

Case report

# Dystonia after a bone fracture of the arm in a patient with a history of striato-pallidal ischemic stroke: a case report

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## Abstract

We report on a female with segmental dystonia of the upper limb after an anatomically related ischemic stroke. Dystonia developed almost 3 years after the onset of the stroke and immediately following the removal of the cast because of a bone fracture in the same limb. We discuss the case considering issues such as: delay-onset, lesion topography and pathophysiology, peripheral input and their possible contribution to the development of secondary dystonia.

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## 1. Introduction

Dystonia is a syndrome characterized by sustained and forceful muscle contractions, frequently causing twisting and repetitive movements, or abnormal postures. According to the etiological classification, dystonia can be divided into two major categories: idiopathic or primary and symptomatic or secondary [1]. Delayed-onset focal or segmental dystonia secondary to focal cerebral vascular injury, such as infarction of the basal ganglia is a rare but well-described entity [2–6]. Moreover selectivity of the lesions within the basal ganglia that result in dystonia is a matter of great interest because it may give clues to the functional organization of the lesions. Finally, there is a growing body of evidence that defective sensory inputs could play a crucial role in the development of dystonic dyskinesia [7]. Our case describes a patient with a history of right striato-pallidal ischemic stroke, who developed segmental dystonia of the left upper limb 2 months after a peripheral injury (bone fracture) of the left elbow and immediately following the removal of the plaster cast. Our purpose is to consider among others issues the following crucial question; was the

onset of the movement disorder related to the peripheral trauma or could it have occurred by chance?

### 1.1. Case history

A 58-year-old right handed female was referred to a movement disorder outpatient clinic complaining of ‘pain and discomfort’ as well as marked involuntary movements restricted to the left hand. Three years before she had a stroke of the right basal ganglia territory (Brain CT scan: hypodense area at the anatomical position of the right putamen). resulting in a left hemiparesis involving mainly the upper limb, together with mild paralysis of the left lower face (asymmetry of the nasolabial fold). At the time the patient had no sensory complaints or defects. An NIH stroke scale performed immediately after the insult showed only minor motor impairment [score 1 for ‘facial facial palsy’ (item 4) and score 1 for ‘motor arm and leg’ (item 5 and 6)] and the neuropsychological evaluation revealed some degree of apathy and loss of drive. Apart from mild hypertension for the previous 5 years, medical history was unremarkable and she had no family or personal history of psychiatric or movement disorders. Following the stroke she was seen every 6 months in the outpatient clinic, and by the end of the first year the patient had no residual weakness of the left hand.

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About two and a half years after the stroke she fractured left elbow and she was in a plaster cast for the next 2 months in order to immobilize the joint. Immediately after the removal of the cast at the end of 2 months, the patient experienced marked discomfort of the hand, which was not helped by physiotherapy. The discomfort ‘forced’ the patient to make continual ‘irregular, mainly twisting,

movements’ of the hand mostly characterized by finger extension and abduction, or flexion of fingers III and IV in the metacarpophalangeal joints with extension of the other finger joints, pronation of the forearm, and to a lesser degree by internal rotation, as well as adduction of the arm. She also stated that when walking she had less discomfort when the limb ‘goes’ at behind her. Muscle strength in the hand was almost normal and the clinical examination did not reveal additional signs. The continuous writhing character of the movements added an athetotic component to them. There were no signs of autonomic dysfunction of the limb as seen in cases of complex regional pain syndrome (CRPS). There was no history of toxic exposure or treatment with neuroleptic drugs. The patient underwent a brain MRI (Fig. 1), which showed the old lesion of the right putamen and the globus pallidus with additional involvement to a minor degree of the right internal capsule and gliosis at the border zone around the lesion. The pattern of the dystonic movements did not change over a 2-year follow-up period.

