Patterns of limb apraxia in primary progressive aphasia

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Accepted 7 May 2003

Abstract

Primary progressive aphasia (PPA) is a syndrome characterized by a progressive language deficit without other dementia features for at least two years (Weintraub et al., 1990). Other deficits that are likely to coexist with aphasia, such as apraxia, have only been investigated in a few case studies and only at a rudimentary level for the most part. In this study we investigate the frequency and severity of apraxic deficits in PPA patients. Ten PPA patients and twelve aged-matched healthy adults performed eight transitive gestures to pantomime and to imitation. Gesture performance was measured along five movement dimensions and a composite score based on the arithmetic mean of the five dimensions was calculated. Overall, PPA patients performed worse than controls with both pantomime and imitation. Furthermore, individual comparisons revealed that out of the three apraxia patterns described by Roy (1996) (pantomime alone, imitation alone, or apraxia in both conditions), the most frequent pattern in PPA patients was apraxia in both conditions. This result corresponds with previous findings in populations of stroke and Alzheimer's patients. Considering the occurrence of apraxia in this population, this study supports the idea that a comprehensive apraxia assessment should be administered in cases of PPA.

1. Introduction

Primary progressive aphasia (PPA) is a syndrome characterized by slowly progressive language deficits without accompanying dementia for at least two years (Weintraub et al., 1990). Since being conceptualized as a unique clinical syndrome by Mesulam (1982), there have been many reports of PPA in the literature (for a review see Westbury & Bub, 1997). However, only a few of these studies have systematically looked at other deficits that are likely to coexist with aphasia. For instance, apraxia, a movement disorder in performing gestures that is not attributable to any primary sensory or motor dysfunction, has often been simultaneously observed in patients exhibiting aphasia after focal brain lesions in the left hemisphere (Basso et al., 1981). Some authors have argued the overlap of aphasia and apraxia in these patients is due to a central language deficit that affects both language and gestural production (Duffy & Duffy, 1981; Glosser et al., 1986). Alternatively, other authors counter that aphasia and apraxia are separable behavioral deficits that often coexist due to the close proximity of the anatomical substrates that underlie language and gesture production (Feyereisen et al., 1988; Kertesz et al., 1984; Lehmkuehl et al., 1983). Since aphasia and apraxia have been observed independently of each other the second hypothesis appears more likely.

Despite the comorbidity of aphasia and apraxia, it is interesting that only a small number of investigations have examined the presence of apraxia in PPA (Karbe et al., 1993; Kempler et al., 1990; Kertesz et al., 1994; Hudson et al., 1994). For instance, in a study of three PPA patients, Kempler et al. (1994) identified limb apraxia in two patients and orofacial and truncal apraxia in the third patient. Kempler also identified limb apraxia in two PPA patients while Fuh et al. (1994) observed the development of orofacial apraxia three years after the onset of language difficulties. Other studies have found negative results of apraxia in PPA patients, at least in the early stages of the disease (Beland & Ska, 1992; Craenhals et al., 1990). However, based on this limited literature, it is difficult to form strong conclusions about the presence...
and attributes of apraxia in PPA. As stated in one overview, ‘praxis has not been well studied in this group of patients’ (Black, 1996).

1.1. Patterns of limb apraxia in brain lesion patients

Previous praxis research has shown that apraxia can be exhibited under conditions of pantomiming (responding to verbal command) and imitation (Leipmann, 1908). Dissociations have been observed in which some patients have greater difficulty with pantomiming than imitation (Schnider, Hanlon, Alexander, & Benson, 1997; Watson, Fleet, Rothi, & Heilman, 1986) while other patients show the opposite pattern (Barbieri & De Renzi, 1988). Extending the research of others (Barbieri & De Renzi, 1988; De Renzi, Motti, & Nichelli, 1980; Leipmann, 1908), Roy and colleagues (Roy, 1996; Roy & Hall, 1992; Roy & Square, 1985) have attempted to account for apraxic deficits by developing a multi-stage model of gesture performance.

Subsequent research by Roy (Heath, Roy, Black, & Westwood, 2001; Roy, 1996; Roy & Square, 1994; Roy et al., 2000) described three patterns of apraxia (i.e., selective pantomiming deficits, selective imitation deficits and combined pantomiming and imitation deficits), which were hypothesized to reflect disruptions at different stages of the gesture production model. Firstly, specific deficits in pantomiming were believed to involve disruptions to the conceptual system (i.e., recalling the knowledge of a tool and its associated action) or the early stages of the production system in which image generation and response selection are required. Secondly, explicit deficits of imitation, which according to the model bypasses the conceptual system and early stages of the production system, were proposed to involve the sensory/perceptual pathways in which analyzing visual information and translating visual gestural information into an action are affected. Finally, deficits in both imitation and pantomiming were theorized to affect the later stages of the production system, which organizes and controls the execution of movement patterns.

