



Slowly progressive aphasia: a four-year follow-up study

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Received 14 October 1999; received in revised form 18 September 2000; accepted 20 December 2000

Abstract

This paper reports the long-term follow-up of GC, a patient with primary progressive aphasia of the fluent type. GC presented at onset with an anomia characterized by sparing of first letter knowledge, that applied mainly to proper names and living categories. No semantic deficits were observed in the first stage of the disease, and MRI showed a left temporal lobe atrophy with a gradient from the pole to the posterior regions, the latter being less involved. We now report the clinical evolution of GC from the 2nd to the 4th year of disease. As the disease progressed, the anomia became more severe and the phenomenon of first letter sparing was no longer detectable. Also semantic knowledge was gradually affected and, eventually, was dramatically lost. However, no other cognitive deficits were seen at the last examination. By that time, the temporal atrophy shown by MRI was bilateral, although still more evident on the left side. © 2001 Elsevier Science Ltd. All rights reserved.

Keywords: Semantic dementia; Temporal lobe atrophy

1. Introduction

Primary progressive aphasia (PPA) is a gradual dissolution of language, being the only salient feature that compromises life for at least 2 years [14]. The disease is associated with left perisylvian atrophy. Hodges et al. [9] use interchangeably the terms semantic dementia or progressive fluent aphasia, considering the fluent form of progressive aphasia to overlap with semantic dementia. In this condition there is a progressive atrophy of the anterior temporal lobes, particularly of the inferior temporal gyri [21].

There are a few detailed follow-up studies on PPA. Mazzone et al. [13] described a woman who, over a period of 9 years, showed no signs of dementia and had only a language disturbance. The aphasia was apparently a non fluent form, and the evolution was toward a progressive reduction of words, mainly verbs, and a tendency to produce very short sentences. A 14-year follow-up study is reported by Schwarz et al. [20]. The

profile of this case was similar to that shown by patients with semantic dementia. The follow-up of RC, a very similar patient to the one we have described [17], is reported by Kertesz et al. [10]. The focus of their paper, however, is to demonstrate the selective impairment of the central semantic system, suggesting a fractionation of semantic memory: this was demonstrated by the relative preservation of visual categorization. Other patients included in group studies are reported at different intervals from the onset of the disease, but a detailed review of the literature is out of the aim of this study.

In this paper, we report the follow-up of GC, a previously described patient [17], whose first symptom was a proper name anomia. A particular feature of this patient was a sparing of initial letter knowledge, while other characteristics of the unretrievable words were not equally available. Our aim in the follow-up study was to assess:

1. the general neuropsychological evolution;
2. the evolution of naming impairment;
3. the evolution of the first letter sparing phenomenon; and
4. the possible evolution of the formerly spared semantic knowledge, including the related anatomo-functional aspects.

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2. Case report

2.1. April 95–May 96

When first seen in April 1995, GC was a 59 year-old right-handed woman, with 5 years of education, who suffered a progressive difficulty in retrieving proper names, including those of her relatives. Since January 96, the naming impairment also began to affect common names, without a discrepancy between animate and inanimate stimuli. The most impressive feature of this patient was a first letter sparing, which mainly characterized proper names, but was present even for animate category stimuli. GC produced the spared first letter by giving the name of the letter (e.g. 'emme' for 'M'). In Italian orthographic–to-phonological mapping is largely consistent, apart from a few exceptions such as 'C', but even in words starting with 'C' that could have two different phonological forms, GC always gave the name of the letter and then started to pronounce the correct sound, often producing nonwords that were very similar in sound to the target. Semantic knowledge about the people and objects she could not name was perfectly preserved (for a detailed clinical history and discussion of this case from April 1995 to May 1996 see Ref. [17]).

2.2. October 96–October 97

A summary of the results obtained in the general neuropsychological examination is given in Tables 1 and 2. Episodic memory was not examined due to the language difficulties of the patient.

From October 1996 to October 1997, GC showed a mild but progressive deterioration of naming; there was a category effect on first letter sparing, which was more evident for living than for non living categories.

Naming performance from October 96 until October 97 is reported in Fig. 1. Auditory–verbal comprehension (see Fig. 2) and semantic knowledge remained normal until October 97, when a verbal comprehension (an 80-item pointing to picture test, in which the subject has to point to the target among four foils of the same category) appeared to be slightly under the cut-off for living categories (83%, normal value > 87%). Non living categories were still in the normal range (86%, normal value > 83%).

The pattern of GC's impairment changed substantially in December 1998, so a description of her behaviour at that time is given in greater detail.

2.3. December 1998

At this stage GC seemed less concerned about her

illness than she had been before. However, she reported that she was no longer able to go shopping, because she could not access the names of vegetables. She had started to buy packages in which all vegetables were mixed together. In this way, if her husband had asked for celery, she was sure that among all the different components, this one was included. She was unable to retrieve not only the name, but also what that particular thing looked like, and this made it difficult to cook: if she had to follow a recipe, and this included for example an onion, she could not decide what an onion was. This difficulty emerged also with inanimate categories: during the testing session, the examiner asked for a hammer, but the patient did not know what a hammer was. When she was told that it was the instrument to insert nails, she asked what a nail was. The same clinical picture is described by Kertesz et al. [10] in their patient RC. Interestingly, even in their patient the first symptom was a proper name anomia, 2 years before the appearance of a more general word finding impairment. At the same time, GC did not show episodic memory deficits and no prospective memory impairment was reported. The appointment was arranged with the patient herself, she came by train alone to the hospital from another town, she remembered the examiner perfectly, she was able to find the testing room by herself and she recognized the tests that she had previously performed.

Table 1

General assessment: the results obtained in April 95 are also reported for comparison

	April 95	October 97	December 98	Normal score
Non verbal intelligence				
Rven PM 47 Matrices	36	36	32.5	≥ 18
Perception				
Benton Line Orientation	28	27	23	≥ 17
Benton Face Recognition	50	49	42	≥ 39
Memory				
Corsi span	4.25	4	4	≥ 3.50
Language				
Token Test	32	26 ^a	25 ^a	≥ 29
Praxis				
Oral praxis	20	n.e. ^b	20	≥ 17
Ideomotor apraxia	68	n.e. ^b	65	≥ 53

^a Pathological.

^b Not examined.

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