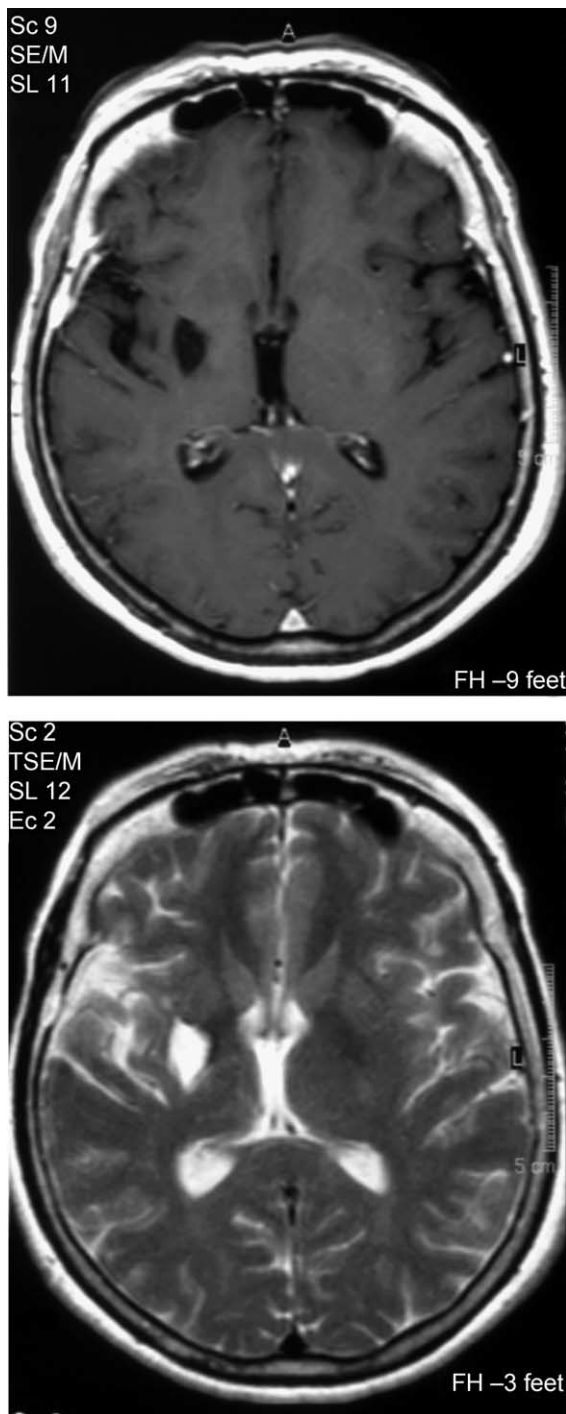


Fig. 1. Brain MRI of the patient. The lesion is visible as hypodense and hyperdense area on T1 and T2 weighted images, respectively, involving the posterior part of the right putamen and the globus pallidus.

## 2. Discussion

Delayed-onset dystonia is a rare sequel of stroke. Delayed-onset of a movement disorder after ischemic-hypoxic injury may reflect the time required for remyelination, inflammatory changes, ephaptic transmission, oxidation reactions, maturation or aberrant synaptic reorganization, trans-synaptic neuronal degeneration, or denervation supersensitivity [11].

The anatomical basis and pathogenesis of secondary dystonia are both uncertain. The involvement of the internal capsule together with the adjacent pallido-striatal complex in stroke patients with dystonia confined to the upper limb has been also demonstrated by others [5,6,13]. Moreover, the literature concerning the question of which structure among putamen and globus pallidus, internal (GPi) or external (GPe), is mainly responsible for the development of secondary dystonia, is conflicting and confusing. It has been suggested that putaminal lesions might affect both direct and indirect pathways [14]. This disruption may predominantly affect the indirect pathway resulting in dystonia by increased thalamocortical drive due to disinhibition of the thalamo-cortical projections [6,15]. On the other hand, there is indirect evidence in primates and humans showing that, at least in primary dystonia, both the direct and indirect pathways are overactive, and certain models of primary dystonia share this notion [15]. If this is a valid point, then secondary dystonias associated with putaminal lesions are difficult to explain based on striatal overactivity. The same difficulty exists in the case of involvement of the external globus pallidus (GPe) since lesions of GPe tend to produce parkinsonism by disinhibition of the subthalamic nucleus (STN) [17], and certain movement disorders like chorea

