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Famous faces and voices: Differential profiles in early right and left semantic dementia and in Alzheimer's disease

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ABSTRACT

Background: Famous face and voice recognition is reported to be impaired both in semantic dementia (SD) and in Alzheimer's Disease (AD), although more severely in the former. In AD a coexistence of perceptual impairment in face and voice processing has also been reported and this could contribute to the altered performance in complex semantic tasks. On the other hand, in SD both face and voice recognition disorders could be related to the prevalence of atrophy in the right temporal lobe (RTL).

Objective: The aim of the present study was twofold: (1) to investigate famous faces and voices recognition in SD and AD to verify if the two diseases show a differential pattern of impairment, resulting from disruption of different cognitive mechanisms; (2) to check if face and voice recognition disorders prevail in patients with atrophy mainly affecting the RTL.

Materials: To avoid the potential influence of primary perceptual problems in face and voice recognition, a pool of patients suffering from early SD and AD were administered a detailed set of tests exploring face and voice perception. Thirteen SD (8 with prevalence of right and 5 with prevalence of left temporal atrophy) and 25 CE patients, who did not show visual and auditory perceptual impairment, were finally selected and were administered an experimental battery exploring famous face and voice recognition and naming. Twelve SD patients underwent cerebral PET imaging and were classified in right and left SD according to the onset modality and to the prevalent decrease in FDG uptake in right or left temporal lobe respectively. Correlation of PET imaging and famous face and voice recognition was performed.

Results: Results showed a differential performance profile in the two diseases, because AD patients were significantly impaired in the naming tests, but showed preserved recognition, whereas SD patients were profoundly impaired both in naming and in recognition of famous faces and voices. Furthermore, face and voice recognition disorders prevailed in SD patients with RTL atrophy, who also showed a conceptual impairment on the Pyramids and Palm Trees test more important in the pictorial than in the verbal modality.

Finally, in 12SD patients in whom PET was available, a strong correlation between FDG uptake and face-to-name and voice-to-name matching data was found in the right but not in the left temporal lobe.

Discussion: The data support the hypothesis of a different cognitive basis for impairment of face and voice recognition in the two dementias and suggest that the pattern of impairment in SD may be due to a loss of semantic representations, while a defect of semantic control, with impaired naming and preserved recognition might be hypothesized in AD. Furthermore, the correlation between face and voice recognition disorders and RTL damage are consistent with the hypothesis assuming that in the RTL person-specific knowledge may be mainly based upon non-verbal representations.

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1. Introduction

Loss of knowledge for famous persons is one of the presenting features of Semantic Dementia (SD), a neurodegenerative disorder characterized by anterior temporal lobe (ATL) atrophy (Snowden et al., 1996; Neary et al., 1998; Hodges et al., 1992). An alteration of this domain can be detected from the earliest stages of disease (Hailstone et al., 2011), mainly affecting person recognition from face when the atrophy is prominent in the right ATL and from their name when it mainly affects the left ATL (Snowden et al., 2004, 2012; Gainotti, 2007; Gainotti, 2015a). Results of Van Lanker and Canter's (1982) early investigations and of Gainotti's (2011) recent review of voice recognition disorders showed that these defects, just as face recognition disorders, are mainly due to right temporal lesions. Loss of knowledge of famous persons is one of the clinical features listed in the diagnostic criteria of Semantic Dementia (Neary et al., 1998).

In Alzheimer's Disease (AD) the earliest and most frequent sign is loss of episodic memory. Famous person knowledge is neither a presenting symptom nor is usually a prominent feature occurring during the course of the disease and is not mentioned as one of the clinical key features in the revised diagnostic criteria of AD (McKhann et al., 2011).

It follows that to detect an impairment in this domain should help the clinician in the differential diagnosis since this disturbance is a prototypical feature of SD whereas it is presumably spared in AD. However, even if the prototypical clinical pictures of the two diseases seems to support this assumption, neuropsychological investigations of this domain in SD and AD reveled that both SD and AD patients show problems on tests of famous person knowledge.

