Semantic amnesia without dementia: documentation of a case

Rusconi M.L.1, Zago S.2, Basso A.2

1 Dipartimento di Psicologia Generale, Università di Padova; 2 Istituto di Clinica Neurologica, Università di Milano, Italy.

We described the case of a patient affected by a progressive semantic memory disorder associated with prevalent temporal lobe atrophy. This deficit seems to be “pure” in the sense that it has not been found to overlap with other cognitive deficits (intellectual, linguistic, perceptual, visuo-spatial etc.) for a long time. Furthermore, despite his impaired semantic knowledge, the autobiographical memory of the patient was largely intact. This case therefore represents a form of “semantic amnesia” without dementia, and supports the hypothesis that there is a partial distinction between “semantic” and “episodic” memory.

Key Words: Semantic amnesia — Semantic memory — Episodic memory.

Introduction

A number of clinical descriptions of patients showing a progressive decline in only some cognitive functions, in the absence of widespread cognitive deterioration, have recently been published. These are interesting cases because, unlike demented patients, they show a focal neuropsychological presentation and progression over time, which is presumably due to their underlying anatomical locus. Some of them are represented by the syndrome of “slowly progressive aphasia” [6, 7] in which patients experience a gradual deterioration of language abilities without other cognitive and behavioural disturbances, with the cerebral degeneration predominantly involving the left perisylvian region. Case reports of isolated and progressive dysfunction confined to other cognitive abilities have also been published: for example, visual perception impairments or progressive generalized apraxia due to a degenerative process mainly involving the posterior regions [3], and a selective semantic deficit due to a focal cerebral degeneration predominantly affecting the temporal lobes [5, 10, 11]. This last syndrome is characterized by a progressive and selective breakdown of semantic knowledge that contrasts with a widespread preservation of the phonology, grammar and syntax of spoken language, autobiographical and episodic memory, as well as other cognitive functions, such as non-verbal memory, and perceptual and visuo-spatial abilities. The deficit seems to involve the component of long-term memory called “semantic memory”, which is recognised as being the repository of knowledge concerning concepts and facts, as well as words and their meaning [12].

The first well-documented cases of selective semantic memory deficit were reported by Warrington [13], who studied three patients with cerebral atrophy and progressively impaired lexical-semantic knowledge. Careful neuropsychological examination revealed a loss of verbal vocabulary and an impaired knowledge of animals and objects, in the context of well-preserved linguistic, visuo-perceptual and general intellectual functions. Some years later, similar observations were made by Schwartz et al. [9] in a single case and more recently, various studies have contributed towards improving our understanding of both the neuropsychological and anatomical aspects of this syndrome [5, 10, 11].

In these last cases the semantic disorder was selective, unrelated to the type of stimulus or input modality, and characterized by severe anomia and impaired spoken and written single-word comprehension. The other components of speech production (notably syntax and phonology) and other cognitive abilities such as autobiographical and episodic memory, and perceptual and spatial skills, were largely unaffected.

Snowden et al. [10] used the term “semantic dementia” to denote the syndrome, but we prefer the term “semantic amnesia” used by De Renzi et al. [4] in their description of a patient with a semantic deficit, because it underlines the fact that the disorder more specifically concerns the memory system than the widespread cognitive involvement underlying the slowly progressive evolution from semantic amnesia to dementia (see Patterson and Hodges) [8].

It would seem that the most frequent pathological pattern is a focal and often asymmetric temporal lobe atrophy which, at least initially, may be confined to the left hemisphere. In some cases the locus also involves the
frontal and temporo-parietal areas [5, 10], but CT and/or MRI imaging and functional brain imaging (SPECT/PET) have shown that it is always predominant in the lateral temporal regions, see Patterson and Hodges [8]. We have recently had the opportunity of studying and following up for two years a patient in whom the neuropsychological data were indicative of a selective impairment of semantic knowledge.

Case report and neuropsychological follow-up

M.U., a 60-year-old right-handed ex-manager with eight years of schooling, first visited a neurologist in June 1992, one year after the onset of a slowly progressive difficulty in finding names for objects. His medical history was unremarkable except for gastric ulcer; his family history was negative for dementia and other neurological diseases. The results of neurological examinations were normal and EEG revealed no abnormalities. CT scans and MRI showed atrophy of the temporal lobes with a prevalent enlargement of the left temporal horn and mild frontal lobe atrophy (Figure 1).

The patient underwent a first neuropsychological test in October 1992 (the results are reported in Table 1). He was alert, cooperative, and fully oriented in time and place. No behavioral abnormalities were observed. He retained good insight into his naming disorder and was perfectly capable of reporting his personal history. Spontaneous speech was fluent and well articulated; its syntactic structure was complex but characterized by word-finding difficulties. The comprehension of spoken commands, and reading and repetition abilities were normal. However, the ability to name objects and people was severely impaired; the patient recognized objects but used paraphrases to describe them. To remember the names of people, he resorted to numbering the letters of the alphabet: for example, the number “5” corresponds to the fifth letter “E”, “10” to “L” (in the Italian alphabet), and so on until he had reconstructed the name of his wife ELSA. Written naming was poor due to the word-finding disorder, and naming by verbal description and other oral fluency task performances were very poor. Verbal memory evaluation indicated normal short-term memory, with defective learning and recall (the latter can be explained in terms of a loss of receptive and expressive vocabulary). The patient's performance in spatial memory tests was normal, and detailed examination of motor and constructional praxis revealed no defect. His visuo-spatial and calculation abilities were also preserved.

Although no standardized tests evaluating semantic memory were performed, during the first examination, a mild degree of semantic impairment could be inferred from some hesitations concerning the definition and comprehension of single words, and in the word-to-picture matching task; however, at this stage, the predominant feature was anomia, meaning that the patient was capable of identifying but not of naming the stimuli (for example: “Egyptian building” for “pyramid”; “it falls from the trees” for “gland”).

Evaluations were also made in April and October 1993, and in July 1994 (the results are summarized in Table 1). The first and second neuropsychological re-examinations did not reveal any further deterioration but confirmed the severely deficient lexical-semantic retrieval. For the examination performed in October 1993, a formal test of semantic memory was devised: this was the object decision test, in which the patient had to decide whether a given picture represented a real or non-existent object. The test consisted of 84 pictures of objects and animals, half of which represented real things. The score of the patient was 31/84 (36.9%), whereas those of five control subjects ranged from 77 to 81. Furthermore, during an informal test of single word comprehension, our patient showed a profound deficit in semantic knowledge retrieval. Some (spoken and written) words had a familiar sound but were lacking in semantic content (for example, “grapes” ... “I recognize the word but I don't know what it is... it should be a fruit”). At this stage, the loss of fine-grained subordinate information, with the preservation of superordinate broad categorical information was detectable, although no category-specific effects were observed.

During the neuropsychological assessment performed in July 1994 (two years after the first), the patient was still well oriented in time and place but showed a mild decline in cognitive functions (Table 1). Furthermore, a series of