Agnosia for accents in primary progressive aphasia

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ABSTRACT

As an example of complex auditory signal processing, the analysis of accented speech is potentially vulnerable in the progressive aphasia. However, the brain basis of accent processing and the effects of neurodegenerative disease on this processing are not well understood. Here we undertook a detailed neuropsychological study of a patient, AA with progressive nonfluent aphasia, in whom agnosia for accents was a prominent clinical feature. We designed a battery to assess AA's ability to process accents in relation to other complex auditory signals. AA's performance was compared with a cohort of 12 healthy age and gender matched control participants and with a second patient, PA, who had semantic dementia with phonagnosia and prosopagnosia but no reported deficits involving accents. Relative to healthy controls, the patients showed distinct profiles of accent agnosia. AA showed markedly impaired ability to distinguish change in an individual's accent despite being able to discriminate phonemes and voices (apperceptive accent agnosia); and in addition, a severe deficit of accent identification. In contrast, PA was able to perceive changes in accents, phonemes and voices normally, but showed a relatively mild deficit of accent identification (associative accent agnosia). Both patients showed deficits of voice and environmental sound identification, however PA showed an additional deficit of face identification whereas AA was able to identify (though not name) faces normally. These profiles suggest that AA has conjoint (or interacting) deficits involving both apperceptive and semantic processing of accents, while PA has a primary semantic (associative) deficit affecting accents along with other kinds of auditory objects and extending beyond the auditory modality. Brain MRI revealed left peri-Sylvian atrophy in case AA and relatively focal asymmetric (predominantly right sided) temporal lobe atrophy in case PA. These cases provide further evidence for the fractionation of brain mechanisms for complex sound analysis, and for the stratification of progressive aphasia syndromes according to the signature of nonverbal auditory deficits they produce.

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PPA syndromes span a hierarchy of early perceptual, apperceptive and semantic processing stages, analogous to the processing hierarchy established for visual objects (Warrington & Taylor, 1973; Warrington, 1982; Warrington & Taylor, 1978; Riddoch & Humphreys, 1987; Griffiths & Warren, 2002, 2004; Goll et al., 2010a). Particular PPA syndromes are associated with distinctive profiles of nonverbal auditory deficits: Whereas auditory apperceptive and semantic impairments have been demonstrated in both SD and PNFA, additional early auditory perceptual impairments occur in PNFA and more widespread auditory deficits have been documented in LPA (Goll et al., 2010a, 2011).

The processing of accents is potentially of particular relevance to understanding the PPA syndromes (Hailstone et al., 2012). Accent is a meta-linguistic feature of spoken utterances that conveys information about the speaker’s geographical or socio-cultural background: accent is therefore potentially a rich source of nonverbal semantic information about speakers. In addition, accent modifies the acoustic properties of spoken phonemes, interacting with individual vocal characteristics and prosody (Boula de Mareuil & Vieru-Dimulescu, 2006; Clopper & Pisoni, 2004; Howell, Barry, & Vinson, 2006); if spoken phonemes are regarded as auditory objects (Griffiths & Warren, 2004), then a phoneme spoken in a non-native accent could be considered as a non-canonical ‘view’ of the phoneme for a particular listener, and should therefore engage auditory apperceptive processing. Both recognition of non-native accents and comprehension of words spoken with less familiar accents have been shown to be impaired in patients with PNFA, in keeping with conjoint semantic and apperceptive deficits of accent processing in this PPA syndrome (Hailstone et al., 2012). However, limited information is currently available concerning the brain basis of accent processing and the impact of disease on this processing. In particular, no detailed and systematic comparison of the processing of accent in relation to other kinds of complex auditory signals has previously been undertaken in PPA.

Here we describe a detailed analysis of the processing of accent in a patient, AA, with PNFA. Difficulties with accent recognition and comprehension were early and prominent features of AA’s clinical syndrome. AA’s performance on apperceptive and semantic analysis of accents, voices, speech and environmental sounds was assessed using a novel neuropsychological battery and compared with the performance of healthy control participants and another patient, PA, with a syndrome of SD characterised by progressive anoma, prosopagnosia and phonagnosia, but no reported difficulties with accent processing.

2. Methods

2.1. Participant details

Demographic data for all participants are summarised in Table 1.

