A case of a 50-year-old, right-handed female, monolingual native Spanish-speaker with a university-level education and cognitive changes is reported. Over approximately 2 years, she presented with a progressive deterioration of writing abilities associated with acalculia and anomia. An MRI disclosed a left parietal temporal atrophy. Two years later, further significant cognitive decline consistent with a dementia of the Alzheimer’s type was observed. Amnesia, executive dysfunction, and ideomotor apraxia were found. Writing was severely impaired, and some difficulties in reading were observed. Copying abilities, however, were relatively well preserved, and the patient could drive and go to different city locations without significant spatial orientation difficulties. A second MRI approximately 2 years later showed that brain atrophy had progressed significantly. Spontaneous writing and writing to dictation were impossible. The ability to read words was preserved, but the ability to read pseudowords was lost. Changes in calligraphy were noted. This case clearly illustrates the progression of focal cognitive defects over time and the spread of abnormalities to other domains.

Key words: progressive agraphia, progressive acalculia, progressive anomia

Progressive deterioration of a single domain of cognitive function is unusual in Alzheimer’s disease (AD). Progressive aphasia, alexia, agraphia, amusia, visuospatial disturbances, anarthria, ideomotor apraxia, dressing apraxia, and visual agnosia with initial preservation of most other cognitive functions have been described in histopathologically verified AD and are considered among the atypical clinical variants of Alzheimer dementia (e.g., Ardila, Rosselli, Arvizu, & Kuljis, 1997; Benson, Davis, & Snyder, 1988; Berthier, Leiguarda, Starkstein, Sevlever, & Taratuto, 1991; Buxbaum, Giovannetti, & Libon, 2000; Caselli et al., 1999; Crystal, Horoupian, Katzman, & Jotkowitz, 1982; De Renzi, 1986; Fukui, Sugita, Kawamura, Shiotai, Nakano, 1996; Holf et al., 1993; Infante et al., 2000; Jagust, Davies, Tiller-Borcich, & Reed, 1990; Kertesz, Davidson, & McCabe, 1998; Kiyosawa et al., 1989; Otsuki, Soma, Yoshimura, & Tsuji, 1997; Polk & Kertesz, 1993; Rosen et al., 2002; Ross et al., 1996; Garcia-Sanchez et al., 1998; Wakai et al., 1994). Primary progressive aphasia is a condition of insidious onset and gradual progression in which there is typically a slow progression and isolated impairment in language function for at least 2 years before additional cognitive impairments develop (e.g., Garcia-Sanchez, Estvez-Gonzalez, Catafau, & Escartin, 1998; Graff-Radford et al., 1990; Hodges, Patterson, Oxbury,

Alexia is often mentioned in cases of progressive aphasia and progressive posterior cortical dementia (e.g., Ardila et al., 1997; Chiacchio, Grossi, Stanzione, & Trojano, 1993; Freedman et al., 1991). Several of these reports also refer to agraphia (e.g., Benson et al., 1988; Berthier et al., 1991; Grossman et al., 2001; Kiyosawa et al., 1989; Mendez, Mendez, Martin, Smyth, & Whitehouse, 1990). However, the severity of these manifestations and the level of documentation and assessment vary widely, and the type of alexia and agraphia are usually not specified or documented in sufficient detail. Furthermore, there is a wide range of individual variability in deficits encountered.

Ardila et al. (1997) described a 65-year-old woman with progressive visuospatial dysfunctions associated with alexia and agraphia. MRI revealed cerebrocortical atrophy, which was particularly severe in both parieto-occipital regions. Alexia was significantly more severe than agraphia, fitting the pattern of a letter-by-letter reading alexia. Both lexical and spatial agraphia were evident. The patient’s ability to write deteriorated as the orthographic ambiguity of the target word increased, and a tendency toward regularization in writing was observed (lexical agraphia). Misuse of spaces, iterations of strokes and letters, slanting of the lines, and underuse of the left side of the article were observed (spatial agraphia).

