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Face–name repetition priming in semantic dementia: A case report

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ABSTRACT

Repetition priming (RP) has been employed as a measure of implicit processing in patients suffering from a breakdown of semantic memory, as in the case of semantic dementia (SD), a subtype of frontotemporal lobar degeneration (FTLD). Here, we investigated face–name representation in a case of SD using a paradigm of within- and cross-domain repetition priming. Compared to ten healthy participants, SD patient did not show any facilitation when a famous name was primed by its own face (cross-domain) or when the prime was the same proper name (within-domain). Results are discussed within the hypothesis of a degradation of face and name representation, one of the most consistent accounts explaining semantic deficits in SD.

1. Introduction

Famous face recognition and proper naming impairments are neuropsychological deficits occurring following a wide range of brain damage types, including stroke, traumatic brain injury and neurodegenerative disease (Bauer, 2003; De Haan, 2000; Semenza, Mondini, Borgo, Pasini, & Sgaramella, 2003; Semenza, Mondini, & Zettini, 1995; Werheid & Clare, 2007). When, in dementia, impairments with regard to famous faces and names occur, these are characterised by an insidious onset and a progressive manifestation. In particular, these deficits are common in patients affected by semantic dementia (SD), a clinical subtype of frontotemporal lobar degeneration (FTLD) (i.e., Snowden, Thompson, & Neary, 2004). The diagnostic label of FTLD encompasses a number of heterogeneous clinical manifestations, in which different patterns of impairment, involving linguistic processing, executive functions and action organisation, reflect the location of the underlying pathology (Libon et al., 2007). SD is traditionally the language variants of FTLD and is characterised by loss of word meaning and impaired language comprehension (Gorno-Tempini et al., 2004; Hodges, Patterson, Oxbury, & Funnell, 1992).

SD is a clinical syndrome described first by Pick (1904). In 1975, Warrington (1975) described three patients with progressive impaired recognition of objects and she hypothesised that the deficit was due to a breakdown of conceptual knowledge. Subsequently, Mesulam (1982, 1987) described patients with insidious disturbance of language, beginning with anomia and progressively reduced speech output and comprehension deficit, where he later named this syndrome primary progressive aphasia (PPA). Such patients can be distinguished based on the fluency of their speech, having either progressive nonfluent aphasia or fluent aphasia. The term 'semantic dementia' refers to fluent aphasia with additional loss of word meaning.

Later, Poeck and Luzzatti (1988) described patients with similar deficits. Only a year afterwards, Snowden, Goulding, and Neary (1989) demonstrated that these deficits were not confined to the verbal domain, and they designated these patients as suffering from a "loss of semantic information", coining the term 'semantic dementia'. These patterns of impairment were characterised as degradation of a semantic store (Warrington, 1975).

In substance, SD is characterised by loss of word meaning and impaired language comprehension, with preserved syntactic comprehension, production and fluency of speech output (Gorno-Tempini et al., 2004; Hodges et al., 1992).

SD patients' performances correlate highly between different semantic tasks and show strong item-specific consistency across modalities, suggesting that the anterior temporal lobes underpin a single store of amodal semantic knowledge (Bozeat, Lambon Ralph, Patterson, Garrard, & Hodges, 2000; Rogers et al., 2004). Semantic memory is affected, whereas episodic memory appears intact, where this feature distinguishes SD patients from Alzheimer disease (AD) patients, at least in the first stages of the diseases (Scahill, Hodges, & Graham, 2005).

Voxel-based morphometry studies have shown that SD is characterised by damage to the anterior temporal lobe (Gorno-Tempini et al., 2004). Temporal lobe atrophy in SD is usually bilateral, but most frequently predominant in the left hemisphere. Lateral and medial anterior regions are affected, including the perirhinal cortices and fusiform gyri (Chan et al., 2001; Galton et al., 2001;

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Gorno-Tempini et al., 2004; Mummery et al., 2000; Patterson, Nestor, & Rogers, 2007; Rosen et al., 2002). In addition, the hypometabolism, related to the semantic memory impairment in SD patients is associated with the bilateral rostral temporal lobes, in contrast to a widespread hypometabolism in AD patients (Nestor, Fryer, & Hodges, 2006).

In several reports of single SD cases, patients presenting a progressive prosopagnosia always showed unfamiliar face recognition, with visual basic abilities preserved, in contrast to famous face recognition impairment. As such, the prosopagnosic deficits are of associative-like type and never apperceptive in nature (Barbarotto, Capitani, Spinnler, & Trivelli, 1995; Evans, Heggs, Antoun, & Hodges, 1995; Gainotti, Barbier, & Marra, 2003; Gentileschi, Sperber, & Spinnler, 1999; Joubert et al., 2003; Sperber & Spinnler, 2003; Tyrrell, Warrington, Frackowiak, & Rossor, 1990). The cortical atrophy is always within the temporal lobe (bilateral, greater on the right), in its antero-inferior parts (Gainotti et al., 2003), superior temporal gyrus (Tyrrell et al., 1990) or the right fusiform gyrus (Joubert et al., 2003). Other studies have reported SD patients presenting proper naming deficit and person-specific knowledge impairment (Papagno & Capitani, 1998; Papagno & Capitani, 2001; Poeck & Luzzatti, 1988; Schwarz, De Bleser, Poeck, & Weis, 1998). The presence of these deficits in SD patients might depend on the site of the brain atrophy. Snowden et al. (2004) found that SD patients with predominant left temporal lobe atrophy were better at recognising famous faces than famous names, whereas those with right temporal predominance showed the reverse pattern. Moreover, Thompson et al. (2004) reported that specific person knowledge deficit could persist when the atrophy is predominantly on the right, whereas a general impairment knowledge is present when the temporal atrophy is predominantly on the left.

