

Reading and Spelling Impairments in Progressive Aphasia: Evolving Behavioral Deficits and Neural Substrates

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Abstract

The nature and progression of written language impairments were examined in one individual with semantic dementia (SD) and one with progressive nonfluent aphasia (PNFA). Participants were tested on reading and spelling of regular words, irregular words, and nonwords at two points in time. Voxel-based morphometry (VBM) was performed to determine regional cortical atrophy at each time point. Results indicate diverging patterns of behavioral performance and underlying cortical atrophy in SD versus PNFA. Lexical-semantic impairments were associated with extrastriangular temporal lobe atrophy in SD, while phonological impairments were associated with perisylvian atrophy in PNFA.

Introduction

Impairments of written language may result from damage to the brain caused by stroke or neurodegenerative disease. The nature of such impairments in individuals with focal lesions has been well-established^{1,2}; however, reading and spelling deficits in individuals with progressive aphasia due to neurodegenerative disorders have been less thoroughly examined. Such deficits have been explored in the context of SD, where the loss of conceptual representations results in surface dyslexia/dysgraphia³. In contrast, difficulty with nonwords as well as irregular words has been noted in individuals with PNFA⁴.

Methods

- **Participants**
 - One individual with SD and one with PNFA (Table 1; assessed at two points in time approximately 20 months apart)
- **Language battery**
 - Tests of naming, semantic processing, and overall language performance
 - Reading and spelling of regular words, irregular words, and nonwords
 - Effects of regularity (regular vs. irregular words), lexicality (real words vs. nonwords), frequency (high vs. low frequency words) and imageability (high vs. low imageability words) were calculated
- **High-resolution, 3D MRI scans**
 - Areas of regional cortical atrophy (relative to 13 controls) determined using voxel-based morphometry (VBM) implemented in SPM5

Table 1. Demographic characteristics and performance on standardized language measures

Sex	Age	Educ.	Handed-ness	WAB AQ and aphasia type	PPT	AZ Semantic Test	BNT
SD	F	78	R	T1→T2 anomic→ 72.6 anomic	T1→T2 42→38	T1→T2 32→28	T1→T2 6→2
PNFA	F	71	R	88.8 anomic→ 76.5 conduction	T1→T2 52→52	T1→T2 40→40	T1→T2 37→35

NAZ tested on the WAB. T1/T2 = testing time points one and two. AZ = Arizona Reading Test (out of 52). BNT = Boston Naming Test (out of 60). WAB AQ = Western Aphasia Battery Aphasia Quotient (out of 100).

Figure 1a,b. Reading performance on regular words, irregular words, and nonwords

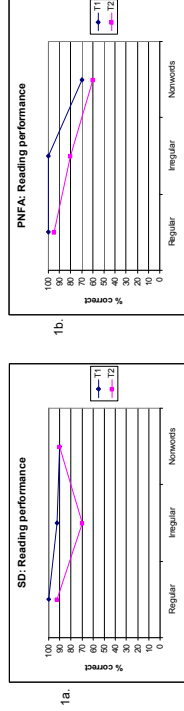
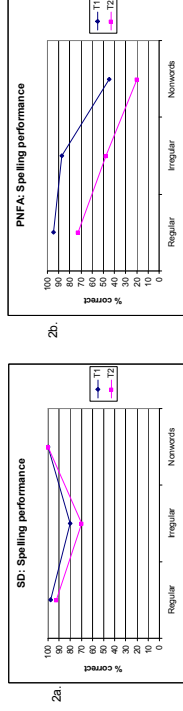


Figure 2a,b. Spelling performance on regular words, irregular words, and nonwords



Interval between T1 and T2 approximately 20 months.

Tables 2a,b. Effects of linguistic variables on reading and spelling performance at time 1 and time 2

	Semantic Dementia			Progressive Nonfluent Aphasia		
	T1	T2	T1	T2	T1	T2
Regularity effect	*	*	*	*	*	*
Lexicality effect			*	*	*	*
Frequency effect	*	*	*	*	*	*
Imageability effect						*

*Indicate a significant effect as determined by Fisher's exact tests

Figure 3a,b. Results of VBM (p<0.01, uncorrected) for SD patient at time 1 (a) and time 2 (b); color bar indicates t-values

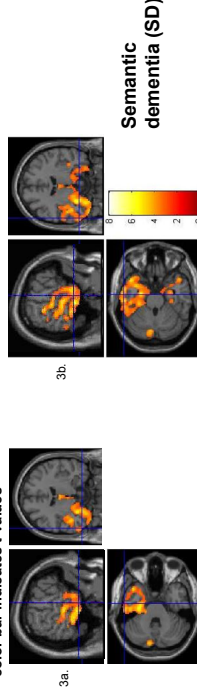
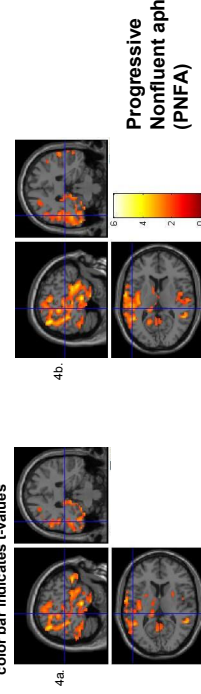


Figure 4a,b. Results of VBM (p<0.05, uncorrected) for PNFA patient at time 1 (a) and time 2 (b); color bar indicates t-values



Results

- **Diverging patterns of performance in SD versus PNFA** (Figures 1,2; Tables 2a,b)
 - SD patient
 - regularity effect at T2 for reading
 - regularity effect at T1 that became more pronounced at T2 in spelling
 - PNFA
 - lexicality effects at T1 that persisted at T2 for reading and spelling
 - regularity effects at T2 for both modalities
- **VBM at T1 revealed typical patterns of atrophy for SD and PNFA** (Figures 3, 4)
 - SD patient- atrophy primarily of left anterolateral and anteromedial temporal cortex
 - PNFA- atrophy primarily in left perisylvian cortical regions
- **VBM at T2 revealed changes in both the spatial extent of atrophy as well as the degree of atrophy observed in regions affected at T1.** Of particular interest were changes in regions thought to be critical to phonological and lexical-semantic processing of written language.
 - SD patient- increased atrophy in the left temporal lobe, with extension into posterior inferior temporo-occipital cortex and superior temporal regions
 - PNFA patient- increase in degree and extent of atrophy affecting left perisylvian cortex

Discussion

These findings suggest distinct patterns of progression for the reading and spelling impairments in SD versus PNFA. In SD, the evolving semantic deficit likely contributes to an increased reliance on phonology and thus a progressively larger regularity effect over time. The underlying pattern of atrophy in left temporo-occipital cortex is consistent with previous findings of lexical-semantic reading and spelling impairments in patients with stroke affecting the same brain regions.

Conversely, findings in the PNFA patient suggest that declining phonological abilities related to progressive perisylvian cortical atrophy may contribute to increasing difficulty reading and spelling nonwords. These results are consistent with the well-documented association between phonological dyslexia/dysgraphia and perisylvian cortical damage in stroke. The emergence of regularity effects in PNFA may suggest the development of a subtle lexical-semantic impairment necessitating reliance on residual phonological abilities. An alternative interpretation is that phonological processing is critical to both nonword and real word reading/spelling and that, when the phonological deficit reaches a critical level of severity, low frequency, low imageability, irregular words may be affected in a manner similar to nonwords⁵.

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