Grossman et al. (2001) described a patient who presented with writing difficulty that deteriorated over time. Although her graphemes were typically legible, her writing was extremely slow, and her letters were written in an inconsistent and heterogeneous manner. She also produced allophonic and spelling errors (mainly omissions and perseverations) during her writing. Using a positron emission tomography scan, superior parietal occipital and superior frontal defects were more evident at the left. The authors proposed that the patient was experiencing a deficit in retrieving physical letter forms as manifested by her heterogeneous and slow production of letter forms.

In the current case, a progressive loss of writing skills associated with acalculia and anomia were noted. Initial MRI scans showed a focal left frontal-parietal temporal atrophy. Two different evaluations were performed, the first in August 1999 and the second in July 2001. During the first evaluation, memory difficulties were mild; during the second evaluation, memory difficulties were significant, and acalculia, agraphia, and anomia were severe.

Case Presentation

Background

The case was a 50-year-old, right-handed female (A.R.A.), monolingual native Spanish-speaker with a university-level education. No familial history of neurological or psychiatric illness was found in the two past generations. A.R.A. had two sisters and three brothers, all of whom were grown and married and had successful lives without any evident cognitive deterioration. A.R.A. was married with two healthy sons, aged 25 and 17 years. A.R.A. worked as a successful psychologist in a government educational institute in Mexico. Her job duties included writing and lecturing frequently. Approximately 2 years before the first evaluation, A.R.A. noted difficulties in correctly writing words and using Spanish orthography. She mentioned thinking hard how to write the words. A.R.A. frequently wrote the words several times and corrected them. Her job required budget management, and she made errors in calculations. She had to ask someone else to review the accounts and to repeat the arithmetical operations. In addition, she began to have some word-finding difficulties that prevented her from lecturing.

Because of these difficulties, A.R.A. left her job 1 year before the first evaluation and began to help in her brother’s business. According to her family, difficulties became significant from approximately 1½ years before the first evaluation. Her family mentioned difficulties in using numbers, handling money, word finding, and maintaining the topic although she was speaking. They reported that arithmetical errors were frequent but mild; that is, that she usually got a result that was close to the correct answer. Her family also emphasized her writing disturbances, which made it difficult for her to perform her daily working duties in her brother’s business. At this time, A.R.A. consulted a neurologist. The neurological exam was normal. MRI scans taken in June 1999 showed a left parietal-temporal atrophy (Figure 1). She was referred for neuropsychological testing.

First Evaluation

During the first evaluation performed in June 1999, the patient was fully oriented. Spontaneous language was nearly normal. Speech articulation, speed, and voice
volume were normal. Fluency, phonology, grammar, and sentence length were normal during conversational speech. Mild word-find difficulties resulted in occasional semantic paraphasias, and circumlocutions, however, were noted. Verbal IQ (Wechsler, 1981) was 86, Performance IQ was 103, and Full Scale IQ was 93 (Table 1). Total score in the Neuropsi–Brief Neuropsychological Test Battery for Spanish Speakers (Ostrosky, Ardila, & Rosselli, 1999; Ostrosky-Solís, Ardila, & Rosselli, 1997) was 91 (mildly abnormal for her age and education). Decreased scores were observed in successively subtracting 3s from 20, naming, phonological verbal fluency, semantic verbal fluency, writing, arithmetical problems solving, categories recall, language comprehension, sequences, changing right-hand position, and opposite reactions subtests. Verbal and nonverbal memory-recall condition were borderline.

Language repetition, reading, verbal memory–coding, copying a semicomplex figure, similarities, digits backwards, visual detection, words recognition, changing right-hand position, and hands alternating movements tasks were within the normal range for her age and educational level. Scores on the Wechsler Memory Scale were within the normal range. Score on a Spanish naming test (Gardner, 1987) containing 80 items was 42 (abnormal). A.R.A. recognized right and left in her body and the examiner’s body. Errors were noted in naming and pointing to fingers. Without any evident difficulty, A.R.A. recognized the faces of relatives, friends, and people around her.

