Chapter 7

'Mirror' dystonia in writer's cramp

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Abstract

In some patients with writer’s cramp, writing with the asymptomatic arm induces dystonia in the symptomatic arm. In a patient with this ‘mirror’ dystonia we recorded angulation movements of both wrists simultaneously during writing. Abnormal movements occurred only in the symptomatic wrist. The abnormal movements during writing with the asymptomatic hand resembled closely abnormal movements during writing with the symptomatic hand. The feature of ‘mirror’ dystonia may help identify primary involved muscles and help elucidate the pathophysiology of writer’s cramp.
Introduction

In the treatment of writer's cramp with botulinum toxin injections, identification of muscles primarily responsible for the dystonic movements is essential. Mostly, identification of responsible muscles is done by careful clinical observation during writing, sometimes supported by electromyographical recordings (Cole et al.1995). Identification of responsible muscles, however, remains difficult in many patients. This difficulty is mainly due to compensatory movements and postures necessary to be able to write despite the dystonic movements. In a patient with writer's cramp, we recorded dystonic movements in the symptomatic arm induced by writing with the asymptomatic arm. We want to draw attention to these movements, as they may be helpful for the identification of responsible muscles for treatment with botulinum toxin.

Patient and methods

A 55-year old right-handed man presented with writing difficulty, due to involuntary movements and posturing of the right hand and arm, which had gradually increased for four years. The last two years the patient experienced increasing difficulties in other tasks, such as typing, stirring, cutting, and spreading bread with butter, as well. He had taught himself to write with the left hand, but he had noticed involuntary movements in the right hand while doing so. To prevent these movements he had grown accustomed to sit on his right hand while writing. On examination, when writing with the right hand it appeared difficult to hold the pen on the paper due to prominent extension and ulnar deviation movements of the right wrist. In addition there was abduction of the upper arm. When writing with the left hand extension and ulnar deviation movements were also present in the right hand, but there was no abduction of the upper arm. Except for the involuntary movements in the right arm, neurological examination was normal.

The wrist movements were studied with the Greenleaf wrist system (Greenleaf Medical Systems, Palo Alto, California). This system consists of a glove fixed around the wrist with two transducers for detection of flexion/extension and ulnar/radial deviation movements. Recordings were performed in both wrists simultaneously. After calibration of angles the patient was asked to write first with the right and subsequently with the left hand. Angulation movements were displayed on line on a mini-computer.
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Results

An example of the handwriting with the right and the left hand is given in figure 1. The movement analysis is depicted in the figures 2 and 3. The registration and the writing start at 0 seconds and so the wrist posture at this moment represents the resting position. During writing with the right hand there are gross ulnar deviation and extension movements in the right wrist, while virtually no movements are seen in the left wrist (figure 2a). Writing with the left hand is largely done by movements of the fingers. During writing with the left hand only minor wrist deviations are present on the left side and these minor deviations thus reflect the normal writing movements for this patient (figure 2b). However, again gross abnormal movements are seen in the right wrist (figure 2b). They resemble closely the abnormal movements, which occurred during writing with the right hand, but corrective movements, seen as rapid flexion movements towards the baseline position in figure 2a, are made less often. When drawing circles with the right and left hand as well, movements in the right wrist were much less pronounced than during writing (figure 3).
Figure 2.
(a) writing with the symptomatic right arm elicits gross extension (R E/F) and ulnar (R R/U) deviations in the right wrist, while the left wrist remains quiet (L E/F, L R/U).
(b) writing with the asymptomatic left hand again elicits gross extension (R E/F) and ulnar (R R/U) deviations in the right wrist, while again the left wrist (L E/F, L R/U) remains quiet. The figures are redrawn as the original figures had colored lines.)
Figure 3.
(a) Drawing circles with the symptomatic right (R) hand elicits only minor extension (E/F) and ulnar (R/U) deviations in the right wrist, the left wrist (L E/F, L U/R) remains quiet.
(b) Drawing circles with the left hand elicits only minor extension (R E/F) and ulnar (R R/U) deviation movements in the right wrist. (The figures are redrawn as the original figures had colored lines)
Discussion

In treatment of writer’s cramp with botulinum toxin it is often difficult to select muscles that are primarily responsible for the abnormal movement. This is predominantly due to compensatory movements, necessary to hold the pen and to keep the pen on the paper. ‘Mirror’ dystonia in patients with writer’s cramp was first described by Sheehy, Rothwell and Marsden (1988), but has received little attention since. As the movements in ‘mirror’ dystonia have no compensatory component, dystonic activity in the symptomatic arm, disclosed by writing with the asymptomatic arm is likely to be that of the primary involved muscles. Therefore, when present, ‘mirror’ dystonia may be helpful in selecting muscles for botulinum toxin injections in writer’s cramp. In the presented patient this is demonstrated by the abduction movements in the shoulder present during writing with the right arm, but absent in ‘mirror’ dystonia. Therefore, abduction movements in the shoulder are probably compensatory, necessary to keep the pen on the paper. The ‘mirror’ dystonic movements in our patient were evident by clinical observation. Recording with the Greenleaf wrist system facilitated quantification and insight into the combined direction of the movements.

The pathophysiology of ‘mirror’ dystonia is unclear. The abnormal movements in ‘mirror’ dystonia may be confused with mirror movements. Mirror movements are defined as involuntary movements on one side of the body that occur as mirror reversals of an intended movement on the other side of the body (Zülch and Müller 1969; Cohen et al. 1991). In patients with congenital mirror movements there is an aberrant organization of cortical motor representation areas and corticospinal pathways with ipsilateral as well as contralateral control of voluntary movement (Cohen et al. 1991). However, mirror movements differ from the movements in mirror dystonia as the latter are not homologous and imitative and only occur during the performance of a specific task.

Idiopathic dystonia commonly begins with a specific action dystonia. As the dystonic condition progresses, less specific actions of the affected arm may activate the dystonia and, with further evolution, actions in other parts of the body can induce dystonic movements of the involved arm, so-called ‘overflow’ (Fahn et al. 1987). In this respect ‘mirror’ dystonia is likely to be a task specific overflow phenomenon. Writing requires a high level of differentiation in the generation of muscle activity. Recently, in a primate
genesis model of focal dystonia it has been shown in a passive paradigm of repetitive strain, that cortical dedifferentiation of sensory information occurs (Byl et al. 1996). Although in writer’s cramp sensory information processing may be disturbed (Hallett 1995), ‘mirror’ dystonia suggests the presence of task specific dedifferentiation of executive (motor) rather than of receptive (sensory) functions. ‘Mirror’ dystonia may show to be helpful in the clinical selection of responsible muscles in the treatment with botulinum toxin and in the study of the pathophysiology of dystonia as well.

References


