Objective: To describe pure alexia and auditory comprehension problems in a young woman with multiple sclerosis (MS).

Patient: A 33-year-old woman with MS who complained of difficulties in reading and comprehending spoken language was referred for a neuropsychological examination. Reading difficulties were confirmed and most of the reading errors were additions, omissions, and substitutions of single letters. While the patient reported that the letters seemed to disappear before her eyes, no general problems with visual attention, visual discrimination, or scanning were detected. No difficulties with spelling were reported. The auditory comprehension deficit is interpreted as a form of a semantic access disorder and is not due to generalized slowing in information processing or conceptual disintegration.

Conclusions: Pure alexia is unusual in MS and to our knowledge only 1 other case has been reported (in Japanese). Memory impairments and slowed information processing are probably the most frequent cognitive sequelae of the disease and, consequently, the literature is biased toward the study of those cognitive domains. However, given the wide distribution of sclerotic plaques in MS, it could be argued that we should expect some variability of cognitive changes in MS. Striking deficits as seen in this patient should make us more sensitive to this possibility.

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It is now acknowledged that 40% to 65% of patients with multiple sclerosis (MS) are cognitively impaired. Memory deficits are the most frequently reported cognitive sequelae of MS, but deficits in other cognitive domains are also well documented. Multiple sclerosis can impair speech production but aphasia is relatively rare, although aphasia and naming problems have been reported. Reading ability is generally unaffected, and only 1 case of alexia in MS is reported in the literature.

We describe a patient with MS whose main cognitive complaints were difficulties in reading and comprehending spoken language.

REPORT OF A CASE

The patient was diagnosed as having MS in 1986 when she was 26 years old. She had been healthy until 4 years earlier when she was diagnosed as having optic neuritis. Later she experienced diminished sensation in her upper extremities for several weeks. At the time of diagnosis she had paresthesia and pain in her lower extremities, which had progressed over several weeks. Both visual and somatosensory (tibial nerve) evoked responses were abnormal on the left side of the patient’s body. Brainstem auditory responses were normal. Cerebrospinal fluid examination in February 1988 revealed 13 mononuclear cells (1-3 x 10^6/L). Gamma fraction was slightly elevated but no oligoclonal bands were observed. In the years following, the patient had several periods of relapses and remissions with secondary progression. In 1995, visual and brainstem evoked responses were abnormal bilaterally. The patient had no history of stroke-like episodes.

A magnetic resonance imaging scan taken in February 1994 showed bilateral lesions in the periventricular white matter. There were also bilateral lesions in the occipital lobe white matter (tractus opticus and cuneus) and small subcortical lesions in the superior temporal gyri on both sides. When the patient was referred for a neuropsychological examination, her main complaint was that she was having difficulty reading. She found this distressing and reported that she had difficulties reading to her children. She also reported problems in comprehending spoken language and feeling as if people were speaking in a foreign language. She had also noticed that her memory had become worse. She underwent a neuropsychological examination in November 1993 and February 1994. Results from the 2 testing sessions were comparable. Despite her complaints of dwindling memory, performance on standard verbal and nonverbal memory tests was within normal limits. However, that did not rule out a change from premorbid functioning. Information processing and psychomotor speed were slowed.
We describe a patient with MS who has alexia as well as auditory comprehension difficulties. It has been proposed elsewhere that decreased information processing speed in MS can indirectly cause difficulties with concept formation. Presumably, impaired information processing speed could also make other cognitive functions less efficient. We would not consider such an explanation for the comprehension disorder in this case, much less the alexia. Herein we provide a clear example of a patient with pure alexia, and we think it more appropriate to consider the comprehension deficit a form of a semantic access disorder than to attribute the problem to generalized slowing in information processing. The comprehension problem in this case is no different from what one sees in many aphasic patients with cerebrovascular disease and should not be labeled differently because the patient has MS. However, it may be appropriate to consider difficulties with semantic access the result of slowed processing in a single component of the language system. Magnetic resonance imaging results are consistent with the clinical picture seen here because there are lesions in the occipital lobe white matter, as well as in the superior temporal lobe gyri in the left hemisphere. However, there are other lesions as well, and it is difficult to make anatomical-clinical correlations in the presence of a widespread disease.

