

NOTE

PROGRESSIVE ANOMIA WITHOUT SEMANTIC OR PHONOLOGICAL IMPAIRMENT

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ABSTRACT

We describe a 59-year-old woman, M.T., with a progressive language impairment and neuroimaging findings of decreased perfusion (SPECT) and focal atrophy (MRI) in the left temporal region. The most prominent feature of her cognitive profile was a profound and progressive impairment in naming. In spite of this, she performed normally on tests of semantic processing and phonological output. Her spontaneous speech was fluent with preserved syntax and articulation but with notable word-finding problems. All other cognitive abilities were relatively stable and intact. These features are not typical of either fluent or non-fluent forms of neurodegenerative language disturbance. The cognitive mechanisms that may underlie this case are discussed.

Key words: primary progressive aphasia, semantic dementia, anomia, semantic memory, speech production

Over two decades ago, Mesulam (1982) described six patients with a slowly progressive decline in language function without the involvement of other cognitive domains. Mesulam and Weintraub (1992) later termed the syndrome “primary progressive aphasia” (PPA) and proposed that it be diagnosed when language function progressively deteriorates over at least a two-year period with relative preservation of other cognitive abilities and activities of daily living, other than those that place a premium on language function. The syndrome of PPA is heterogeneous, however, and two major subtypes of language disorder can be identified: fluent and non-fluent progressive aphasia.

In a systematic review of 112 PPA cases, Westbury and Bub (1997) determined that anomia was the most common initial deficit in both PPA subtypes. In fact, anomia was the presenting symptom in five of Mesulam’s (1982) original cases. In fluent PPA, also known as “semantic dementia” (Hodges et al., 1992a; Snowden et al., 1989), anomia is generally thought to result from a progressive loss of knowledge about the meaning of words and is typically accompanied by impaired single-word comprehension. Speech remains fluent in these patients with relative preservation of phonology and syntax, but it becomes increasingly “empty” and circumlocutory with semantic paraphasias (Hodges et al., 1992a, 1995, 1998; Kertesz et al., 1998; Snowden et al., 1989). By contrast, in non-fluent PPA, anomia is thought to result from disturbed phonological representations and is usually accompanied by effortful, agrammatic

speech containing phonemic paraphasias. Comprehension of single-words remains relatively intact, although these patients are often impaired in their comprehension of syntax (Hodges and Patterson, 1996; Karbe et al., 1993; Thompson et al., 1997; Weintraub et al., 1990).

In this paper, we report a patient, M.T., with a progressive language deficit that was not typical of either the fluent or non-fluent forms of PPA. M.T. displayed a profound anomia but with no evidence of impairment in semantic processing or phonological output at this point in her disease.

CASE REPORT

M.T. was a 58-year-old right-handed woman with grade 12 education who first presented to us in October, 2002 with a five-year history of word-finding difficulty. Her chief complaints included problems recalling names of family members, friends and plants in her garden. She reported some benefit, however, from the use of a self-taught strategy in which she sequentially went through the phonemes of the alphabet to try and cue her retrieval of a word. She also reported difficulties in recalling day-to-day events and maintaining her train of thought, particularly while reading. She described numerous difficulties with instrumental activities of daily living including managing household finances, medications, grocery shopping and cooking. She had previously worked as an interior designer but had resigned one-year after the onset of her cognitive problems.

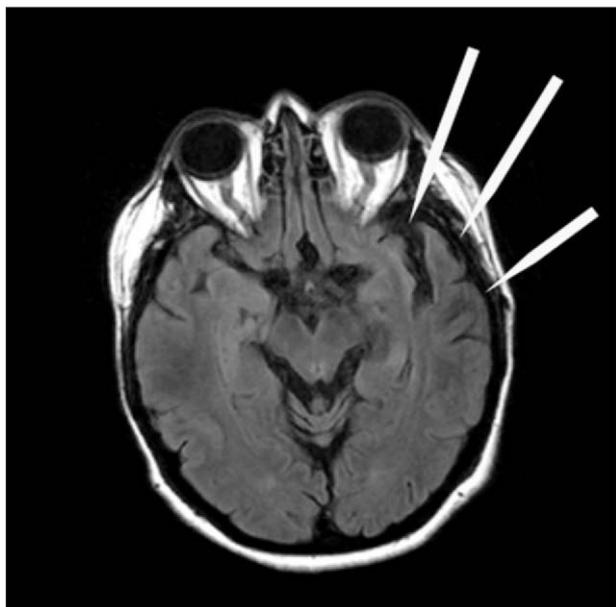


Fig. 1 – MRI scan (T2 Flair) from April 2003 showing left anterior temporal lobe atrophy.

