Agnosia for accents in primary progressive aphasia

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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, Theuissen, & Warren, 2012; Hailstone, Crutch, Vestergaard, Patterson, & Warren, 2010; Luzzit al., 2007; Omar, Hailstone, Warren, Crutch, & Warren, 2010; Piwim-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 joint 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 anomia, 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 dif...
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