

A Case of Oral Spelling Behavior: Another Environmental Dependency Syndrome

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Objective: To present the case of a woman with a probable vascular dementia and a unique oral spelling behavior (OSB). Our aim was to better characterize the mechanisms underlying OSB in this patient with advanced cognitive and noncognitive deficits.

Background: Different kind of language deficits have been described in dementias and other related diseases. Other nonaphasic acquired communication disorders include a heterogeneous group of verbal impairment that are mainly due to attentional and executive impairments.

Methods: We describe the neurolinguistic and neuropsychologic analysis of an 88-year-old woman with spontaneous OSB. Computed tomography-scan at the time of the clinical analysis is also presented.

Results: OSB occurred preferentially in spontaneous discourse, manifested in other language tasks, concerned all lexical categories, and was not accompanied by spelling deficit although language was not intact and disclosed foreign accent syndrome. Environmental dependency syndrome or utilization behavior was prominent and concerned various sensorimotor activities.

Conclusions: We postulate that OSB was not secondary to a linguistic or more specifically to a spelling deficit but to another variant of environmental dependency syndrome and probably associated with a cortico-subcortical lesion including the left prefrontal region.

Key Words: spelling, foreign accent syndrome, environmental dependency syndrome, dementia, aphasia, utilization behavior

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Although language impairments are obviously more difficult to interpret than in focal lesion studies, patients with dementias and related disorders present with various kind of anomias, aphasias, surface dyslexia, and semantic deficits, depending on localization, type,

and extension of the pathology.¹ Other nonaphasic acquired communication disorders have been described and include a heterogeneous group of verbal impairment that does not imply predominant semantic, syntax, or grammar deficits. For example, the so-called empty speech production mostly encountered in Alzheimer disease, is mainly due to attentional and executive impairments.^{2,3} We present a case with a unique spontaneous oral spelling behavior (OSB) that occurred in the course of a probable vascular dementia. Our aim was to better characterize the mechanisms underlying OSB in this patient with advanced cognitive and noncognitive deficits.

CASE REPORT

A right-handed 88-year-old woman with 18 years of education, polyglot, known for high blood pressure and already diagnosed a year ago with probable vascular dementia was hospitalized for agitation and aggravation of cognitive impairment. On neurologic examination, visual field was complete, cranial nerves were normal. There were bilateral palmomental, suck and grasp reflexes. She had slight bilateral bradykinesia, paratonia, a right Babinski sign, and a frontal gait with small steps but no stooped posture or arm swing disturbances. She was not collaborative, at times resistant, irritable, agitated, unaware of her deficits, and suspicious but without hallucinations or well-formed delusions. During the following weeks, the whole staff noticed recurrent episodes whereas she would spontaneously spell out loud words she heard. We called this behavior OSB.

At the time of clinical assessment, her brain computed tomography-scan showed a profound subcortical vascular leukoencephalopathy with typical sparing of U-fibers that connect adjacent cortico-cortical regions. The lesions were confluent and pronounced in bilateral prefrontal regions, particularly on the left (Fig. 1).

RESULTS

Mini-mental state examination⁴ was 5/30 and Dementia Rating Scale⁵ 26/144 but those tests were unequivocally lowered by disruptive behavior and lack of collaboration. She was, however, not severely demented. This was corroborated by the fact she could mention recent political facts or main biographical events, correctly manipulate real objects, imitate gestures, recognize objects, pictures or texts, and that basic activities of daily living were preserved. Nevertheless, for the same reasons mentioned above, any other standardized psychometric tests could not be performed.

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FIGURE 1. Brain computed tomography-scan showing important cortico-subcortical lesions of vascular origin in the prefrontal regions and more pronounced on the left side (arrow).

