Neuropsychologia 50 (2012) 1327–1334

Contents lists available at SciVerse ScienceDirect

Neuropsychologia

journal homepage: www.elsevier.com/locate/neuropsychologia

Preserved visual language identification despite severe alexia

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ARTICLE INFO

Article history:
Received 19 May 2011
Received in revised form 26 January 2012
Accepted 22 February 2012
Available online 3 March 2012

Keywords:
Pure alexia
Letter-by-letter reading
Language
Orthography
Covert reading
Occipital cortex

ABSTRACT

Patients with letter-by-letter alexia may have residual access to lexical or semantic representations of words despite severely impaired overt word recognition (reading). Here, we report a multilingual patient with severe letter-by-letter alexia who rapidly identified the language of written words and sentences in French and English while he had great difficulty in reading them, judging their lexical status or extracting semantic information. Lexical decision was strongly influenced by the orthographic structure of stimuli: whereas he easily determined the lexical status of illegal nonwords (e.g., '*rsdo*'), he had random performance with legal pseudowords (e.g., '*binus*'). When asked to determine the language of meaningless letter trigrams with high frequency in the English or French orthography (e.g., '*oth*' or '*iqu*') his performance was significantly above chance. In contrast, similarly to healthy participants his language decision was at chance with low-frequency trigrams. These findings suggest that written language identification relies on sublexical processing of orthographic rules specific to each language.

1. Introduction

Letter-by-letter alexia (LBL) is an acquired reading disorder consecutive to left occipito-temporal damage centered on the fusiform gyrus (Cohen et al., 2003; Damasio & Damasio, 1983). Though naming of isolated letters is often preserved in patients with LBL alexia word reading is laborious and characterized by a letter-by-letter identification strategy. As a result, the time to identify a word is linearly related to the number of its constituting letters, with an increase of up to 2 s or more for each additional letter (Behrmann, Nelson, & Sekuler, 1998; Leff et al., 2001). Current accounts posit that the impairment preventing patients with LBL reading from adopting the parallel process observed in normal readers affects peripheral processes such as the visuo-orthographic level (Arguin & Bub, 2005; Behrmann, Plaut, & Nelson, 1998; Patterson & Kay, 1982).

In spite of their laborious reading some LBL readers are able to categorize words according to their semantic and/or lexical status, suggesting at least partial activation of higher-level lexical-semantic representations (Behrmann, Plaut, et al., 1998; Lambon Ralph, Hesketh, & Sage, 2004). Evidence for this phenomenon – termed the Saffran effect – is the fact that some LBL readers perform well above chance in lexical or in semantic decision tasks (Coslett, Saffran, Greenbaum, & Schwartz, 1993; Landis, Regard, & Serrat, 1980; Saffran & Coslett, 1998). Here, we present a LBL patient (AL) who demonstrates a form of Saffran effect that has not been described yet: he was able to identify rapidly and without effort the language of words and sentences that he failed to read. Several neuropsychological and neurophysiological studies support the idea that the human brain, as a result of perceptual experience, encodes the orthographic structure of a specific language (McClelland & Rumelhart, 1981). More precisely, Binder et al. (Binder, Medler, Westbury, Liebenthal, & Buchanan, 2006; see also Dehaene, Cohen, Sigman, & Vinckier, 2005) demonstrated that the left lateral fusiform gyrus, a region whose damage is critical for the occurrence of pure alexia, is sensitive to the familiarity of letter combinations. According to these findings, we hypothesize that AL's language decision might be based on the detection of local orthographic regularities that are specific to each language. For example, the letter trigram '*oth*' is common in English, but does not exist in French whereas the trigram '*iqu*' is frequent in French, but not in English. Based on this hypothesis we expected that AL is capable of identifying the language of nonsense letter combinations provided they have a high frequency of occurrence in a specific language.

2. Material & Methods

2.1. Patient description

AL was a highly educated, 72-year-old right-handed man who suffered a left medial occipito-temporal ischemic stroke (Fig. 1) causing pure alexia and right superior quadrantanopia. He was a native French-speaker. He had excellent knowledge of English and

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doi:10.1016/j.neuropsychologia.2012.02.017