Two main theories explain these semantic deficits in SD patients. The semantic memory loss could be due to an impairment of explicit retrieval of knowledge or to a degradation of the internal representation of the semantic network (Hodges, Salmon, & Butters, 1992; Hodges et al., 1992). At present, the most consistent hypothesis refers to a representational deficit due to a progressive semantic degradation (Jefferies, Patterson, & Lambon Ralph, 2006; Rogers & Friedman, 2008). In addition to the usual explicit measures of neuropsychological testing, the semantic system integrity has been investigated through implicit tasks, such as priming. The advantage of this technique is that it does not require a behavioural/overt response. In the case of repetition priming (RP), it is postulated that the processing of a stimulus (target) is facilitated when the same stimulus (prime) was encountered before. Results issuing from studies of word priming in neurodegenerative disease patients are mixed. Cumming, Graham, and Patterson (2006) found a greater facilitation effect of priming (hyperpriming) for degraded words (those not identified in an explicit recognition task) in SD patients, when compared to controls. Conversely, in Alzheimer's disease, some effects of facilitation have been found since there is a preserved semantic memory, at least at the onset of the disease (Nebes, 1989). Recently, Rogers and Friedman (2008) compared AD and SD patients using a priming task. AD patients showed a hypopriming, whereas SD patients did not show any priming effect. This has been interpreted as a relatively spared semantic network in AD, in contrast to a clear semantic degradation in SD.

In order to study the face and name representation in SD it would be interesting to explore the use of a priming paradigm. Typically, in this task (see for example: Burton, Kelly, & Bruce, 1998; Johnston & Barry, 2006), the subject is presented with a name of a famous or unknown person (prime) preceded by a related or unrelated face (target). The subject is required to make a decision (for example, a familiarity judgement task) on the target. The prime preactivates the related items, as demonstrated by the way the subject is faster to respond to the target compared to a control condition where there is no relationship between prime and target. According to this hypothesis, if a patient's explicit knowledge system is broken, but some lesser activation persists, facilitation in responses (reaction times, or RTs) could be found in RP. Conversely, if a degraded representation occurs, any kind of facilitation should be found (Shallice, 1988).

In the present study, we investigated face–name processing using an RP paradigm for faces and names in a patient with SD and in ten age-matched healthy controls. We studied the repetition effect using name–name pairs (within-domain) and face–name pairs (cross-domain). In order to investigate the semantic memory related to person representation, we decided to employ a repetition priming technique, exploiting implicit processes. This paradigm permitted us to verify if such a representation is broken or relatively spared but not accessible. The absence of a priming effect could be considered an index of a degraded representation of faces and names, instead of an impaired access to information.

Studies in healthy participants have shown that no priming effect occurred when prime and target are cross-domain (i.e., face–name, name–face) (Bruce & Valentine, 1985; Ellis, Flude, Young, & Burton, 1996). In contrast, more recent studies have shown that priming can cross-domain inputs when the face of a famous person is immediately preceded by the same person's name (or vice versa). Calder and Young (1996) demonstrated a clear effect of cross-domain repetition priming when short intervals occurred between the prime and target of the same famous person ('self-priming'). The amount of priming was larger in within- than in cross-domain condition. Burton et al. (1998) reported evidence of cross-domain priming when the task was semantic in nature (e.g., nationality decision, dead/alive).

There is also evidence of cross-domain priming in prosopagnosic patients, in which overt face processing is impaired. De Haan, Young, and Newcombe (1992) described a prosopagnosic patient (NR) who overtly did not recognise famous faces but performed above chance in a forced-choice familiarity task. NR showed a priming effect in a cross-domain task (face–name), where this effect was restricted to those faces categorised as 'familiar' in the forced-choice task. In addition, Young, Hellawell, and De Haan (1988) described a prosopagnosic patient (PH) who could not overtly recognise familiar faces but showed facilitation of responses to targets (names or faces) preceded by semantically related primes.

We hypothesised that, in our SD patient, no effect of priming would occur in cross-domain priming; specifically, that a famous face would not facilitate the access to its own name. There could also be no effect in within-domain priming. Consequently, responses to targets (names) will not be facilitated because prime (faces and names) representation is degraded.

2. Materials and methods

2.1. Subjects

2.1.1. CMR

CMR, a 67-year-old woman with 8 years of education was diagnosed with SD according to frontotemporal dementia diagnostic criteria (McKhann et al., 2001; Neary et al., 1998). The patient underwent a structural brain MRI, and visual rating of MRI images was compatible with the clinical diagnosis. MRI scanning (July 2007) revealed left temporal lobe atrophy, with consequent enlargement of ventricles, associated with signal abnormalities within the right inferior parietal cortex (see Fig. 1). The patient had no clinical signs of motor or sensory deficits.

Extensive neuropsychological assessment was performed, including global functioning, leaning and memory, non-verbal rea-