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SHORT REPORTS

A Case of Masticatory Dystonia Following Cerebellar Haemorrhage

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Introduction

Dystonia is a movement disorder characterized by sustained or intermittent muscle contractions causing abnormal, and often repetitive, movements, postures, or both [1]. It has been attributed to basal ganglia abnormalities and to dysfunctional cortico-striato-thalamo-cortical circuitry [2–4]. Dystonia has a number of possible etiological causes. Focal dystonia is more common than generalized dystonia. In cranial and cervical focal dystonia, neurophysiological investigations have revealed abnormally excitable interneuronal pathways in the brainstem. For instance, several studies reported changes in the length of the recovery cycle of the eye blink reflex and the masseter inhibitory reflex [5–7]. Cortical dysfunctions have also been explored using transcranial magnetic stimulation [8, 9].

Acquired forms of dystonia have been associated with a wide variety of vascular, traumatic, infectious, toxic, and degenerative processes, which can affect either the central or peripheral nervous system [10–14].

The underlying mechanisms of acquired dystonia are not yet completely understood, although several different studies have implicated a neural network linking the cerebral cortex, basal ganglia, and cerebellum [2–4]. In such a network, dysfunctional activity at any level could lead to the onset of dystonia. While the role of the basal ganglia is well known and had been widely demonstrated with respect to the pathogenesis of acquired dystonia, the role of the cerebellum has not been well defined. Both animal studies [15–17] and clinical reports [3, 18–34] have explored the role of the cerebellum in the pathogenesis of dystonia. Overall, there is consensus regarding the idea that acquired dystonia is caused by lesions, leading to abnormal cerebellar output (Table 1).

In this case report, we describe our diagnostic and therapeutic processes for a patient with masticatory dystonia resulting from cerebellar haemorrhage.

Case Report

We assessed a 59-year-old Caucasian man with a medical history of hypertension. The patient was currently taking dihydropyridine and ACE inhibitors. He had been diagnosed with HIV 6 years prior to the study and was receiving HAART therapy for pulmonary sarcoidosis. Finally, he was receiving fluoxetine for chronic psychosis.

The patient had experienced a left cerebellar spontaneous haemorrhage in 2004. Following the haemorrhage, he developed hydrocephalus, for which he received external ventricular derivation, and the haemorrhage was surgically evacuated. Several months later, he received a definitive ventricular-peritoneal shunt. During this period, the patient started to complain of persistent activity of the jaw-closing muscles, preventing jaw opening and limiting speaking and feeding. For this reason, the patient was referred to our department in 2006.

A neurological examination showed signs of left hemispheric cerebellar dysfunction: nystagmus in the right lateral gaze, left hand dysmetria, and left ataxic gait. The dystonic symptom was clinically represented by persistent tonic activity of the jaw-closing muscles that allowed a maximum jaw opening of just 5 mm. This was associated with hypertrophy of the left temporalis muscle.

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Table 1 Review of acquired dystonia cases reported in literature

Reference	Site of lesions	Type of lesion	Time of onset of dystonia after lesion occurrence	Type and side of dystonia
Vascular diseases				
Tranchand et al. [31]	R cerebellum	Cavernous hemangioma	n.d.	Cervical dystonia
Waln, Le Doux [34]	R cerebellar	Haemorrhage	20 years	Oromandibular dystonia
Usmani et al. [33]	Cerebellar vermis	Haemorrhage arterovenous malformation	15 months	L torticollis
Rumbach et al. [30]	L cerebellum	Ischemic stroke	2 months	L hemidystonia
Alarcon et al. [19]	R cerebellum/L thalamic	Ischemic stroke	2 months	R hemicorea
O'Rourke et al. [28]	R and L cerebellum	Ischemic stroke	3 days	R sided torticollis and bilateral blepharospasm
Zadro et al. [35]	L cerebellum	Ischemic stroke	2 days	R sided torticollis
Khooshnoodi et al. [24]	L dentate nuclei (case 3) L cerebellar (cases 4 and 13) L inferior cerebellar peduncle (case 10)	Cystic (case 3) Ischemic stroke (cases 4, 10, and 13)	More than 40 years	Blepharospasm
Occupying space lesions				
Le Doux, Brady [3]	L cerebello-pontine angle	Arachnoid cyst	–	Cervical dystonia
Kojovic et al. [25]	R cerebellar	Cystic	–	L torticollis R laterocollis
Boisen [22]	Cerebellar (cases 1 and 2)	Hemangioblastoma (cases 1 and 2)	–	Cervical dystonia
Turgut et al [32]	Posterior fossa	Medulloblastoma, glial tumours, arachnoid cyst, and tuberculoma	–	Cervical dystonia
Kumandas et al. [26]	(Cases 1 and 4) posterior fossa (mesial)	Astrocytoma	–	R sided torticollis (case 1) L side torticollis and posterior anterocollis (case 4)
Caress et al [23]	Cerebellar	Gangliocytoma	–	Cervical dystonia
Krauss et al. [27]	Cerebello-pontine angle (case 1) L cerebellar (case 2) R cerebellar (case 3)	Schwannoma (cases 1 and 2) Meningioma (case 3)	Several months (case 1) (Cases 2 and 3)	L side head jerk (case 1) R side torticollis (case 2) L side torticollis (case 3)
Alarcon et al. [18]	L (vermian and paravermian) cerebellum	Tuberculoma	2 weeks	L arm dystonia
Other causes				
Plant [29]	R midbrain R cerebellar	Multiple sclerosis	1 year	L cervical torticollis
Alcalay et al. [20]	R L dentate nuclei	Cerebrotendinous xanthomatosis	3 years	Oromandibular dystonia
Baik [21]	R cerebellar	Atrophy	–	R arm dystonia