2.1.1. Patient AA

This 67 year old right handed retired teaching assistant, who had lived in the London area for the whole of her life, presented with a two year history of progressive word finding difficulty and hesitant, effortful speech. In addition, she had noticed prominent difficulty identifying a speaker’s accent and in understanding non-native accents. For example, when watching a film or television programme she was unable to follow the conversation of actors speaking in foreign accents or to identify their accents. In the last year she had also experienced some difficulty recognising individual voices. This was particularly evident when using the telephone, though remained relatively mild in relation to her difficulties with accents. On examination her speech was nonfluent and agrammatic, with speech apraxia and frequent phonetic errors. She exhibited prominent orofacial apraxia; the general neurological examination was normal. When asked to identify the examiner’s accent (Australian) she reported that she had not realised that this was non-native but when pushed, suggested that he might be ‘Northern’. AA fulfilled current clinical diagnostic criteria for the nonfluent-agrammatic variant of PPA, here designated PNFA (Gorno-Tempini et al., 2011). Brain MRI showed predominantly left-sided peri-Sylvian atrophy (Fig. 1).

2.1.2. Patient PA

This 71 year old right handed retired medical secretary, who had lived in the South East region of England for the whole of her life, presented with a seven year history of progressive difficulty recognising people. When first assessed, she had difficulty recognising close relatives and friends. In addition, for the past two years she had developed difficulty recognising voices over the telephone and had begun to notice problems recalling the names of things. She had recently developed an obsessional interest in puzzles and crossword books. Family members also reported that she was less empathic. On examination her speech was garrulous and circumlocutory with anoma. The general neurological examination was unremarkable. PA was diagnosed clinically with a semantic dementia syndrome led by progressive prosopagnosia. Brain MRI showed marked bilateral anterior temporal lobe atrophy, more severe on the right (Fig. 1).

2.1.3. Healthy control participants

Twelve healthy age and gender matched individuals (mean age 66 years, range 57–71 years) participated. All were native English speakers. Eleven had grown up in the South East of England and had lived in the London area for the majority of their lives; one participant had originally grown up in New York but had lived in London for the last forty years. No participant had a history of neurological or psychiatric illness. The healthy control group had, on average, higher educational attainment than the patients (see Table 1): The patients had 10 and 11 years of education (corresponding to finishing school aged 15 or 16, prior to O-Levels/G.C.S.E.s) whereas the control group had on average 16 years of education (corresponding to Degree level education).

All participants were recruited via the Cognitive Disorders Clinic at the National Hospital for Neurology and Neurosurgery. The study was approved by the local institutional research ethics committee and all participants gave informed consent in accord with the principles of the Declaration of Helsinki.

2.2. General neuropsychological assessment

A comprehensive assessment of general neuropsychological functions covering language, executive functions, working memory and posterior cortical cognitive domains was undertaken in all participants. Details of the neuropsychological tests administered are summarised in Table 1.

Table 1 Summary of general demographic and cognitive data for all participants.

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>AA</th>
<th>PA</th>
<th>Healthy controls*</th>
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**General**

| Age (years) | 67 | 71 | 66 (57–71) |
| Education (years) | 11 | 10 | 16 (10–20) |
| Symptom duration (years) | 2 | 3 | N/A |
| MMSE (max 30) | 26 | 28 | N/A |
| Verbal IQ | 78 | 84 | 121 (106–130) |
| Performance IQ | 97 | 93 | 120 (88–141) |

**Language**

| BPVS (max 150) | 126 | 136 | 147 (129–150) |
| GNT (max 30) | 0 | 3 | 26 (19–29) |
| NART (max 50) | 12 | 27 | 44 (30–49) |

**Arithmetical and spatial**

| GDA addition (max 12) | 5 | 5 | 6.9 (4–11) |
| GDA subtraction (max 12) | 6 | 6 | 8.7 (6–12) |
| VOSP (max 20) | 19 | 18 | 17 (13–20) |

**Executive**

| Stroop: Colour naming (time in seconds) | 48 | 27 | 28 (24–36) |
| Stroop: inhibition (time in seconds) | 72 | 60 | 52 (36–70) |
| Digit span reverse (maximum string length) | 5 | 6 | 5 (4–7) |

Key: *mean (range) data shown. Patient data below healthy control range are shown in bold. †two healthy control participants did not complete general neuropsychological assessments; BPVS, British Picture Vocabulary Scale (McCarthy & Warrington, 1992; Lloyd et al., 1982); GNT graded naming test; GDA, Graded Difficulty Arithmetic (Jackson & Warrington, 1986); IQ, scores calculated from the Wechsler Abbreviated Scale of Intelligence (Wechsler, 1999); MMSE, Mini-Mental State Examination score; NART, National Adult Reading Test; Stroop, D-KEFS Stroop test (Delis, Kaplan, & Kramer, 2001); VOSP, Visual Object and Spatial Perception battery. |
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