Good calligraphy was observed in writing. Spatial distribution was appropriate, and no spatial neglect was noted. Three different writing tasks were used:

- spontaneous writing: to report her history in writing (Figure 2)
- copying: written words of a 44-word story
- dictation: a short text (28 words) with 8 words, and 10 pseudowords

In reporting her history, 130 words were used. The report contained 20 corrections. Orthographic errors were abundant (e.g., cercanas → sercanas). Letter omissions and additions were noted (e.g., ortografia → hortografia). Nonhomophone errors also were observed (e.g., máquina → magina). In writing words under dictation, similar errors were found. In copying a text, 1 word from 44 was incorrectly written, and the patient wrote an extra word. No reading errors were observed in reading the 109-word story included in the Neuropsi. Reading understanding was normal; she correctly answered all the Neuropsi reading understanding questions.

Calculation abilities were abnormal. Defects were observed in basic arithmetical operations, both written and oral. Reading numbers was correct up to three-digit numbers. Writing single digits to dictation was correct, but writing two-digit numbers was abnormal (e.g., 45 → 25). Four-digit comparisons could not be made (e.g., What four-digit number is bigger in an eight-number series; for example, 2541–1859). In addition to the calculation subtest included in the Neuropsi, and the Wechsler Adult Intelligence Scale–Arithmetic subtest, the Wide Range Achievement Test (WRAT)–Revised (Jaskat & Wilkinson, 1984) was administered. Her score of 7/40 was regarded as abnormal. Three two-digit numbers could not be added. Multiplication tables could not be used. To the problem 3 × 4, she answered 18. Arithmetical signs were confused (e.g., 6/2 = 12, 33 – 17 = 50), and fractions could not be used.

Over the next 2 years, the patient attended a cognitive rehabilitation program. She continued helping in her brother’s business. She continued driving, and no spatial or topographic difficulties were noted. In the rehabilitation program, special emphasis was placed on calculation abilities, writing, and language.

Second Evaluation

In July 2001, a further neuropsychological evaluation was conducted (see Table 1). The patient was alert, oriented, and cooperative, with fluent speech. Attention level was normal. No articulation defects, apraxia of
speech, or agrammatism were observed. However, a significant decrease in most test scores was found. Neuropsi scores in word recognition, language repetition, similarities, left-hand position, and alternating movements with both hands subtests remained within the normal range. All the WAIS subtest scores were more than one standard deviation below the expected score. Total score in the Neuropsi Neuropsychological Test Battery score was 79/130 (severely abnormal). Interestingly, her spontaneous recall of the semicomplex figure corresponded to the normal range, but her score during the Wechsler Memory Scale Visual Reproduction subtest was 5/14. Some nonverbal memory defects, in consequence, were observed. Using verbal information