R right, *L* left

A T1-weighted MRI scan revealed vermian atrophy and left paramedian cerebellar encephalomalacia to be outcomes of a previous stroke (Fig. 1).

Although fluoxetine therapy was stopped, in consideration of its possible effect on dystonia, we saw no subjective or objective clinical improvements.

A neurophysiological assessment including needle electromyography (EMG) of the masticatory muscles, the blink reflex recovery cycle, the masseter inhibitory reflex, and the masseteric silent period revealed a dystonic activation pattern in the left masseter and left temporalis muscles during jaw opening. We also found an altered R2 in the recovery cycle of the blink reflex, recorded from the left orbicularis oculi muscle, with a normal masseter inhibitory reflex and masseteric silent period.

In consideration of the observed clinical and electromyographic patterns, we injected an abobotulinum neurotoxin into the dystonic muscles, with the aim of reducing dystonic activity during jaw opening. This treatment produced an improvement in drinking, feeding, and speaking. The infiltration of the abobotulinum toxin was realized using an EMG guide to identify the exact injection point in the target muscle.

We assessed the treatment efficacy by measuring the jaw opening and using the Baylor Rating Scale [36].

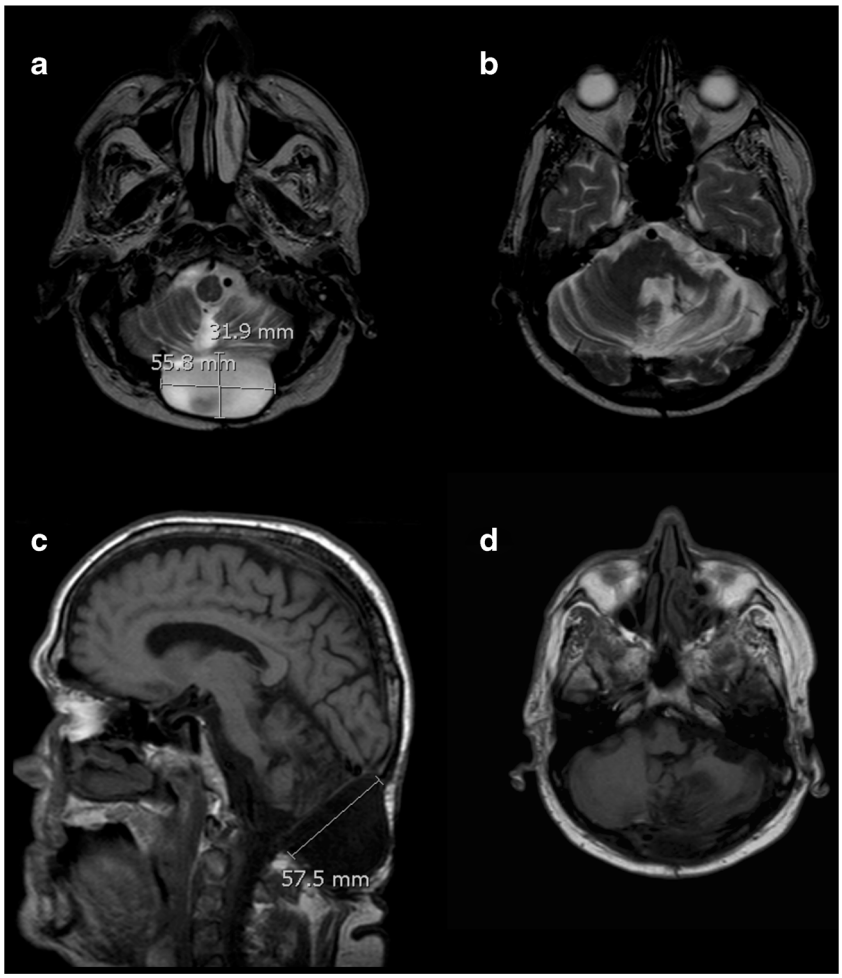

Fig. 1 T2- (**a** and **b**) and T1-weighted (**c** and **d**) MRI scan showing vermian and left paramedian cerebellar encephalomalacia and encephaloceles as stable outcome of a cerebellar haemorrhage occurred in 2004

