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 aphasias. 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 in accent processing. 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.

1. Introduction

The progressive aphasias (PPA) are a diverse group of neurodegenerative syndromes with characteristic clinico-anatomical signatures and heterogeneous histopathology (Mesulam, 1982; Gorno-Tempini et al., 2008, 2011). Three canonical PPA syndromes are recognised (Gorno-Tempini et al., 2011): progressive nonfluent aphasia (PNFA), characterised by impaired speech production and agrammatism associated with predominant left peri-Sylvian atrophy; semantic dementia (SD), characterised by impaired single word comprehension and loss of vocabulary, associated with asymmetric, selective anterior temporal lobe atrophy; and logopenic aphasia (LPA), characterised by prolonged word-finding pauses and impaired auditory verbal working memory, associated with predominant left temporoparietal atrophy. By definition, PPA syndromes are primarily defined by language deficits; however, nonverbal deficits are increasingly recognised and are likely to be integral to the pathophysiology of PPA, reflecting a profile of brain network disintegration in these diseases. Examples of such non-linguistic impairments include the breakdown of multi-modal object and conceptual knowledge in SD (Bozeat, Lambon Ralph, Patterson, Garrard, & Hodges, 2000; Goll et al., 2010a; Goll, Crutch, & Warren, 2010b; Goll, Ridgway, Crutch, Theunissen, & Warren, 2012; Hailstone, Crutch, Vestergaard, Patterson, & Warren, 2010; Luzzi et al., 2007; Omar, Hailstone, Warren, Crutch, & Warren, 2010; Piwnica-Worms, Omar, Hailstone, & Warren, 2010; Josephs, 2008; Fletcher & Warren, 2011) and deficits of nonverbal sound processing across the PPA spectrum (Hailstone et al., 2010, 2011, 2012; Goll et al., 2010a, 2011; Rohrer, Sauter, Scott, Rossor, & Warren 2012). With respect to nonverbal sounds, deficits in...
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 concurrent 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.

### Table 1

| Characteristics | AA | PA | Healthy controls |
|-----------------|----|----|------------------|
| Age (years)     | 67 | 71 | 66 (57–71)       |
| Education (years)| 11 | 10 | 16 (10–20)       |
| Symptom duration (years)| 3 | 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) | 4 | 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.

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 anomia. 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.
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