### Table 1. Results Obtained in the First and Second Neuropsychological Testing

<table>
<thead>
<tr>
<th></th>
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</tr>
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<tbody>
<tr>
<td>Neuropsi</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total score</td>
<td>91/130</td>
<td>80/130</td>
</tr>
<tr>
<td>Semi-complex figure: copy</td>
<td>12/12</td>
<td>9.5/12</td>
</tr>
<tr>
<td>Semi-complex figure: memory</td>
<td>9.5/12</td>
<td>7.5/12</td>
</tr>
<tr>
<td>Verbal Fluency</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Phonologic (letter F)</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Semantic (animals)</td>
<td>8</td>
<td>9</td>
</tr>
<tr>
<td>Repetition</td>
<td>4/4</td>
<td>4/4</td>
</tr>
<tr>
<td>WAIS</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Information</td>
<td>9</td>
<td>5</td>
</tr>
<tr>
<td>Comprehension</td>
<td>12</td>
<td>4</td>
</tr>
<tr>
<td>Arithmetic</td>
<td>4</td>
<td>3</td>
</tr>
<tr>
<td>Similarities</td>
<td>7</td>
<td>6</td>
</tr>
<tr>
<td>Digits</td>
<td>8</td>
<td>4</td>
</tr>
<tr>
<td>Digits forward natural score</td>
<td>(6)</td>
<td>(3)</td>
</tr>
<tr>
<td>Digits backwards natural score</td>
<td>(5)</td>
<td>(2)</td>
</tr>
<tr>
<td>Vocabulary</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>Digit-Symbol</td>
<td>8</td>
<td>4</td>
</tr>
<tr>
<td>Picture Completion</td>
<td>9</td>
<td>2</td>
</tr>
<tr>
<td>Block Design</td>
<td>9</td>
<td>4</td>
</tr>
<tr>
<td>Picture Arrangement</td>
<td>9</td>
<td>6</td>
</tr>
<tr>
<td>Object Assembly</td>
<td>8</td>
<td>5</td>
</tr>
<tr>
<td>Verbal IQ</td>
<td>86</td>
<td>65</td>
</tr>
<tr>
<td>Performance IQ</td>
<td>103</td>
<td>74</td>
</tr>
<tr>
<td>Full Scale IQ</td>
<td>93</td>
<td>67</td>
</tr>
<tr>
<td>Wechsler Memory Scale</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Logical Memory</td>
<td>11/45</td>
<td>4/45</td>
</tr>
<tr>
<td>Associative Learning</td>
<td>17/30</td>
<td>6/30</td>
</tr>
<tr>
<td>Visual Reproduction</td>
<td>14/14</td>
<td>5/14</td>
</tr>
<tr>
<td>Wisconsin Card Sorting Test</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Categories</td>
<td>6</td>
<td>0</td>
</tr>
<tr>
<td>Total number of errors</td>
<td>83</td>
<td>98</td>
</tr>
<tr>
<td>Perseverative errors</td>
<td>22</td>
<td>25</td>
</tr>
<tr>
<td>Naming Test</td>
<td>42/80</td>
<td>32/80</td>
</tr>
<tr>
<td>WRAT</td>
<td>7/40</td>
<td>7/40</td>
</tr>
<tr>
<td>Ideomotor praxis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Right hand</td>
<td>—</td>
<td>2/11</td>
</tr>
<tr>
<td>Left hand</td>
<td>—</td>
<td>3/11</td>
</tr>
<tr>
<td>Mouth</td>
<td>—</td>
<td>1/4</td>
</tr>
<tr>
<td>Right-left discrimination</td>
<td>correct</td>
<td>2/10</td>
</tr>
<tr>
<td>Fingers</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Naming</td>
<td>abnormal</td>
<td>2/5</td>
</tr>
<tr>
<td>Pointing</td>
<td>abnormal</td>
<td>3/5</td>
</tr>
</tbody>
</table>

Note: Scores in the Neuropsi (excepting verbal fluency, referring to the total number of elements in that category) are “patient score/total score.” Scores in the WAIS (excepting “natural scores” in digits) are scaled scores. Scores in the Wechsler Memory Scale are “patient score/total score.” Scoring in the Wisconsin Card Sorting Test is according to the manual. For the rest of the tests, scores are “patient score/total score.”
spontaneous recall was abnormal (1/6 words), but recognition was normal (6/6). No defects in encoding were observed, but problems in retrieving verbal information were assumed. Her score on the WRAT was again 7/40. Adding and subtracting simple quantities were performed, likely because this ability was intensively trained during the rehabilitation program, but performing any other arithmetical operation was impossible. Ideomotor apraxia was tested by requests

(a) to mimic different movements by oral command or by copying the examiner’s movements (seven items): to throw a ball, to ‘say goodbye’ with her hand, to ‘say come here’ with her hand, to brush her teeth and comb her hair;

(b) to imitate nonsense movements presented by the examiner (four items): to make “horns” with her fingers, to show her fist, to make a circle with her thumb and index finger, and to make a “bunny” with her fingers; and (c) to perform buccofacial movements (four tasks): to whistle, to inflate her cheeks, to blow, and to show her teeth.

Performance in this test was frankly abnormal (see Table 1) indicating an evident ideomotor apraxia. A.R.A., however, continued to recognize the faces of her relatives, friends, and people around her without difficulty.

