The Broader Role of Basal Ganglia in the Semantic Aspect of Language: an Interesting Finding in the Neuropsychological Assessment of Huntington's Disease

Omid Mohamadi* and Farhad Torabinezhad

Department of Speech and Language Pathology, School of Rehabilitation, Iran University of Medical Sciences, Iran

*Corresponding author: Omid Mohamadi, PhD Candidate, Department of Speech and Language Pathology, School of Rehabilitation, Iran University of Medical Sciences, Iran

Introduction

Huntington's disease is an inherited autosomal dominant degenerative CNS disorder. This disease usually begins by the fourth or fifth decade of life and half of the children of individuals with the gene are affected [1]. Pathologically, severe loss of neurons in the caudate nucleus and putamen, as well as diffuse cortical neuronal loss along with impaired activity of the striatum and its frontal lobe projection areas is evident [2,3]. A group of studies attributed language deficits in Huntington's disease only to non-linguistic impairments such as movement disorders, visuospatial impairments, slow retrieval processes, and constructional disability. Moreover, there is disagreement about whether Huntington's disease affects some language skills, such as semantic processing.

Case Description

The patient was a 64-year-old man with movement disorder and dysarthria, which was referred to the Speech Therapy Clinic of Hazrat Rasoul Hospital affiliated with Iran University of Medical Sciences. The level of education of the patient was PhD in political science and his job was to teach part-time at the university as well as high school management. The onset of obvious symptoms in this patient dates back to about 4 years and 7 months ago. At that time, he was not able to keep the names of his students and colleagues as before, and he experienced the first problems and long pauses during speech. Years ago, the patient's wife noticed sudden movements in the patient's limbs and occasional distractions, but these symptoms were so subtle and rare that the person seemed completely normal. The patient's family history indicated that his first-degree relatives had Huntington's disease. Because of the patient's personality traits and his refusal to accept the problem, his first visit to a neurologist was 4 years ago. In neurological and physical examinations, the most prominent clinical manifestation of the disease was choreiform movements in the limbs and face. In this case, the head MRI and genetic testing were performed only once. Brain imaging results showed mild brain atrophy with neurodegenerative disease. Based on this, doctors referred the patient for a Huntington's disease molecular diagnosis test. The results of a laboratory test with 98% sensitivity and accuracy showed that the patient had Huntington's disease. Therapeutic interventions prescribed to the patient included medication and rehabilitation. At present, the patient's medications are as follows: Sertraline 50, Amantadine 100, Depakene 500 mg, Olanzapine 5, and Risperidone 2 mg.

The patient's speech profile indicated high speech rate, inappropriate pauses, and articulation disorders. Consequently, the speech and laryngeal Alternating Motion Rates (AMRs) also had a significant reduction. Moreover, the perceptual characteristics of the monopitch, struggled and harshness voice along with the sudden arrests were quite obvious. The patient had dif-

Citation: Mohamadi O, Torabinezhad F (2020) The Broader Role of Basal Ganglia in the Semantic Aspect of Language: an Interesting Finding in the Neuropsychological Assessment of Huntington's Disease. Int J Neurodegener Dis 3:016. doi.org/10.23937/2643-4539/1710016
Accepted: May 30, 2020; Published: June 01, 2020
Copyright: © 2020 Mohamadi O, et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.
Huntington's disease, the patient's performance was impaired in skills such as attention, calculation, mental control, and memory. This finding is consistent with previous studies [5-8]. The patient's problems with copying 3D shapes (square and cylindrical shapes), assembling parts of an object, making designs with blocks similar to the examiner's pattern, and spatial recognition persuaded researchers to refer him to the occupational therapy service for more accurate assessments. Previous studies have acknowledged perceptual problems in people with Huntington's disease [9,10]. Progressive impairment of executive function in the early stages of Huntington's disease [8,11] and its association with caudate and insular atrophy [12] has been previously reported. Moreover, impaired executive function may account for many of the cognitive impairments in Huntington's disease, caused by striatal and corticostriatal involvement. Assessment of executive functions is considered as an indicator to monitor progression of disease [13]. The poor performance of the patient in tests such as GO-NO GO, FAS, Stroop Test, Digit Symbol, and Verbal abstraction indicated impairment to executive functions, which was consistent with previous research.

While the role of cortical structures in language processing has been well studied, the importance of subcortical structures such as basal ganglia in language processing has not been fully elucidated. In addition to the letter fluency, patient performance in the semantic or category fluency was also severely impaired. This reduction in verbal fluency is one of the word finding problems that seems to be related to semantic memory loss. The patient's problems with naming skills were both visually and auditory based. However, the patient was able to repeat the words immediately after examiner was intact. On the second day, based on the test instructions, the patient was shown a picture of each item to name, that is, the naming process was purely visually-based. The patient was able to name 33 items correctly, 13 items with semantic cues, and 4 items with phonetic cues. Accordingly, the naming errors were of both auditory and visual based. The test scores showed that the patient's naming disorder was moderate.

