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Preservation of autobiographical memory in a case of pure progressive amnesia

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Abstract

G.D., a 79 year-old female, presents with a severe and slowly progressive amnesia although she remains entirely independent in daily life and is perfectly well spatially oriented. Her amnesia is relatively isolated and her deficit does not embrace other cognitive domains. G.D. underwent extensive neuropsychological evaluation including language, executive functions, perceptual, and memory tests. Based on clinical observation, the purpose of this study was to determine whether there was a dissociation between her autobiographical and semantic memory. Results point out a severely degraded semantic knowledge of famous public events and persons while autobiographical memory of personally experienced and relevant information remains intact. Results from this study and from previous studies seem to suggest that relative sparing of hippocampal structures may be related to the preservation of autobiographical memory.

1. Introduction

The medial temporal lobe is a large area that includes the hippocampal formation, subhippocampal structures such as the entorhinal, perirhinal, parahippocampic cortices, and the amygdaloid complex. The role of these various structures remains a matter of debate. However, lesions to the hippocampal or subhippocampal cortices are thought to result in different memory impairments (Aggleton & Brown, 1999). For instance, in a recent study by Vargua-Kadem et al. (1997), patients with selective hippocampal pathology and preserved subhippocampal region presented with a dissociation between impaired recall of everyday autobiographical memory and a preserved ability to learn new semantic facts. In contrast, patients with semantic dementia present with a progressive and selective deterioration of semantic memory (general knowledge about the world) while autobiographical memory remains largely intact (Hodges, Patterson, Oxbury, & Funnel, 1992; Snowden, Griffiths, & Neary, 1994, 1995, 1996). In the early stages, semantic dementia often results from an atrophy of bilateral anterior and inferior temporal structures with a relative preservation of the hippocampal formation.

Progressive focal cortical atrophies are degenerative processes which are characterised in the early stages by a selective impairment in one cognitive domain. Focal cortical syndromes affect language (primary progressive aphasia), speech (progressive dysarthria), semantic memory (semantic dementia), episodic memory (pure progressive amnesia), vision (progressive visuospatial and visuoperceptual disorders) or gesture (progressive apraxia) (Didic, Felician, Ceccaldi, & Poncet, 1999). The neuropathological processes inherent to these syndromes are heterogeneous: they include non-specific degenerative lesions, Pick's disease lesions and Alzheimer's disease lesions. Pure progressive amnesia, a rare form of focal cortical atrophy, is characterised by the gradual onset of an isolated amnestic syndrome in the relative absence of dysfunction in other domains of cognition at least during the first 2 years of the disease (Didic, Ali Cherif, Gambarelli, Poncet, & Boudouresques, 1998). Such patients typically present with an impaired deficit in recalling context-free semantic

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knowledge, maintain a striking independence in everyday life and show preserved spatial memory. Bilateral focal atrophy of the subhippocampal region with relative preservation of the hippocampi was found on MRI in one such patient (Didic et al., submitted).

In the present article, we report the case of such a patient with pure progressive amnesia of undetermined aetiology who shows a dissociation between her severely impaired recall of semantic facts and her strikingly preserved ability to recall spatially and temporally determined autobiographical facts. For instance, during a first general neuropsychological assessment, she could describe with great precision moments she had lived during the Second World War (she was in her early twenties at the time) yet could not provide very basic semantic facts about the war such as who was Adolf Hitler or who was persecuted during that war. In the present study, patient G.D. underwent detailed neuropsychological testing evaluating her semantic and autobiographical memory.

2. Methods

2.1. Case history

G.D., a 79 year-old retired florist, was referred to the Service de Neurologie et Neuropsychologie de la Timone for increasing memory complaints. According to her husband, her memory problems had appeared more than 2 years ago and had been gradually but slowly worsening since then. Changes in behaviour were absent. G.D. is completely independent in daily life. She still manages her household chores efficiently and continues to pursue her hobbies such as choir, fishing, and gardening. G.D. is well oriented in space and does not get lost in her own environment nor in new environments. She still drives on her own and is able to find her way from her hometown to the Timone Hospital more than an hour away without any help.

2.2. General neuropsychological evaluation

G.D. underwent extensive neuropsychological evaluation including tests of language, memory, executive functioning, visuoperceptual, and visuospatial abilities. F.G scored 19/30 on the Mini-Mental State Examination (Folstein, 1975).