In spontaneous writing (describing her disease) she reported aloud what supposedly was written, but no word was recognizable (see Figure 2). Letters were well formed and could be easily recognized, but calligraphy had changed in contrast to the first assessment. During the first evaluation, normal cursive writing was used, whereas during this second evaluation, uppercase block letters were used. No clear relationship between the length of the spoken and written to dictation word was evident. Writing words and writing text to dictation had similar characteristics. The patient could report what she was going to write, letters were relatively well-formed, but no word could be recognized. A few self-corrections were noted. Nonetheless, seemingly there was some tendency to alternate consonants and vowels, as most frequently observed in Spanish. Copying, however, was virtually perfect (Figure 3).

Writing to dictation was tested using exactly the same procedure developed in the first evaluation. Initially a short text (28 words) was dictated (Figure 4). Even though the patient repeated what she was going to write, only a weak relationship between what she repeated and what she wrote was observed. For example, the first sentence was La mañana era fría (the morning...
Figure 3. Copying during the first (A) and second (B) evaluations.

Figure 4. Writing by dictation during the first (A) and second (B) evaluations.

Table 2. Words and Pseudowords Reading and Writing (Second Evaluation)

<table>
<thead>
<tr>
<th>Writing to Dictation</th>
<th>Reading</th>
</tr>
</thead>
<tbody>
<tr>
<td>Words</td>
<td></td>
</tr>
<tr>
<td>Ir</td>
<td>Correct</td>
</tr>
<tr>
<td>Sol</td>
<td>Correct</td>
</tr>
<tr>
<td>Tren</td>
<td>TEL</td>
</tr>
<tr>
<td>Venir</td>
<td>GEMIR</td>
</tr>
<tr>
<td>Niño</td>
<td>NISO</td>
</tr>
<tr>
<td>Casa</td>
<td>SASA</td>
</tr>
<tr>
<td>Escribir</td>
<td>ESTIMIR</td>
</tr>
<tr>
<td>Paleta</td>
<td>TAREA</td>
</tr>
<tr>
<td>Caminar</td>
<td>CAEIAR</td>
</tr>
<tr>
<td>Mariposa</td>
<td>MARIOPA</td>
</tr>
<tr>
<td>Papalote</td>
<td>PAGISOE</td>
</tr>
<tr>
<td>Estacionar</td>
<td>UGIOM</td>
</tr>
<tr>
<td>Pseudowords</td>
<td></td>
</tr>
<tr>
<td>Ul</td>
<td>IL</td>
</tr>
<tr>
<td>Pal</td>
<td>PL</td>
</tr>
<tr>
<td>Cron</td>
<td>RIM</td>
</tr>
<tr>
<td>Conir</td>
<td>OMIR</td>
</tr>
<tr>
<td>Teño</td>
<td>EIO</td>
</tr>
<tr>
<td>Bosa</td>
<td>OPA</td>
</tr>
<tr>
<td>Alercibir</td>
<td>ALRIR</td>
</tr>
<tr>
<td>Suleta</td>
<td>POETA</td>
</tr>
<tr>
<td>Chuminar</td>
<td>GOTIMAR</td>
</tr>
<tr>
<td>Turiposa</td>
<td>SIROM</td>
</tr>
<tr>
<td>Dipalote</td>
<td>SIEMOP</td>
</tr>
<tr>
<td>Oltacionar</td>
<td>OSAONA</td>
</tr>
</tbody>
</table>

Note: The words and pseudowords that were used appear in the first column. In the subsequent two columns is presented what the patient wrote and read.
was cold), and the patient wrote LA EINC EL FIEA (nonsense). Only the first word, the feminine article LA, was correctly written. The patient's writing contained 24 letter groups that may be considered to correspond to words.

