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Case report

Obsessive-compulsive disorder and executive deficits in two patients with primary dystonia

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Abstract

In spite of the high prevalence of behavioral and cognitive disturbances found in most basal ganglia disorders and attributed to fronto-striatal dysfunction, the existence of psychiatric and cognitive symptoms in patients with primary dystonia remains controversial. We present a 42-year-old female with primary writer's cramp and obsessive-compulsive disorder (OCD) and a 59-year-old male with Meigs syndrome, idiopathic torticollis and OCD. Both patients had mild executive dysfunction. The coexistence of psychiatric, cognitive and motor symptoms of different intensity may be explained by variable dysfunction on different frontal–striatal loops, as proposed by the open interconnected model of fronto-striatal circuits.

Keywords: Primary dystonia; Obsessive-compulsive disorder; Executive dysfunction; Fronto-striatal circuits

1. Introduction

Frontal lobe deficits [1] and psychiatric disorders (obsessive-compulsive disorder—OCD, depression, psychosis) [2] are frequently found in Huntington's disease, Progressive Supranuclear Palsy and Parkinson's disease, and have been related to dysfunction in circuits linking prefrontal cortex and basal ganglia. Alexander's model of five structurally and functionally segregated cortical–basal circuits (anterior cingulate, dorsolateral prefrontal, lateral orbitofrontal, motor and oculomotor loops) [3] implies that the coexistence of behavioral and motor symptoms in these disorders would necessarily be due to dysfunction in more than one loop and its corresponding structures. Recently, Joel and Weiner model of open loops with bridging connections between motor, associative (anterior cingulated and ventrolateral and dorsolateral prefrontal) and limbic (orbitofrontal) circuits solved this problem by predicting that dysfunction in a single circuit may eventually produce changes in more than one functional domain [4].

Primary dystonia is a syndrome of sustained muscle contractions, frequently causing twisting and repetitive movements, or abnormal postures, for which no etiology can be found, and where dystonia is the sole phenotypical manifestation [5]. Primary dystonia's pathophysiology is less well known than in other movement disorders. Evidence points to fronto-striatal dysfunction, with reduced frontal cortical inhibition by the basal ganglia (motor loop dysfunction) [6]. However, the existence of cognitive and behavioral disturbances in primary dystonia is still a mater of debate. We report two cases in which primary dystonia, OCD and mild executive dysfunction coexist.

2. Case reports

2.1. Case 1

At the age of 20, this 42-year-old female secretary developed a right hand tremor when holding objects or

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performing precision movements. She had no major difficulty in writing until she was 34, when she began to feel involuntary contractions of her right hand fingers during handwriting, with excessive griping of the pen. The patient found that placing her left index finger over the dorsum of the right hand when writing could ease muscular contractions. These symptoms aggravated over several years, becoming more intense and spreading to other muscle groups, affecting wrist, arm and shoulder movements, eventually disturbing many of her daily activities. She became nearly unable to write and stopped working at the age of 37. She denied the use of neuroleptics or other dystonia-inducing drugs prior to disease-onset. Family history was unremarkable.

Neurological examination: during writing, the patient developed involuntary contraction of the right fingers and dystonic movements of her right arm. Similar involuntary movements appeared when grabbing an object or drinking from a bottle. She alleviated her dystonic postures by placing her left index finger over the right hand's dorsum. Further neurological examination was unremarkable.

Brain MRI, serum copper and coeruloplasmin levels, CBC and ESR where normal, excluding secondary causes. Botolinium toxin was started, with temporary improvement. She is currently doing one session every 3 months.

Interviewing revealed onset of psychiatric symptoms at age 38, with growing obsessive concerns about cleanliness and compulsive, elaborated hand-washing rituals that consumed most of her day. She also had increasing difficulty in going to places more than a few meters away from home due to fears that a drop of dirty water might fall on her head from a balcony. This fear invariably made her return home and compulsively wash her hair, usually several times in one afternoon. She eventually developed dermatitis in her hands and scalp. She also eventually developed checking and repeating rituals (e.g., repeatedly closing a door or switching off a light). At the time of first examination she had been on paroxetine 40 mg i.d. for 2 years, with subjective relief from obsessive-compulsive symptoms. She nonetheless still scored 24 on the Yale Brown Obsessive-Compulsive Rating Scale (YBOCS; cut-off 16). She fulfilled DSM-IV criteria for OCD. No further significant psychiatric symptoms were found.

Cognitive function was evaluated by means of: MMS Examination; Stroop test and Wisconsin Card Sorting Test (WCST) for executive functions; Benton's Visual Memory Test (BVM) and the cube test from Wechsler's Adult Intelligence Scale (WAIS) for visuo-spatial functions. All results were normal except for the WCST, where she scored in the *mild dysfunction range* for 'total number of errors' and in the *mild to moderate dysfunction range* for 'number of perseverative responses' and 'number of perserverative errors'.

2.2. Case 2

At the age of 58, this 60-year-old retired man developed excessive blinking, especially when exposed to bright light and wind, which gradually evolved to forced unsuppressable closure of the eyelids. A few months later, further occasional involuntary grimacing appeared, and finally forced contraction of the neck muscles, with flexion movements of the head. The blinking, grimacing and neck movements became more intense over time, limiting many of his daily activities.

The first neurological examination showed increased blinking frequency, sometimes with forced, prolonged eye closure, brief occasional lip-contraction and grimacing, and involuntary contraction of the sternocleidomastoideus muscles bilaterally. One year later, Meig's syndrome was evident, with occasional blepharospasm and pronounced dystonic movements of the lips, tongue and several muscles of the lower face, as well as pronounced anterocollis. Pressure over the chin alleviated anterocollis. He had no other signs of neurological dysfunction. He denied having ever used neuroleptics or other drugs suspected of causing dystonia. Brain MRI and copper metabolism studies were unremarkable. He was started on botulinium toxin, with temporary improvement.

Although the patient initially denied any psychiatric complaints, search for obsessive-compulsive psychopathology revealed excessive concern about hand-perspiration leading to frequent hand washing which the patient himself found exaggerate. He also was deeply concerned about pollution and the presence of garbage near his house. He therefore spent 3–4 h daily walking about his neighborhood collecting garbage he would then dispose of in the way he felt proper. He did this during the night so that nobody could see him, as he felt his conduct to be abnormal. He also carefully selected "toxic" items from his own domestic litter and kept them indefinitely awaiting "adequate" disposal. He scored 29 in the YBOCS (cut-off 16). OCD was diagnosed, as defined by DSM-IV criteria. Meaningful psychopathology other than OCD was absent. He was on no psychiatric medication at the time of observation.

The patient scored in the mild dysfunction range for 'total number of errors' and 'number of nonpersevarative errors' and mild to moderate dysfunction for 'number of perservarative responses' and 'number of perseverative errors' in the WCST. He scored normally in the MMS, BVM, Stroop and WAIS cube tests.

3. Discussion

Both patients present primary segmental dystonia (writer's cramp and Meig's syndrome plus spasmodic torticollis), as well as severe OCD. Although neither