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Short communication

Identification of environmental sounds and melodies in syndromes of anterior temporal lobe degeneration ☆

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ABSTRACT

Recognition of nonverbal sounds in semantic dementia and other syndromes of anterior temporal lobe degeneration may determine clinical symptoms and help to define phenotypic profiles. However, nonverbal auditory semantic function has not been widely studied in these syndromes. Here we investigated semantic processing in two key nonverbal auditory domains – environmental sounds and melodies – in patients with semantic dementia (SD group; n = 9) and in patients with anterior temporal lobe atrophy presenting with behavioural decline (TL group; n = 7, including four cases with MAPT mutations) in relation to healthy older controls (n = 20). We assessed auditory semantic performance in each domain using novel, uniform within-modality neuropsychological procedures that determined sound identification based on semantic classification of sound pairs. Both the SD and TL groups showed comparable overall impairments of environmental sound and melody identification; individual patients generally showed superior identification of environmental sounds than melodies, however relative sparing of melody over environmental sound identification also occurred in both groups. Our findings suggest that nonverbal auditory semantic impairment is a common feature of neurodegenerative syndromes with anterior temporal lobe atrophy. However, the profile of auditory domain involvement varies substantially between individuals.

1. Introduction

Semantic dementia (SD) is a focal neurodegenerative syndrome characterised by insidiously progressive impairment of semantic memory due to selective, asymmetric antero-medial temporal lobe atrophy [1]. SD is a canonical syndrome of frontotemporal lobar degeneration (FTLD) yet its phenotypic boundaries remain unclear: while semantic processing of words and visual objects has been studied intensively, much less is known concerning other knowledge modalities in SD that are likely also to contribute to symptoms and disability [1–9]. Furthermore, although research consensus diagnostic criteria have been developed for stratifying the major syndromes of FTLD [10,11], in practice the SD syndrome often shows substantial overlap clinically and anatomically with other syndromes of FTLD, in particular behavioural variant frontotemporal dementia (bvFTD) [12,13]. While not the presenting feature, semantic deficits are often prominent in such cases; moreover this spectrum includes cases with selective (particularly nondominant) temporal lobe atrophy [12–15] and the important subgroup represented by MAPT mutations, characteristically accompanied by focal bilateral anterior temporal lobe atrophy [12,16,17]. Conversely, SD is commonly associated with early and prominent behavioural abnormalities that overlap closely with bvFTD [18].

Among the nonvisual sensory modalities of semantic knowledge, nonverbal sound is of particular interest as a potential probe of this SD-like phenotypic spectrum, on both clinical and neuroanatomical grounds. The nonverbal auditory domain encompasses both environmental sound sources and events (including highly biologically and socially salient signals) and music (which exemplifies a nonverbal, abstract and autonomous rule-based semantic system [19,20]). Studies of auditory agnosia associated with focal brain damage and functional imaging studies of nonverbal sound processing in the healthy brain have implicated distributed temporo-parietal networks that closely overlap those damaged in FTLD [21–29]. The available evidence further suggests that these auditory semantic domains are at least partly dissociable neuropsychologically and neuroanatomically [21,30,31]. In line with this neuropsychological and neuroanatomical evidence, impaired identification of environmental sounds has been documented as part of more generalised semantic impairment in SD [2,4,7]. Findings for

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music processing in SD and other syndromes of FTLD appear somewhat inconsistent [3,5,32–35]: several well documented case studies show relatively-spared identification of melodies in patients with SD despite widespread semantic deficits [3,5,35], though group studies have qualified this [6,34]. In addition, a substantial proportion of patients with SD and bvFTD develop abnormal craving for music ('musicophilia') and/or aversion to environmental sounds [36,37], suggesting that these nonverbal auditory domains may underpin a repertoire of clinically salient symptoms in these disorders. However, previous neuropsychological investigations have relied largely on cross-modal labelling (word or picture matching) or in the case of melodies, familiarity judgement, rather than assessing specific within-modality identification and comparing domains directly. Accordingly, the true status of nonverbal sound processing in SD and related syndromes has not been fully defined.