Discussion

As expected, due to the degenerative nature of Huntington's disease, the patient's performance was impaired in skills such as attention, calculation, mental control, and memory. This finding is consistent with previous studies [5-8]. The patient's problems with copying 3D shapes (square and cylindrical shapes), assembling parts of an object, making designs with blocks similar to the examiner's pattern, and spatial recognition persuaded researchers to refer him to the occupational therapy service for more accurate assessments. Previous studies have acknowledged perceptual problems in people with Huntington's disease [9,10]. Progressive impairment of executive function in the early stages of Huntington's disease [8,11] and its association with caudate and insular atrophy [12] has been previously reported. Moreover, impaired executive function may account for many of the cognitive impairments in Huntington's disease, caused by striatal and corticostriatal involvement. Assessment of executive functions is considered as an indicator to monitor progression of disease [13]. The poor performance of the patient in tests such as GO-NO GO, FAS, Stroop Test, Digit Symbol, and Verbal abstraction indicated impairment to executive functions, which was consistent with previous research.

While the role of cortical structures in language processing has been well studied, the importance of subcortical structures such as basal ganglia in language processing has not been fully elucidated. In addition to the letter fluency, patient performance in the semantic or category fluency was also severely impaired. This reduction in verbal fluency is one of the word finding problems that seems to be related to semantic memory loss. The patient's problems with naming skills were both visually and auditory based. However, the patient was able to repeat the words immediately after the examiner. These two findings could indicate an impairment of auditory and visual information processing skills in terms of their semantic properties. In other words, the patient may have difficulties in processing semantic representations of pictures and verbal descriptions of words. Studies of language disorders in Huntington's disease fall into two general categories. The findings of the current case study contrast with studies that only believed in the role of non-linguistic impairments for patients' language deficits. Wallesch and Fehrenbach (1988) attributed the patient's deficits in the picture naming and comprehension to dysarthria and their cognitive decline [14]. In this regard, Podoll, et al. also claimed that language impairments in Huntington's disease are the result of disorders such as dysarthria, visuospatial impairment, constructional disability, and chorea [15]. Hodges, et al. found that the naming errors in Huntington's disease were visually-based and a sign of impaired perceptual analysis. The same researchers acknowledged that phonemic processes remained relatively intact [16]. Some studies also reported that people with Huntington's disease show greater impairment in letter fluency rather than semantic fluency [17-19].
The discrepancy between the findings of the current case report and the studies of the first group is evident. The second group of studies pointed to a deeper role of these basal ganglia in language processing but there are still differences between them about the role of basal ganglia in semantic processing. In a study on mildly and moderately demented patients with Huntington’s disease, Smith, et al. reported a disruption in the system of spreading activation in a lexico-semantic network in a group [20]. In another study, people with Huntington’s disease were assessed by language tests. The researchers found that patients were impaired in lexico-semantic tasks [21]. Paraphasic errors and word-finding difficulty [22] and occasional production of neologisms and semantic errors [23] were some obvious problems in the spontaneous speech of people with Huntington’s disease. The present study also confirmed the impaired semantic processing of both auditory and visual information in a person with Huntington’s disease. On the other hand, there are studies that have refuted the damage to semantic processing in Huntington’s disease [24-26]. The various manifestations of Huntington’s disease at different stages of its development, the lack of consistency among the methods of examining the language skills of these patients and the use of different tests and diagnostic tools are among the causes of existing contradictions. Although this case report provides the lowest level of evidence, the role of basal ganglia in language processing seems to be broader than our previous knowledge. It is possible that the caudate nucleus and putamen, which are more involved in Huntington’s disease, play an important role in providing sufficient cognitive resources for semantic processing and facilitating access to its representations. Researchers suggest that interdisciplinary studies be designed with larger sample sizes to achieve stronger evidence.

Conflict of Interest Statement

The authors have no conflicts of interest to declare. No financial support was received for this study.

References

1. Caviness JN (2000) Huntington’s disease and other cho- reas. In: Adler CH, Ahlskog JE, (edn) Parkinson’s disease and movement disorders: Diagnosis and treatment guidelines for the practicing physician. Totowa, NJ: Humana Press.

2. Bartenstein P, Weindl A, Spiegel S, Boecker H, Wenzel R, et al. (1997) Central motor processing in Huntington’s disease: A PET study. Brain 120: 1553-1567.

3. Weeks RA, Ceballos-Baumann A, Piccini P, Boecker H, Harding AE, et al. (1997) Cortical control of movement in Huntington’s disease: A PET activation study. Brain 120: 1569-1578.