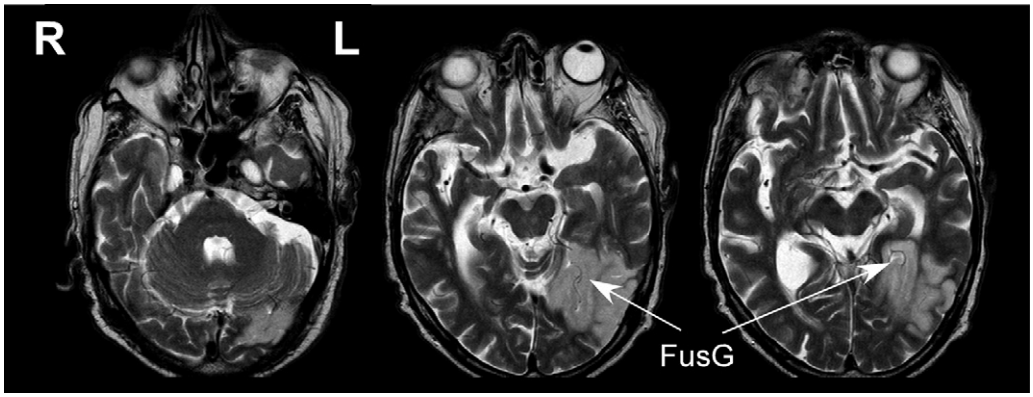

Fig. 1. T2-weighted MRI-scan showing ischemic damage to the left occipito-temporal cortex involving the fusiform (FusG), lingual, lateral occipital, and posterior parahippocampal gyrus.

Table 1
Results of neuropsychological evaluation.

Function	Test	Patient raw score (%)	Percentile	Classification
Naming on definition	Boston Naming Test (1)	19/20 (95%)	>50	High average
Repetition	Clinical protocol	44/44 (100%)		High average
Oral comprehension	MT-86 (2)	43/47	–	High average
Writing (oral spelling)	Items from de Partz (3)	Regular words 19/20	–	Average
		Irregular words 18/20	–	Average
Reading from oral spelling	Examen des Dyslexies Acquises (4)	97/99 (98%)	–	High average
Visual recognition	VOSP incomplete letters	14	18.8	Impaired
	VOSP Object Decision	16	17.7	Low average
	VOSP Dot Counting	10	9.9	High average
	VOSP Position Discrimination (5)	20	19.6	High average
Memory	Warrington words (6)	43	43	High average
Executive functions	Verbal fluency (7)	34	34.2	Average
	Figural fluency (8)	16	26.3	Low average

1. Kaplan, Goodglass, & Weintraub (1983); 2. Nespoulous et al. (1992); 3. de Partz (1994); 4. Lemay (1990); 5. Warrington and James (1991); 6. Warrington (1984); 7. Thurstone and Thurstone (1963); 8. Regard (1981).

good knowledge of German (languages he had often used during his professional life), though he had mostly stopped to use these languages since the time of his retirement (seven years prior to this study). Using the classification of Roux et al. (2004) at the time of this study AL would be classified as LIII (acquisition of English and German after the age of 7 years, not spoken every day). Standardized neuropsychological testing revealed moderate deficits of verbal and non-verbal memory, executive functions and visual identification (Table 1). Other cognitive functions, including ideomotor praxis, arithmetic skills and short-term memory, were normal. Oral production of language, comprehension and repetition were normal, and writing was intact. The most apparent deficit was an inability to read visually presented material, while identification of words orally spelled was virtually perfect. He showed extremely laborious letter-by-letter reading, sometimes confusing even single letters, and was often unable to identify a word even under unlimited exposure. For example, AL would identify the word "étoile" (star) as /e/ /tO/ /i/ /b°/ /e/ (corresponding to: é-to-i-beé) in a first attempt, and subsequently as /etwab/ (corresponding to: étoibe) which took him more than 10 s. Amazingly, when asked to indicate in what language a text that he was unable to read was written (English, French, or German), he immediately recognized the language. Given the absence of aphasia in French, and the fatigability of the patient, we decided not to examine spoken English.

2.2. General methods

The study started 3 months after the stroke and lasted for 6 months. During this period the patient's reading deficit remained stable. Evaluation of reading was performed in French and English. The patient's performance was compared to three age- and gender-matched healthy volunteers (3 males, mean age, 71 ± 2.7 years). All were native French speakers with excellent knowledge of written English which had been acquired after the age of 7 years and not spoken every day (proficiency classified as LIII according to Roux et al. (2004)). The study was approved by the Ethics Committee of the University Hospital of Geneva, and all participants gave informed, written consent.

Before every experiment, several practice trials were performed. Item presentation in each experiment was pseudo-randomized and counterbalanced across control-subjects. Stimuli were presented centrally on a 17″ computer-screen in black letters (Arial, size 36) on white background. A central fixation cross was presented for 1000 ms, followed by a blank screen (100 ms), and the stimulus (either 400 ms or unlimited, as specified below). In the naming and reading tasks, responses were registered with a voice-sensitive key with millisecond accuracy. In the forced-choice tasks, participants responded by pressing one of two keys on a button box with their left and right index finger.

2.3. Reading

2.3.1. Methods

Reading of letters was evaluated by asking participants to name each letter of the Latin alphabet presented for unlimited time. Upper and lowercase letters were presented in pseudo-random