Neuropsychologic and neurolinguistic assessments were multiple and limited in time. She had a profound loss of insight. Orientation was partial. Language was fluent. Disruption in segmental and prosodic elements, especially changes in the articulation of words and alterations in syllable structure were perceived as a Japanese accent by listeners including nurses, neuropsychologists, medical staff, and family members. Denomination and auditory comprehension tasks were impaired, but most of semantic aspects were intact as shown with the Laiacona battery,⁶ a multiple choice questionnaire measuring semantic knowledge of different kind of categories, where she obtained 80% of correct responses. Repetition was partially preserved for words, nonwords, and short sentences. Orally, she could produce very sophisticated and complex sentences with highly preserved syntax. Written productions demonstrated preservation of orthographic processes. Under dictation, she wrote correct words with some semantic paraphasias. Oral text reading disclosed semantic errors, morphophonological paralexias, omissions of words or lines, and frequent tendency to vocalize punctuation marks. She could perfectly spell polysyllabic words and nonwords, and identify them when spelled out loud. OSB occurred preferentially in spontaneous discourse. Most of the time she would first pronounce the integral word, and then spell it without error. OSB manifested in other language tasks such as repetition, confrontation naming, writing to dictation, and text or isolated word reading and concerned all lexical categories (proper names, common nouns, grammatical words, and nonwords).

She manifested frequent utilization behaviors with all kind of objects placed in her peripersonal space (handbag, glasses, chocolate bar that she would eat, her son's business card that she would read and comment). Any written stimulus would activate oral reading behavior, including restroom panel, books, or newspapers. In addition, she manifested some abnormal repetitive motor behaviors that would last a few minutes and without any reason such as continuously going to the restroom or putting her shoe on and off.

OSB did last during her stay at the hospital then she was discharged and went home with full paramedical staff. A year later she was hospitalized again in the same unit. Although she presented at that time with an

important decrease in spontaneous word fluency and global autonomy, OSB persisted but was less frequent.

DISCUSSION

Oral spelling is a neurolinguistic skill that is developed during the writing learning process either in childhood or when we acquire a new language. Mechanisms that underlie spelling are assumed to hold graphemic representation of a word for conversion into specific letter shapes (for written spelling) or letter names (for oral spelling).⁷ Name letter codes and allographic codes are translated by specific motor programs; speech code for oral spelling and graphemic motor pattern for written spelling into an articulatory and a graphic code for realization of oral and written output. Relative preservation of oral spelling but impairment of written spelling and the opposite profile⁸ or preserved oral spelling without semantics in the context of a vascular dementia⁹ have been described. We hypothesize that OSB was not secondary to a spelling impairment per se, but to an environmental dependency syndrome (EDS) because of the following arguments: (1) OSB occurred spontaneously in different language tasks (writing to dictation, reading, ...) and concerned regular or nonregular words in all grammatical categories such as words, nonwords, verbs, ... (2) The function of OSB was not to give a phonological form or generate semantic access such as in Snowden's patient with severe anomia but preserved oral spelling.¹⁰ Our patient did not have a spelling deficit because she could spontaneously produce the entire appropriate word before she would spell it out loud. (3) She presented with numerous cross-language interferences and code switching when she spoke in French. These disturbances are common in bilingual or multilingual patients with dementia and are commonly attributed to parallel activation of concurrent languages and subsequent loss of inhibition of one language over the other.¹¹ (4) OSB was accompanied by the foreign accent syndrome (FAS), or pseudo-accent syndrome. FAS is not due to a compensation or adaptation to a voice or speech problem but linked to a deficit in speech production and, not surprisingly, most of the time associated with a cortical or subcortical lesion in the dominant hemisphere.¹² (5) She exhibited symptoms similar to EDS and utilization behavior^{13,14} or hyperphasia,¹⁵ loss of behavioral control to read silently,¹⁶ and verbal response to next patient question.¹⁷ In our patient, the extensive utilization behavior concerned various sensorimotor activities, language such as reading out loud and particularly spelling out loud. (6) The presence of concomitant EDS and FAS, and its association with cortico-subcortical lesions in the prefrontal regions, particularly on the left was certainly not incidental.

To conclude, a unique type of human EDS explained our OSB. That original pattern of deficits might have theoretical relevance for the debate on linguistic versus nonlinguistic misproductions in the human brain and in dementia.

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