



CASE REPORT

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

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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 mon-pitch, struggled and harshness voice along with the sudden arrests were quite obvious. The patient had dif-



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faculty in oral preparation and transfer stages. One of the family's complaints was that: "The patient does not chew the bits well and there is always some food left in the mouth."

Over the last few years the patient has had mental problems such as distractions and memory deficits. He was fully oriented, but there was severe impairment in the functions such as writing a book that needed to set goals, organize, plan, execute, monitor steps, and problem solving. The tests used in the neuropsychological evaluation section were: Wechsler Memory Test, Persian Picture Naming Battery, Patient's Mini-Mental Status Examination (MMSE), FAS test, category fluency test, Go-No Go test, Stroop Test, Verbal Abstraction Test and Digit Symbol Test. The results of semantic assessments are the main discussion of this case report; therefore they will be explained in more detail.

In the semantic assessments section, the researchers used FAS test, category fluency test, and Persian Picture Naming Battery. The FAS test is actually a test of mental flexibility and is used to probe executive function while the category fluency test assesses the organization of the semantic network [4]. The researchers performed each test twice in two consecutive days. The results showed that the patient, in addition to weakness in letter fluency skills, also had significant problems in category fluency. In other words, the semantic aspect of language was also exposed to obvious impairment.

In order to further evaluate the semantic aspect of language, the research team decided to perform the Persian Picture Naming Battery in both auditory and visually forms. This test consisted of 50 color images of different classes such as fruits, objects, animals and nature. The patient's ability to repeat words immediately after examiner was intact. On the first day, the examiner provided verbal descriptions of the semantic and functional characteristics of each item, and then asked the patient to name it without seeing the picture. In other words, the only way for the patient to respond correctly was through the processing of auditory, phonological, and lexical information to access the semantic system. Out of a total of 50 items provided, the patient was able to name 31 items correctly and eight items with the phonetic cues from the examiner (first letter of the word). The patient had a semantic paraphasia in seven items and could not answer for the remaining 4 items. 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 sub-cortical 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 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.

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