4. Critchley M (1979) The divine banquet of the brain and other essays. New York: Raven Press.

5. Rosser AE, Hodges JR (1994) The dementia rating scale in Alzheimer’s disease, Huntington’s disease and progressive supranuclear palsy. J Neurol 241: 531-536.

6. Pillon B, Deweer B, Agid Y, Dubois B (1993) Explicit memory in Alzheimer’s, Huntington’s, and Parkinson’s diseases. Arch Neurol 50: 374-379.

7. Brandt J, Spencer M, Folstein M (1988) The telephone interview for cognitive status. Neuropsychiatry Neuropsychol Behav Neurol 1: 111-117.

8. Ho AK, Sahakian BJ, Brown RG, Barker RA, Hodges JR, et al. (2003) Profile of cognitive progression in early Huntington’s disease. Neurology 61: 1702-1706.

9. Brouwers P, Cox C, Martin A, Chase T, Fedio P (1984) Differential perceptual-spatial impairment in Huntington’s and Alzheimer’s dementia. Arch Neurol 41: 1073-1076.

10. Lawrence AD, Watkins LH, Sahakian BJ, Hodges JR, Rob- bins TW (2000) Visual object and visuospatial cognition in Huntington’s disease: Implications for information processing in cortico-striatal circuits. Brain 123: 1349-1364.

11. Lawrence AD, Sahakian BJ, Hodges JR, Rosser AE, Lange KW, et al. (1996) Executive and mnemonic functions in early Huntington’s disease. Brain 119: 1633-1645.

12. Peinemann A, Schuller S, Pohl C, Jahn T, Weindl A, et al. (2005) Executive dysfunction in early stages of Huntington’s disease is associated with striatal and insular atrophy: A neuropsychological and voxel-based morphometric study. J Neurol Sci 239: 11-19.

13. Lenniere J, Decruyenaere M, Evers-Kiebooms G, Vandenbussche E, Dom R (2004) Cognitive changes in patients with Huntington’s disease (HD) and asymptomatic carriers of the HD mutation: A longitudinal follow-up study. J Neurol 251: 935-942.

14. Wallesch CW, Fehrenbach RA (1988) On the neurolinguis- tic nature of language abnormalities in Huntington’s dis- ease. J Neurol Neurosurg Psychiatry 51: 367-373.

15. Podoll K, Caspary P, Lange HW, Noth J (1988) Language functions in Huntington’s disease. Brain 11: 1475-1503.

16. Hodges JR, Salmon DP, Butters N (1991) The nature of the naming deficit in Alzheimer’s and Huntington’s disease. Brain 114: 1547-1558.

17. Hodges JR, Salmon DP, Butters N (1990) Differential impair- ment of semantic and episodic memory in Alzheimer’s and Huntington’s diseases: A controlled prospective study. J Neurol Neurosurg Psychiatry 53: 1089-1095.

18. Randolph C, Braun AR, Goldberg TE, Chase T (1993) Se- mantic fluency in Alzheimer’s, Parkinson’s, Huntington’s disease: Dissociation of storage and retrieval failures. Neuropsychology 7: 82-88.

19. Rosser AE, Hodges JR (1994) Initial letter and semantic category fluency in Alzheimer’s disease, Huntington’s dis- ease and progressive supranuclear palsy. J Neurol Neuro- surg Psychiatry 57: 1389-1394.

20. Smith S, Butters N, Lyon L, Granholm E (1988) Priming sem-antic relations in patients with Huntington’s disease. Brain Lang 33: 27-40.

21. Chenery HJ, Copland DA, Murdoch BE (2002) Complex language functions and subcortical mechanisms: Evidence from Huntington’s disease and patients with non-thalamic subcortical lesions. Int J Lang Commun Disord 37: 459-474.

22. Gordon WP, Illes J (1987) Neurolinguistic characteristics of language production in Huntington’s disease: A preliminary report. Brain and Language 31: 1-10.

23. Illes J (1989) Neurolinguistic features of spontaneous lan- guage production dissociate three forms of neurodegener-
24. Teichmann M, Dupoux E, Kouider S, Brugieres P, Boisse MF, et al. (2005) The role of the striatum in rule application: The model of Huntington's Disease at early stage. Brain 128: 1155-1167.

25. Teichmann M, Dupoux E, Kouider S, Bachoud-Levi AC (2006) The role of the striatum in processing language rules: Evidence from word perception in Huntington's Disease. J Cogn Neurosci 18: 1555-1569.

26. Teichmann M, Dupoux E, Cesaro P, Bachoud-Levi AC (2008) The role of the striatum in sentence processing: Evidence from a priming study in early stages of Huntington's disease. Neuropsychologia 46: 174-185.