2.3. Memory

On the Weschler Memory Scale Revised, her performance was severely impaired. She obtained a score of 73 on verbal memory (percentile 5), 65 on visual memory (percentile 1), 56 on general memory (percentile 0.1), 95 on attention/concentration (percentile 37), and 76 on delayed recall (percentile 5). Her digit span was 5 forward and 4 backward. Her score on the Grober et Buschke Test of episodic memory was also severly impaired: she scored 10/16 on cued immediate recall (control mean = 15.8, $SD = 0.49$), 1/16 on immediate recall I (control mean = 11.3, $SD = 1.2$), 4/16 on immediate recall II (control mean = 13.4, $SD = 1.6$), 1/16 on immediate recall III (control mean = 14.4, $SD = 1.9$), 3/16 on delayed recall (control mean = 14.8, $SD = 1.6$), and 12/16 on recognition (control mean = 15.9, $SD = 1.6$).

2.4. Language

Overall, her language was well preserved. She scored 74/80 on the DO80 naming test (control mean = 74.56, $SD = 4.78$); she produced 29 words in 2 min on the lexical fluency test (letter P: control mean = 14.71, $SD = 7.56$), 21 names of animals (control mean = 25.78, $SD = 5.32$), and 16 names of vegetables (control mean- = 17.21,$SD = 4.97$) on the category fluency test. In spontaneous speech, she had occasional but slight word finding difficulties. Comprehension was intact. Her performance in reading aloud indicates a pattern of surface dyslexia: she read 100% of low, mid, and high frequency regular words correctly but scored 58.8% on low frequency irregular words, 76.2% on mid frequency irregular words, and 100% on high frequency irregular words.

2.5. Visuoperceptual and visuospatial abilities

G.D. scored 8/10 on the *Identical figures* subtest and 29/30 on the *Entangled figures* subtest of the Protocole d'évaluation des gnosies visuelles (Protocole Montréal-Toulouse, 1985). Furthermore, she scored 19/20 on the *Object decision* subtest and 20/20 on the *Position discrimination* subtest of the Visuoperceptual and space perception battery (Warrington & James, 1985). She scored 43 on the Benton Face Recognition Test (normal range = 41–54) (Benton & Van Allen, 1968). When asked to remember a 150 m trajectory in the hallways of the Timone hospital, G.D. was perfectly able to complete the same trajectory without any mistakes 5 min, 15 min, and 1 h after having initially learned that trajectory, thus confirming her well-preserved ability to orientate herself in a new environment.

2.6. Executive functioning

G.D. was able to obtain six categories on the short version of Wisconsin Card Sorting Test, although she made 10 errors of which 7 perseverations. However, these errors clearly reflect an underlying working memory impairment (she kept forgetting the previous

category) rather than a difficulty in switching cognitive strategies.

2.7. Materials and procedure

In order to test G.D.'s autobiographical memory, we used the Kopelman autobiographical Interview (Kopelman, Wilson, & Baddley, 1990), a semi-structured interview which aims at measuring a person's aptitude to recall specific temporally and spatially situated autobiographical events (*autobiographical episodic memory*) and personally relevant semantic information (*personal semantics*) throughout three different life periods: childhood, early adulthood and recent life. We also added a mid-adulthood period that covered a gap of several decades for G.D., and recent life was split into two parts: before and after onset of the disease (past 2 years). This was due to the fact that her episodic memory had considerably worsened since onset of the disease. In order to test a specific aspect of her semantic memory, we used the *Protocole de Mémoire Evenementielle*.

(Thomas-Antérion, Laurent, Lemesle, Laporte, & Michel, 1994), a test that measures an individual's knowledge of famous public events and figures throughout the decades 1920–1990s. Famous events in this test are presented in both the visual and verbal modalities.

3. Results

Overall, G.D. scored on the Kopelman autobiographical Interview remained within normal range. G.D. she scored 100% when she was required to recall specific incidents that had occurred in her childhood, early, and mid-adulthood and recent life before onset of the illness. Her performance dropped to 61.9%, however, when she had to recollect specific incidents that had occurred since the beginning of her illness. When asked to provide personal context-free information such as the names or birth dates of past family members, friends or teachers (personal semantics), her performance was similar to that of episodic autobiographical memory. Her husband, who attended this part of the evaluation, was able to verify and judge as accurate most of the answers she had provided. Her performance was within normal range except for recent life prior to onset of her illness. See Figs. 1A and B for details.