In this study, 10 PPA patients referred to a University Cognitive Neurology Clinic were assessed on tests of praxis. The apraxia tests included performing gestures under conditions of pantomiming and imitation. This study aimed to determine not only the overall frequency of apraxia in PPA patients but also the relative frequency of the patterns of praxis deficits identified by Roy (1996).

2. Methods

2.1. Subjects

Ten PPA and 12 age-matched healthy volunteers participated in this study. All PPA patients met the following criteria; they were previously well, community-dwelling participants with an insidious onset of aphasia gradually progressing in the absence of memory and other signs of global dementia. No patient had a history of other neurological disorders, especially stroke. Nine PPA participants were right-handed (five men, four women) and one woman was left-handed. Mean age and education of the patient group was 64.1 years and 14.1 years, respectively. Based on the Western Aphasia Battery (WAB) (Kertesz, 1982) four patients had Broca’s, five had Anomic, and one had Global aphasia. The 12 healthy controls tested were all right-handed and matched on age (65.7 years) and education (14.8).

2.2. Praxis testing

Tests of apraxia were administered by a trained examiner. Each participant was seated at a table facing a video camera and the examiner who was seated to the right (except for the left-handed patient where the examiner was seated to the left). Two large mirrors (60 x 180 cm) were placed on either side of the participant in order to view the examiner’s gesture during imitation conditions and also to view parts of the participant’s movements that were not directly visible to the camera. Participants were required to pantomime and imitate eight transitive gestures (e.g., using a hammer). In the pantomiming condition, to overcome any confounding effects of poor language comprehension, the participants were shown a picture of a tool and asked how they would use it (e.g., for the hammer, the instructions were ‘show me how you would use this to pound in a nail’). In the imitation condition, the experimenter performed a gesture and instructed the participant to concurrently mimic the action as closely as possible. PPA patients performed all gestures with their dominant hand while the control subjects performed the gestures with both hands.

Praxis performance was analyzed using testing dimensions that were derived from previous work (Roy, Square, Adams, & Friesen, 1985) and are based on movement features found to be important in manual signing (Stokoe, 1972). These five dimensions include: location of hand in space relative to the body, plane of movement of the hand, action (i.e., the movement characteristics of the gesture), hand posture, and orientation of the hand. Each dimension was rated on a three-point scale reflecting the degree of accuracy as follows: two for correct, one for distorted, and zero for incorrect. This method of scoring has previously shown high interrater reliability (Roy, Black, Blair, & Dimeck, 1998). A composite score was derived for each gesture under each performance condition. This score reflected performance across the five dimensions and was expressed as a percentage of the total possible score for
each gesture. Composite scores were then averaged across each performance condition (pantomime vs. imitation) for each subject.

3. Results

3.1. Performance of control participants

Composite scores of control participants were analyzed in a two-hand (right, left) × two-condition (pantomime, imitation) ANOVA. Results revealed no hand differences in gesture performance, $F(1,11) < 1$. Therefore, subsequent comparisons with PPA patients pooled the performance of both hands. There was a significant effect of condition, $F(10.26, p < .01)$. Specifically, controls were more accurate under conditions of imitation versus pantomiming. This finding replicated previous results with control participants (e.g., Roy et al., 2000).

3.2. Comparisons between the control and PPA groups

An averaged composite score (i.e., arithmetic mean of left hand/right hand performance) of control participants was compared with the dominant hand performance of PPA patients in a two-group (control, PPA) × two-condition (pantomime, imitation) mixed-factor ANOVA. There was a main effect for group, $F(1,20) = 23.68$, $p < .001$, illustrating that control participants performed more accurately than PPA patients. This is shown in Fig. 1. In addition, there was a main effect of condition $F(1,20) = 7.14$, $p < .05$, revealing that all participants were more accurate under the imitation condition compared to pantomime. Further pair-wise comparisons (using Bonferroni correction for multiple comparisons) revealed that controls were significantly more accurate than PPA patients under both pantomiming ($t(20) = 4.06$, $p < .01$) and imitation ($t(20) = 4.01$, $p < .01$) conditions. Correspondingly, a group × condition interaction was not significant, $F = 3.53$, $p > .05$.

3.3. Frequency and patterns of apraxia in PPA

To assess the frequency of apraxia in PPA, a cutoff point of the composite scores based on the mean scores of control patients (see Roy et al., 1998) was used for each testing condition. Patients were classified as apraxic on a particular test item if their composite score was ≥2 standard deviations (SD) below the control mean and as borderline if their score was between 1 and 2 SD. Table 1 illustrates the frequency of apraxia in the both testing conditions for all PPA patients (two asterisks indicate apraxic and one asterisk indicates a borderline score).