There are few studies exploring this issue in SD case groups. Most of literature is characterized by detailed studies performed on small groups of patients or on single cases, likely due to the rarity of this syndrome that makes it hard to recruit a high number of patients. The first study focusing on famous faces in a group of SD patients came from Snowden et al. (2004), who tested famous faces and famous names knowledge on 15 SD patients and 17 CE patients. Both patients groups were impaired compared to controls. However, SD patients were considerably more impaired than the AD group and only the SD patients were significantly impaired in forced choice familiarity judgement tasks. Furthermore, SD patients with predominantly left temporal lobe atrophy identified faces better than names, whereas patients with right temporal lobe atrophy showed the opposite pattern of performance. The existence of separate stores for faces and names was confirmed by results of a systematic survey made by Gainotti (2007) of all published cases of patients who showed a selective disorder of famous people recognition due to prevalent damage of the right or left ATLs. Results of this review documented a defective retrieval of person specific semantic information from face stimuli, when the right temporal lobe was damaged and a prevalent impairment in finding their names when the anterior parts of the left temporal lobe were affected. In 2011 Hailstone et al. performed a detailed examination of voice processing in 13 patients with temporal lobe atrophy and 22 CE patients. Both SD and AD patients showed deficits in voice recognition. Face and name recognition were impaired in both groups but deficits were more severe in SD patients. AD patients also showed impaired vocal gender perception and voice discrimination, consistent with the additional presence of perceptual impairments in voice processing. These authors also took into account the problem of asymmetric temporal lobe contributions to different components of person knowledge, but could not resolve this issue, because of small case numbers. In the present introduction we will mainly take into account the problem of face and voice recognition disorders in AD and of the possible underlying semantic mechanisms, because this issue has been thoroughly investigated in the neuropsychological literature. For instance, famous face recognition has been widely studied in AD (Calabria et al., 2012; Werheid and Clare, 2007; Clague et al., 2005; Greene and Hodges, 1996; Hodges et al., 1993), showing that person knowledge deficits are present since the early stages of the disease (Clague et al., 2005). There is also a large agreement to acknowledge that in some degenerative diseases the impairment in famous faces naming and identification is not secondary to name access deficit, but to actual loss or impaired retrieval of stored knowledge of persons. More specifically, the nature of the poor performance in tasks involving semantic knowledge of famous persons is reasonably attributable to a primary semantic impairment in SD, because in these patients semantic knowledge is selectively impaired. On the other hand, in AD it is harder to explain the patients' performance in semantic tasks entirely as a consequence of a semantic impairment for a number of reasons. The extent of degradation of the semantic network in Alzheimer's Disease is questionable (Reilly, Peelle, Antonucci & Grossman, 2011). Some researcher (e.g. Hodges and Patterson, 1995; Rogers et al., 2006) are in favour of a loss of semantic knowledge in AD, because semantic impairment is consistent across a variety of semantic tasks and across distinct modalities (verbal and visual channels). Other authors posit that the poor performance in semantic tasks could reflect an impaired retrieval from the semantic network (Nebes et al., 1984) or a deficit of strategic processing (Rich et al., 2002; Moreaud et al., 2001).

Corbett et al. (2012) have also hypothesized that the semantic impairment of AD patients may be due a defect of semantic control similar to that described by Jefferies and Lambon Ralph (2006) in aphasic stroke patients. These authors have distinguished the semantic disorders of SD patients, which might be due to disruption of amodal representations, from the semantic disorders of aphasic stroke patients, which might result from a defect in the executive processes that help to control semantic activation in a task-appropriate fashion. This hypothesis is supported by the fact that the deficit of executive functions subsuming the defect of semantic control is present both in stroke and in AD patients (Corbett et al., 2012). In any case, the coexistence in AD of an impairment of several neuropsychological domains could influence performance in semantic tasks. In particular the potential presence of perceptual problems could play an important role in famous people recognition tasks, because an alteration of visual and auditory perception, often reported in AD (e.g. Golden et al., 2015; Johnson and Chow, 2015; Kaeser et al., 2015; Moyse et al., 2015) could influence the patient performance in semantic tasks involving face and voice domains.

In most studies of famous face and voice recognition, tasks exploring face and voice perception were not performed and when this domain was investigated, such as in the Hailstone et al.'s (2011) work, it was found that the pattern of voice recognition impairment in AD was a sum of primary perceptual problems and of semantic association disorders. Obviously, when a perceptual impairment is present, it becomes hard to evaluate to what extent the perceptual deficit can influence performance on semantic tasks which are dependent on the perceptual analysis of the shown stimuli.

The aim of the present study therefore consisted in investigating famous faces and voices in early AD and SD excluding the potential impact of perceptual impairment on patient profile. We administered to AD and SD patients a detailed set of neuropsychological tests exploring face and voice perception. Only patients who showed preserved perceptual abilities were included in the study and investigated with tasks exploring naming and recognition of famous faces and voices. We had two distinct predictions, concerning on one hand the differences between AD and SD patients in the recognition of famous faces and voices and, on the other hand, the prevalence within SD patients of face and voice recognition disorders when the atrophy mainly affected the right ATL. As for the first point, we expected to find in SD patients a failure both in naming and in recognition of famous faces and voices, whereas more problematic were the predictions concerning AD patients. To be sure, we expected to find a naming deficit in these patients, but we did not know if their capacity to