The effective dose of abobotulinum toxin was 400 U in the left masseter muscle and 600 U in the left temporalis muscle. We also injected 200 U of abobotulinum toxin A into the non-dystonic right masseter muscle. A clinical evaluation 1 month after the injection showed a significant improvement in dystonic symptoms: the patient was able to articulate words more clearly, chew solid food, and drink from a glass without a straw. We did not observe excessive weakness of the masticatory muscles or any other side effects of the treatment.

Our treatment confirmed the efficacy of abobotulinum toxin for acquired dystonia. Following our treatment, the patient was able to normally chew solid food, drink from a glass, and talk fluently.

Discussion

There are several different pathogenetic and etiological explanations for the dystonic symptoms observed in our patient.

The neuronal network responsible for altered movement control in cases of dystonia includes the basal ganglia nuclei, the cerebellum, and the cortical and subcortical connections between these regions. Dysfunctional activity at any level of this network can lead to dystonia.

As stated by Malone et al. [37], recent human studies and experimental data indicate that cerebellum circuitry could play an important role in the pathogenesis of some forms of dystonia. For instance, irritative cerebellar lesions, which do not involve the loss of cerebellar tissue but rather distort cerebellar output, can lead to dystonia [38]. These lesions are likely to be related to focal haemorrhages or space-occupying lesions that compress and distort cerebellar function.

Rodent models have contributed to an improved understanding of the links between the cerebellum and dystonia [39]. For instance, an electrophysiological study that used dystonic mice revealed aberrant high-frequency bursting activity in cerebellar nuclei [40]. The firing patterns of Purkinje neurons have also been found to be impaired [40]. Another study used a behavioural adaptation paradigm to highlight the links between dystonia and the cerebellum [41]. The researchers described several neurological factors that may be a possible cause of dystonic symptoms. Although it was a

possibility that we considered, there is no evidence in the literature to suggest a relationship between intracranial hypertension and dystonia.

Although the pathophysiological mechanisms of the extrapyramidal symptoms induced by fluoxetine remain unclear, fluoxetine therapy may cause transient or persistent dystonia [42]. An interaction between serotoninergic and dopaminergic pathways may involve serotonin inhibitory actions on extrapyramidal dopaminergic activity. In our patient, fluoxetine was administered after a stroke, and suspension of this drug did not result in clinical improvement.

There is no typical EMG activity pattern that would allow one to distinguish between dystonic contractions and other types of muscle activity; however, based on the definition of dystonia [1], the presence of EMG activity in muscles devoted to jaw closure during voluntary jaw opening is strongly suggestive of a dystonic co-contraction of antagonist muscles.

Although we considered hemimasticatory spasm when making our differential diagnosis, we did not observe typical brief trains of a few single motor unit potentials reaching a discharge frequency of 100 Hz or brief bursts of multiple motor unit potentials associated with hemimasticatory spasm [43]. Our recordings of the masseter inhibitory reflex and masseteric silent period showed normal parameters. These reflex responses often have abnormal parameters in patients with hemimasticatory spasm [44].

We did not expect dystonic activity in our patient to be left lateralized. Our neurophysiological assessment indicated similar lateralization. The recovery cycle of the blink reflex was altered in the left orbicularis oculi muscle, and the left masticatory muscles showed dystonic activation patterns. The concurrence of dystonic and cerebellar abnormalities on the same side of the brain indicates that there is a common pathogenetic mechanism. As suggested in a case report by Waln and LeDoux [34], lateralized dysfunction of the olivocerebellar pathways [45] may have caused the jaw-closing dystonic pattern observed in our patient. However, the timing between the acute lesion and the onset of dystonia was different in the two cases, and we did not observe tongue dystonia in our patient. Although a definitive causality is impossible to establish, we hypothesize that cerebellar lesion plays a fundamental role in the pathogenesis of masticatory dystonia.

Conflict of Interest During the last 2 years, Maurizio Osio received funding from Ipsen SpA, Merz SpA, and Allergan SpA.

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