of hand or (fore)arm with contralateral involvement of the thalamus affecting the

ventral parts of the ventral lateral posterior nucleus with minor damage to the centromedian nucleus, but sparing the ventral lateral nucleus. Although there is a high degree of similarity between the clinical presentation in their patients and in our patient, three of their patients had associated hyperesthesia or hyperpathia. In their series, Krystkowiak *et al.* (1998) reported one patient with hand dystonia due to a focal thalamic infarction affecting the centromedian-parafascicular nucleus, the sensory nuclei (ventral posterior lateral nucleus ; ventral posterior medial nucleus and ventral medial basal nucleus) and pulvinar. The pulvinar was affected in their patients but was spared in ours. Therefore, it seems unlikely that this nucleus would be responsible for the occurrence of dystonia. The clinical and neuroanatomical findings of reported case of isolated hand dystonia associated with a lesion confined to the thalamus are summarized in the table.

The exact role of the thalamus in the occurrence of unilateral dystonia is unclear and remains to be elucidated. There is some experimental evidence that the centromedian nucleus might play an important role in the induction of dystonia. Indeed, experiments revealed the inhibitory role of the mesial thalamus on the ventral lateral nucleus, which receives afferents from globus pallidus and cerebellum (Milhorat, 1967 ; Sakai *et al.*, 1999). In addition, lesions of the pedunculopontine afferents to the centromedian nucleus could also induce dystonia (Deleu, 1997). Krystkowiak's findings (1998) could support the hypothesis that a lesion affecting one or several thalamic nuclei, including the centromedian nucleus, could play a role in the induction of focal dystonia in man. In our patient, the ventral anterior nucleus was spared. The ventral lateral and posterior lateral nucleus were partially affected but there was no sensory deafferentation and the SEP was normal, indicating that both nuclei were functionally intact. Furthermore, lesions affecting sensory nuclei (including ventral posterior lateral nucleus) are unlikely to induce dystonia as evidenced by the observation that positron emission tomographic studies did not display variations of activity in cortical sensory areas receiving thalamic sensory supply (Ceballos-Baumann *et al.*, 1995). Unilateral lesions affecting the dorsomedian nucleus of the thalamus can result in loss of vertical saccades with preserved vertical pursuit and vestibuloocular reflexes, hypersomnolence, amnesia and 'frontal syndrome', but have not been associated with dystonia (Gentilini *et al.*, 1987 ; Mitchell *et al.*, 1990).

The distribution of dystonia may differ with the location of the thalamic lesion (Marsden *et al.*, 1985). In the majority of cases, however, dystonia of the hand is observed and might be explained by selective destruction of neurons, which regulate upperarm movements. Neurophysiological data

Table

Summary of reported cases with pure hand dystonia as sequel of focal thalamic infarction. Those patients presenting with other concomitant movement disorders were not included

Author (Reference)	Clinical presentation of dystonia	Associated features at time of dystonia	Etiology	Thalamic nuclei involved (CT or MRI)
Karşıdağ <i>et al.</i> (9)	Case 3 : 43 y R hand - Dystonic spasm	R hemiparesis	Infarction	CT : L posterolateral thalamus
Krystkowiak <i>et al.</i> (11)	Case 3 : 72 y R hand - Myoclonic dystonia	Mild R hemiparesis	Hemorrhage	MRI : L thalamus (CM-Pf, VPL, VPM, VMB, P)
Lehéricy <i>et al.</i> (10)	Case 5 : 72 y R hand - Myoclonic dystonia	Mild R hypesthesia	Infarction	MRI : L thalamus (Vim, Vc)
	Case 6 : 66 y R hand - Myoclonic dystonia	None	Infarction	MRI : L thalamus (Vim, Vc)
	Case 7 : 71 y L hand - Myoclonic dystonia	L hyperpathia	Infarction	MRI : R thalamus (Vim, Vc)
	Case 8 : 56 y L hand - Myoclonic dystonia	Mild L hypesthesia	Infarction	MRI : R thalamus (Vim, Vc)
	Marsden <i>et al.</i> (2)	Case 10 : 76 y R Hand - Dystonic spasm	Increased pain and temperature threshold in R arm	Infarction
Our patient	Case 12 : 64 y L hand - Dystonic spasm	None	Infarction	CT : R posterolateral thalamus
	18 y R hand - Myoclonic dystonia	None	Perinatal Infarction	MRI : L thalamus (DM, CM-Pf, VL, VPL)

CM-Pf, centromedian-parafascicular nucleus; DM, dorsomedian nucleus; P, pulvinar; VL, ventral lateral nucleus; VPL, ventral posterior lateral nucleus; VPM, ventral posterior medial nucleus; VMB, ventral medial basal nucleus; Vim, ventral intermediate nucleus; Vc, Ventral caudal nucleus. R, right; L, left.

indicated somatotopic organization in the ventral lateral nucleus of the thalamus with a larger representation of upper limbs than lower limbs and face (Strick, 1976). Alternatively, predominance of the topographic representation of the upper extremity in the ventral part of the ventral posterior lateral nucleus could also contribute to this phenomenon (Ohye *et al.*, 1989). However, the latter area was seemingly spared in our patient.

In children, perinatal trauma, inflammatory or congenital vasculopathy, hemoglobinopathy, hypercoagulable states, or embolism caused by heart disease are common causes of cerebral infarction. The pathogenesis of thalamic infarctions in children remains poorly defined and no cases have been reported with isolated infarctions in the thalamus (Brower *et al.*, 1996). As commonly reported, there was a latency between injury and onset of dystonia which can be explained by neuroplasticity including late effects of neuronal synaptic reorganization, transsynaptic neuronal degeneration and slow aberrant neuronal sprouting stimulated by the primary lesion (Burke *et al.*, 1980).

The thalamic areas that are affected in our case receive their blood supply mainly from the anterior choroidal, posteromedial and posterolateral arteries. The former, a branch of the internal carotid artery, supplies the ventral part of the thalamus.

The latter ones, also known as the paramedian arteries (Reilly *et al.*, 1992 ; Chung *et al.*, 1996) usually arising from the posterior cerebral or posterior communicating arteries supply, the anterior, medial and posterior portions of the thalamus (Barr and Kiernan, 1993). In our patient, the lesion was most likely caused by an infarct of the posteromedial artery.

The pharmacological treatment of dystonia is generally disappointing and stimulation of the ventral posterior lateral nucleus or thalamotomy has been recommended particularly for hemidystonia (Andrew *et al.*, 1983). TENS proved to be an effective non-invasive therapy in this patient.

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