result from increased neuronal activity of GPe neurons [19]. Finally in the case of GPi involvement there is also the paradox that although GPi pallidotomy alleviates symptoms in patients with dystonia, by altering the neuronal activity [16], involvement of GPi in secondary dystonia has also been reported by many investigators [4–6,10,13,18]. All these findings suggest the possibility that either a different central pathophysiological mechanism may be responsible for the development of primary vs secondary dystonia, or that in the case of primary dystonia, the palliative result of pallidotomy could be due to the precise lesion location within the GPi (posteroventral) [19].

Another issue in our case has to do with whether the onset of the movement disorder (dystonia) was related to the peripheral trauma or whether it could have occurred by chance? Both peripheral injury and immobilization are responsible for the peripheral input. Although the opposite idea has also been stressed [9], a recent review provides evidence that different types of peripheral injury, including direct trauma and chronic immobilization with plaster cast or splint, may lead to certain movement disorders like focal or segmental dystonia [8].

In contrast to Jankovic assertion [8], namely that peripherally induced post-traumatic dystonia is often characterized by the presence of CRPS, this was not the case in our patient, as has also been reported by others [20]. In addition, our patient's dystonia was not characterized by inconsistency, spontaneous remissions, disappearing with distraction, incongruity of movements, etc. that could suggest a psychogenic origin [24–26].

It has been suggested that one of the functions of the basal ganglia is to regulate sensorimotor interactions in a way that determines which sensory stimuli are used to initiate motor action and which are disregarded [12,14]. Moreover, the participation of the somatosensory system in the mechanism of dystonia has been suggested [7]. One important indication in favour of this notion is the alteration of dystonic muscular patterns after successful botulinum toxin treatment [21]. There is a growing body of evidence that a preexisting dysfunction of the central nervous system plays an important role in the pathogenesis of this kind of movement disorder, and predisposes some patients to develop it. Animal studies also provide evidence that peripheral trauma can 'unmask' subclinical disturbance of basal ganglia function [8], and that localized lesions in the GPe produced certain hyperkinetic syndromes triggered by sensory stimulation [22]. If this is the case, then we can argue that our patient could possibly be an example in which an underlying central nervous system disorder was silent until a peripheral cause triggered the development of the dystonic movements. This may also explain why the dystonia was restricted to the upper limb since we can assume that the bone fracture or immobilization could induce a peripheral sensory modification that 'marked' the left upper limb as the body part that will develop dystonia. Instead, if we reject the contribution of peripheral sensory

input, we can possibly accept that the dystonia was confined to the left hand and arm because of the topography of the lesion within the striato-pallidal complex [23].

In summary, although we realize that it is impossible to draw conclusions in such complicated issues in a single patient report, we suggest that our patient 'meets both criteria' for a mixed delay-onset secondary to stroke and also a peripherally induced dystonia [8,11], and that this case could be an example supporting the notion that in the case of secondary dystonia, the sensory abnormalities are probably not responsible for the primary pathology and that people who develop peripheral trauma-induced dystonia may be somehow predisposed to the development of dystonia [9]. It is possible that changes in neuronal activity in basal ganglia or thalamus or even cortex together with changes in the receptive field properties of neurons of these areas could contribute to the development of dystonia. This perhaps points towards the necessity for studies that can compare baseline characteristics of neuronal activity in putamen and also in GPe and GPi in different clinical and pathophysiological types of dystonia, taking also into consideration the somatosensory input as an important component in the development of this particular movement disorder.

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