

Journal of Neurolinguistics 22 (2009) 83-90

Journal of NEUROLINGUISTICS

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Neurogenic stuttering in corticobasal ganglionic degeneration: A case report

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Received 8 November 2007; received in revised form 30 June 2008; accepted 30 June 2008

Abstract

Corticobasal ganglionic degeneration (CBGD) is a progressive neurological disorder characterized by gradual nerve cell loss and atrophy of the cerebral cortex and basal ganglia. Symptoms of the disorder include verbal apraxia and language disturbances along with bradykinesia and rigidity. There have been no reports to date of acquired or neurogenic stuttering associated with CBGD. We describe a patient whose initial symptom of CBGD was stuttering which worsened as her disease progressed. Neuroimaging including PET scans revealed poor metabolic functioning of the right basal ganglia. This finding, along with bilateral atrophy of the frontal and parietal lobes likely contributed to the disturbance of motor sequencing skills and led to the development and worsening of stuttering, apraxia of speech and swallowing, and eventual aphasia and cognitive decline. We suggest that neurogenic stuttering may be an additional symptom of CBGD.

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Keywords: Corticobasal ganglionic degeneration; Neurogenic stuttering

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^{0911-6044/\$ -} see front matter © 2008 Elsevier Ltd. All rights reserved. doi:10.1016/j.jneuroling.2008.06.003

1. Introduction

Corticobasal ganglionic degeneration (CBGD) is a progressive neurological disorder characterized by gradual nerve cell loss and atrophy of the cerebral cortex and basal ganglia. Symptoms include the progression of tremor, verbal and limb apraxia, bradykinesia and rigidity (Riley et al., 1990). Progression of symptoms in an asymmetric manner is a key feature of the disorder (Jankovic, 2007). The term corticobasal degeneration (CBD) is also used to describe the disease (Litvan et al., 1997). It has been suggested in the literature that CBGD is an under diagnosed disease particularly due to its similarity to Parkinson Disease (PD) and progressive supranuclear palsy (PSP) (Litvan et al., 1997). The best predictors of diagnosis of CBGD include asymmetric akinetic-rigid syndrome with late onset of gait and balance disturbances, limb dystonia, myoclonus and ideomotor apraxia (Litvan et al., 1997). Other symptoms include verbal apraxia, dysphasia, dementia, dysarthria and cortical sensory disturbances (Riley et al., 1990). In a recent study by Frattali, Grafman, Patronas, Makhlouf, and Litvan (2000) the language disturbances of 15 patients with CBD were described. The authors reported that 53% of their patients demonstrated aphasia syndromes including Broca's aphasia, anomic aphasia, and transcortical motor aphasia. Their patients demonstrated left frontal, parietal or temporal lobe atrophy on brain MRI studies.

In this paper we describe the progression of speech deterioration in a patient with a medical diagnosis of CBGD whose initial and continual complaint was stuttering. As her illness progressed, her stuttering worsened, leading to the impression that neurogenic stuttering may be an additional symptom of CBGD.

2. Case Report

Our patient, a 68-year-old right handed female, was initially seen by her neurologist due to concerns of stuttering and word finding problems. She was a fluent speaker throughout her childhood and adulthood without a history of developmental stuttering. She was extensively involved in volunteerism and gave frequent speeches and presentations. She described symptoms of stuttering and not being able to "get the word out" over the course of 1 year prior to her neurological examination. She described the problem as worse when she was anxious. She was becoming embarrassed by her non-fluent speech and began avoiding social speaking situations. She had a longstanding history of Bell's Palsy with a residual right facial droop and ptosis. She reported new numbness in the center of her upper lip. She had a history of bilateral hand tremor since childhood. There was a significant family history of tremor. There was no reported history of stuttering in her family but the patient reported that as her mother aged she developed speech problems, the specifics of which were not identified by the patient. There were no complaints of blurred or double vision, vertigo or dizziness. Gait was normal.

The patient's initial evaluation by the speech—language pathologist revealed stuttering-like behavior in the form of difficulty initiating speech and sound repetition particularly on the initial sound of words during conversation. She also demonstrated inconsistent sound substitutions, additions and repetitions when reciting phonemically easy to complex words and phrases from the verbal agility subsection of the Boston Diagnostic Aphasia Examination (BDAE) (Goodglass & Kaplan, 1983a). The patient achieved a score of 8/14 on the verbal agility subtest of the BDAE due to sound repetition of the first or second syllable of words. A poor oral agility score on the BDAE (6/12) occurred due to slow and uncoordinated rate of isolated and alternating tongue and lip movement during the 5 s allotted time period for the response. Unsteady,

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