M.T. had a long and persistent history of depression despite numerous anti-depressant trials and her cognitive symptoms were initially attributed to depression. A computed tomography (CT) scan in 1997 and a MRI scan in 2001 had been read as normal. Other medical history and medication use were all non-contributory. M.T.'s family history was not known as she was adopted.

M.T.'s husband confirmed her difficulties in word-finding and in consistently performing many instrumental activities of daily living. However, he felt that her difficulties carrying out these latter activities were due to her depression and that she performed them well at times when her mood was improved.

On exam, M.T. displayed significant word-finding difficulties despite fluent conversational speech. Syntax, articulation and prosody appeared entirely normal. Cognitive screening was conducted

using a standardized Behavioural Neurology Assessment Battery which assessed the domains of attention, memory, language, visuospatial function, executive function and praxis (Darvesh et al., 2005). M.T.'s scores in each domain fell above the cut-off scores for dementia with the exception of object naming which was severely impaired. Her neurological and general physical examinations, as well as her blood work, were all within normal limits.

A MRI scan in April, 2003 revealed left anterior temporal lobe atrophy (see Figure 1). A SPECT scan in November, 2003 showed striking hypoperfusion in the left anterior temporal lobe (see Figure 2). Although subtle hypoperfusion in the left frontal lobe was also suggested this may have been due to M.T.'s co-existing depression, and was not clearly related to the left temporal hypoperfusion or atrophy (Mayberg, 2003).

LONGITUDINAL NEUROPSYCHOLOGICAL ASSESSMENTS

M.T. had general neuropsychological assessments on three occasions: October 2002, August 2003 and May 2004. Results from a neuropsychological assessment that was conducted in September 2000 at a different site were also available. Table I shows M.T.'s performance on a range of neuropsychological tests in each of the four assessments.

M.T.'s overall verbal skills as assessed by the Wechsler Adult Intelligence Scale – Third Edition (WAIS-III; Wechsler, 1997) declined over the four assessments. In the first assessment, her scores on the Vocabulary and Information subtests were relative strengths, although they did not differ statistically from the mean of the verbal subtest scores ($p > .05$; Wechsler, 1997). By the second assessment, however, scores on all verbal subtests, with the exception of Digit Span, had declined and

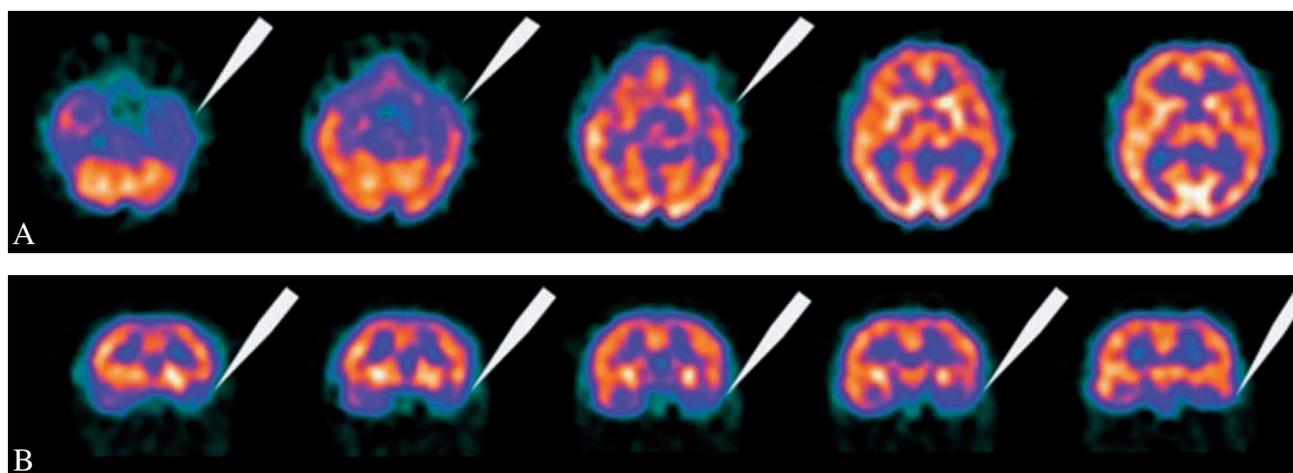


Fig. 2 – SPECT scan from November 2003 showing left temporal and frontal hypoperfusion. (a) Transverse sections; (b) Coronal sections.

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