Although the variability of cognitive changes in MS has been emphasized, the literature is biased toward the study of memory, while other cognitive domains have been somewhat neglected. There is some variability in the distribution of sclerotic plaques in MS, and the focus on group studies may lead to less frequent cognitive sequelae receiving less interest than if that focus were not exclusively on group studies. Similar claims have been made about cognitive impairments in schizophrenic patients. Given the pathological features of MS, it is likely that memory, attentional, and executive deficits dominate, but the study of these disorders to the exclusion of other potential deficits will impede our understanding of the cognitive consequences of MS.

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REFERENCES


TABLE 1

<table>
<thead>
<tr>
<th>Error Types</th>
<th>No. (%) of Errors</th>
<th>Examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>Omissions</td>
<td>11 (23)</td>
<td>fálmâ→fálm</td>
</tr>
<tr>
<td>Complex errors</td>
<td>11 (23)</td>
<td>forþsâlmâ→fors</td>
</tr>
<tr>
<td>Additions</td>
<td>10 (21)</td>
<td>kamar→kambär</td>
</tr>
<tr>
<td>Substitutions</td>
<td>10 (21)</td>
<td>skíningur→skíldingur</td>
</tr>
<tr>
<td>Transpositions</td>
<td>3 (6.4)</td>
<td>pell→epell</td>
</tr>
<tr>
<td>Pronunciation errors</td>
<td>2 (4.3)</td>
<td>NA</td>
</tr>
</tbody>
</table>

*The total number of errors is 47. NA indicates data not applicable.

of frontal lobe functioning revealed no perseveration or difficulties with shifting and conceptualization. However, verbal fluency was decreased.

Sentence repetition and auditory lexical decisions were normal. This finding indicates that both early auditory analysis and the knowledge of which sound patterns constitute real words (lexical decisions) were intact. However, the patient had difficulties in comprehending spoken language, both in normal interaction and during testing. She also occasionally failed to immediately understand words she could read out loud correctly. The patient’s auditory comprehension as measured by the Token Test was poor (29/36 [80%]). On this test she was very slow and sometimes had to repeat the sentences to herself prior to responding. Healthy young individuals obtain a score of at least 34 (94%) of 36 on the Token Test and generally respond quickly. There was no evidence of semantic impairments on an auditory picture-word matching test (40/40 [100%]) or on a test of semantic associations (19/20 [95%]). There was mild output anomia, revealed by abnormally long response latencies. Color naming was also relatively slowed. Spelling was at premorbid levels. When reading, she said she needed to spell the words prior to reading them, and reported that the letters seemed to disappear before her eyes. When tested she could name all letters correctly, both single letters and letters within words. She was asked to read 384 single words out loud and 33 nonwords. Of the words, 87.8% (337/384) were correctly read; of the nonwords, 78.8% (26/33). There were no effects of word class, word frequency, or concreteness. There were no semantic errors and most of the errors were additions, omissions, and substitutions of single letters that resulted in word responses in 85% of the cases. Most of the errors (64%) were on the right side of the words. The error pattern for nonwords was similar. Although the patient claimed she had to spell the words to herself prior to reading them, no clear word length effects were obtained, perhaps because reading speed was not greatly reduced. Rapid presentation of words (1 word per 300 milliseconds) did not increase reading errors.

There was no evidence of visual difficulties on tasks other than reading. Tests of visual search and neglect were easily performed, and on those tasks the patient did not have the impression that stimuli disappeared. Rapid presentation (<2 seconds) of small line drawings did not cause difficulties with identification, even when 3 drawings were presented simultaneously. The patient had no difficulties with arithmetic and reported that numerical stimuli did not disappear from the page as did letters. Therefore, no general problems with visual attention, visual discrimination, or scanning accounted for the reading difficulties experienced by the patient.