Here we assessed nonverbal auditory semantic processing across the domains of environmental sounds and music in a cohort of patients with SD. In parallel, we assessed a group of patients presenting with behavioural symptoms (i.e., a syndrome of bvFTD) who also had selective temporal lobe atrophy on MRI. Our objectives were to assess in detail clinically-relevant domains of nonverbal auditory semantic memory in SD; and to compare the auditory semantic profile in SD with another syndrome (bvFTD) associated with anterior temporal lobe degeneration. Identification of environmental sounds and melodies was compared using a novel, uniform procedure based on within-modality stimulus matching, thereby obviating the need for cross-modal (especially, verbal) labelling. Based on previous neuropsychological and clinical evidence [2–6,35], we hypothesised that patients with SD would show impairments in both nonverbal auditory semantic domains, albeit less severely and consistently for melodies; that relative sparing of melody knowledge might be a hallmark of the SD group; and that patients with a syndrome of SD would show more severe nonverbal auditory semantic deficits than other patients with selective temporal lobe atrophy.

2. Methods

2.1. Participants

Nine consecutive patients fulfilling consensus criteria for typical SD [10] and seven patients who fulfilled consensus criteria for a primary syndrome of probable bvFTD [11] with predominant temporal lobe atrophy on MRI (based on visual assessment by an experienced neuroradiologist blinded to clinical diagnosis) were recruited from a specialist cognitive disorders clinic (representative sections from each patient's brain MRI are shown in Figure S1 in Supplementary material on-line). The latter, bvFTD syndromic group is hereafter designated the 'temporal lobe' [TL] group to emphasise the criterion of relatively focal temporal lobe atrophy used in selecting these patients; it is this subgroup that overlaps most closely with the SD group. The TL group included four patients with a confirmed pathogenic MAPT mutation; this high proportion is in line with previous evidence that these mutation cases usually present with behavioural changes but frequently also exhibit prominent semantic deficits associated with focal antero-medial temporal lobe atrophy [12,16,17]. Twenty age-matched healthy individuals also participated in the study. No participant had a history of clinical hearing loss. All participants underwent comprehensive assessment of musical background and general neuropsychological functions; participant group characteristics including background behavioural data are summarised in Table S1 in Supplementary Material on-line. None of the participants was a professional musician; on the basis of a previously described semi-structured caregiver questionnaire [36], one patient in the SD group and two in the TL group exhibited musicophilia, defined as abnormally increased interest in music compared with premorbid levels sufficient to interfere with daily life functioning.

2.2. Experimental behavioural tests

We adapted a previously described paradigm [4,5] to create tests of environmental sound and melody identification within the auditory modality. These tests were based on presentation of pairs of sound stimuli derived either from the same environmental sound source or tune ('same' condition) or from different sound sources or tunes ('different' condition). Identification in each test was assessed by asking the subject to determine whether the source sounds or tunes for the members of each stimulus pair were the same or different, thereby avoiding the need for cross-modal labelling of particular sounds. The tests are schematised in Fig. 1 (stimulus details are in Supplementary Tables S2 and S3 on-line). Environmental sounds and melodies were all highly familiar to healthy older British individuals based on previous pilot work [4,5] (see also Supplementary Table S4 on-line). Stimuli were chosen such that 'same' and 'different' pairs did not differ in overall perceptual similarity: tunes were all presented as piano melodies controlled for musical attributes such as key, metre and tempo. The

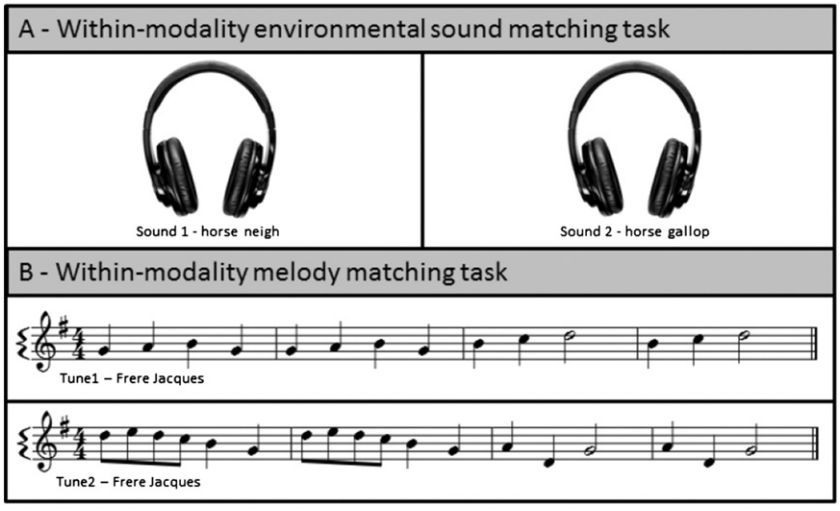

Fig. 1. Examples of environmental sounds (A) and notated tune excerpts (B) used in the identification by within-modality matching tasks (here, examples for the 'same' source sound and melody conditions are shown).