Categorization of patients into each of the three patterns of apraxia proposed by Roy (1996) was based on these cutoff scores. Thus, seven out of 10 (70%) patients were apraxic for both pantomime and imitation conditions, while two patients had selective imitation deficits. One patient, WB, showed no deficits under either testing condition.

4. Discussion

4.1. Apraxia results

In this study we examined the prevalence and patterns of apraxia in a population of 10 PPA patients. Compared to controls, PPA patients performed significantly worse overall under both pantomiming and imitation conditions. Further analyses comparing individual PPA scores to normative data revealed widespread apraxic performance in this group. Specifically, apraxia was evident under conditions of both pantomiming and imitation for 7 of the 10 participants, while selective imitation
Impairments were observed with two patients. According to the model developed by Roy and colleagues (Roy, 1996; Roy & Square, 1985), deficits on both pantomiming and imitation indicate dysfunction in the later stages of the production system involved in controlling and organizing gestures. Selective imitation deficits, on the other hand, are proposed to result from the dysfunction of the systems involved in analyzing and translating visual gestural information. This distribution of the patterns of apraxia reflecting a higher frequency in impairments in pantomiming and imitation is consistent with our work in studies of patients of stroke (Heath et al., 2001; Roy et al., 2000) and Alzheimer's disease (Barbour, Roy, Black, & Scott, 1997).

Overall, the presence of apraxia in PPA was significant. Some may argue that this result supports a unitary language deficit that underlies both apraxia and aphasia (e.g., Duffy & Duffy, 1981; Glosser et al., 1986). However, as others have described before, apraxia and aphasia are observed independently in different patient populations (Beland & Ska, 1992; Lehmkuhl et al., 1983; Feyereisen et al., 1988) suggesting that the co-occurrence of these deficits is more likely to be due to dysfunction of adjacent neural areas. Within this study, the occurrence of two patients with selective imitation deficits illustrates that aphasia is not always associated with pantomime apraxia, as would be predicted with the first hypothesis. Overall, these results illustrate the importance of assessing apraxia in PPA patients, and moreover, when possible, using a multi-task apraxia battery.

4.2. Neural substrates underlying PPA and apraxia

Since the early work of Leipmann (1908), the left parietal lobe has been proposed to store the motor programs involved in performing skilled movements bilaterally. Although this initial hypothesis has been supported in subsequent studies (e.g., De Renzi et al., 1980) more recent reports suggest the laterality of the praxis system may be overstated (Roy et al., 2000). Moreover, a review of the neural structures underlying apraxia show there is no single lesion site of this disorder (Leiguarda & Marsden, 2000). Alternatively, researchers have identified distributed parallel networks that work in concert to form an intact praxis system.

As at least three major pathways have been identified based on functional neuroimaging studies of humans (e.g., Catalan, Honda, Weeks, Cohen, & Hallett, 1998) and neurophysiological studies of non-human primates (Rizzolatti et al., 1996). These circuits include a fronto-parietal network, a fronto-striatal network, and a temporoparietal-frontal network. Each pathway has been hypothesized to contribute to different components of the praxis system. Specifically, the fronto-parietal network has been shown to be involved in sensorimotor integration (Rizzolatti et al., 1996) and movement sequencing (Catalan et al., 1998); the fronto-striatal network for movement sequencing; and the temporoparietal-frontal network in the recognition and imitation of actions (Di Pellegrino, Fadiga, Fogassi, Gallese, & Rizzolatti, 1992).

Studies investigating the neural substrates involved in primary progressive aphasia have most often shown anomalies in the left inferior frontal and temporal lobes (Black, 1996; Chawluk et al., 1986; Kobayashi et al., 1990; Mesulam, 1987; Schletens, Ravid, & Kamphorst, 1994) although other anomalies have also been noted (Black, 1996; Delecluse et al., 1990). Interestingly, all nine of the apraxic PPA showed dysfunction under imitation conditions, a finding consistent with disruptions to the temporoparietal-frontal network (Di Pellegrino et al., 1992). Thus, comparing the overlap of neural structures involved in apraxia and PPA, it seems reasonable to propose that left fronto-temporal dysfunction contributes both to aphasia and apraxia in PPA. Future functional and anatomical studies may elucidate the strength of this hypothesis.

Acknowledgments

This work was partially funded through grants to E. Roy from NSERC and to E. Roy and S. Black from the Ontario Mental Health Foundation and the Heart and Stroke Foundation of Ontario.

References