In contrast with G.D.'s preserved performance on the Kopelman test of autobiographical memory, her performance was significantly impaired when compared to age-matched control subjects in both the visual and verbal modalities on a test of semantic knowledge of famous public events and figures (Protocole de mémoire évenementielle, Thomas-Antérion et al., 1994). G.D.

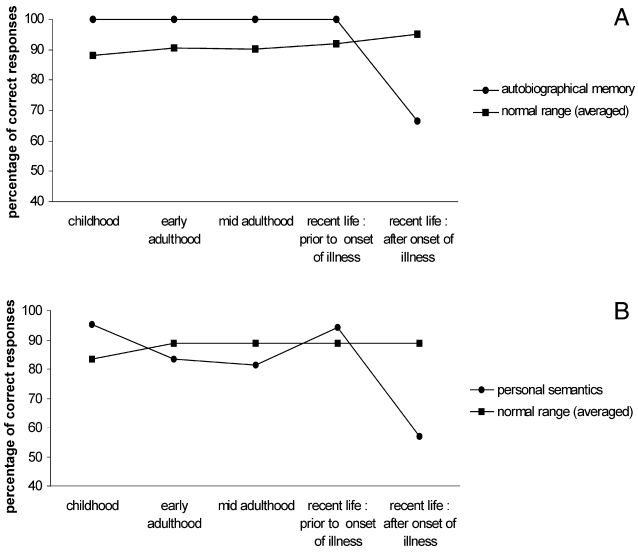

Fig. 1. (A) G.D.'s episodic autobiographical memory. (B) G.D.'s personal semantic memory.

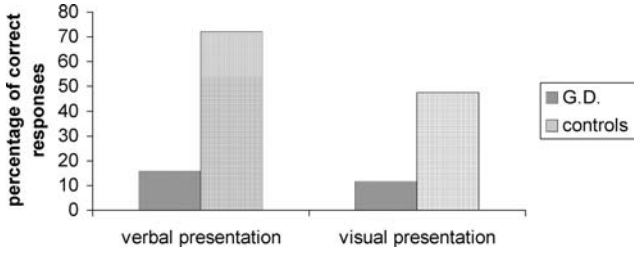

Fig. 2. G.D.'s performance on a test of semantic knowledge of famous public events.

scored 15.7% correct answers in the verbal modality (mean score = 72.2) and 11.7% correct answers in the visual modality (mean score = 47.5%). See Fig. 2 for results.

4. Discussion

G.D. has a slowly progressive amnesia in the absence of any significant impairments affecting her other mental functions. Although her performance on the WMS-R was extremely poor (often inferior to percentile 1), G.D. remains remarkably independent in everyday life and finds her way in her environment. Upon more detailed testing of her amnestic syndrome, it becomes evident that she sustains normal memory for personally experienced and personally relevant autobiographical events and facts while more general context-free semantic information such as knowledge of famous public events and persons is severely degraded. This pattern of dissociation between impaired semantic memory and preserved autobiographical memory is a characteristic feature of semantic dementia (Hodges et al., 1992;

Snowden et al., 1994, 1995, 1996) but to our knowledge remains undocumented in pure progressive amnesia. Such a dissociation may express the relative sparing of hippocampal regions, which are assumed to play a key role in episodic autobiographical memory. Contrary to patients suffering from semantic dementia, however, G.D.'s selective semantic loss of knowledge of famous public events and figures does not appear to extend to a more general semantic impairment. Her word and sentence comprehension is normal, her ability to name pictures and real objects is normal (real object naming was informally tested), and she does not present the important word-finding difficulties that are often found in semantic dementia.

SPECT scanning of G.D. indicates a predominantly left-lateralised hypofixation signal in the medial temporal lobe. MRI scans, although performed ten months after neuropsychological evaluation and indicating an already more extensive atrophy of the brain, involved predominantly bilateral subhippocampal and inferior temporal lobe structures. These clinical and anatomical results, as well as previous results obtained in patients with semantic dementia and pure progressive amnesia, confirm the view that autobiographical episodic memory and semantic memory may be underscored by independent yet interactive neural networks within the medial temporal and inferior temporal regions.

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