Writing words and pseudowords to dictation was defective. Two of 12 words and no pseudowords were written. The correctly written words contained only two or three letters (ir—to go, and sol—sun). On longer words, failure occurred. However, reading was easier and only one error in reading words and 10 errors in reading pseudowords was recorded. Lexicalization (reading pseudowords as real words; e.g., bosa—pseudoword as bolsa—bag) was observed in seven items (Table 2). Seventeen errors occurred when reading the short 109-word story history included in the Neuropsi neuropsychological test battery.

A new MRI demonstrated that the brain atrophy had significantly extended and was not anymore limited to the left parietal-temporal area (Figure 5).

Discussion

Cognitive decline in our patient began with (a) difficulties in using Spanish orthography and (b) errors in performing arithmetical operations. Interestingly, her family pointed out that her arithmetical errors were frequent but mild, and a result that was close to the correct answer was provided. This type of approximative responses when solving arithmetical tasks is not unusual in cases of acalculia (Ardila & Rosselli, 2002). Some word-finding difficulties were recorded.

At the time of the first evaluation, arithmetical abilities were at a frankly abnormal level. Verbal fluency and lexical knowledge (according to the WAIS Vocabulary subtest and the Naming test) were decreased. Nonetheless, writing difficulties were almost limited to orthography in spontaneous writing. Calligraphy was appropriate, and written copying and writing to dictation were nearly normal.

The strong association found between orthography and calculation ability is intriguing. Our case supports the hypothesis that both share a common brain organization. It may be conjectured that both depend on some spatial representation mediated through language. Actually, A.R.A. initially presented an incomplete Gerstmann’s syndrome (acalculia, agraphia, and finger agnosia, but no right-left discrimination defects). Gerstmann’s syndrome has been sometimes interpreted as a defect in performing mental rotations and conceptualize of space through language (Ardila, Concha, & Rosselli, 2000; Gold, Adair, Jacobs, & Heilman, 1995). Some authors have proposed that Spanish orthography is, to a significant extent, a spatial ability (Ardila, Rosselli, & Ostrosky, 1996).

Two years later (i.e., approximately 4 years after the start of her symptomatology), the patient had a significant cognitive decline. Mainly verbal but also nonverbal abilities significantly decreased. Memory scores corresponded to a pathological range. Full Scale IQ decreased 26 points, with a similar decline in both the verbal and performance scales. During the initial evaluation, Full Scale IQ corresponded to a normal performance; during the second evaluation it was found to be more than two standard deviations below the mean. At this time, the patient clearly fulfilled the diagnostic criteria for dementia, according to the Diagnostic and Statistical Manual of Mental Disorders (4th ed.; DSM–IV; American Psychiatric Association, 1994). However, she continued driving and going by herself to different places without any apparent difficulty. Copying abilities were well preserved. She could copy figures and words without any significant difficulty. As a result, even though she was technically demented, there was no global cognitive deterioration yet but rather multifunction impairments.

Calculation abilities decreased between the first and the second evaluation, but in the first evaluation, these abilities were already so seriously impaired that no significant further score decrease was anticipated. Scaled
The correct use of orthography (i.e., selecting between
phy was the initial writing difficulty noted in A.R.A.
Evaluation, she wrote to dictation only 2 of 12 words
difficult than letter writing. During the second
test, the patient
and Z) of six letters (D, J, L, V, and Z). When writing
the alphabet, she wrote A, B, C, H, E, D, I, G, J, L, M,
N, Ñ, O, P, S, Y, T, Z.

It is interesting to note that using Spanish orthography
was the initial writing difficulty noted in A.R.A.
The correct use of orthography (i.e., selecting between
two or more homophone alternatives) represents for
normal people the most difficult aspect in writing Span-
ish. It was not surprising to find that it was the most
fragile writing ability. Further evolution (first evalua-
tion) demonstrated not only orthographic (homophone)
errors but also letter omissions and additions and even
nonhomophone errors. Regardless of inability to write
spontaneously or by dictation noted during the second
evaluation, writing by copy was virtually perfect. It can
be conjectured that writing by copy does not really rep-
resent a linguistic ability but rather visuoperceptual and
visuoconstructive ability.

Although in the second evaluation, the patient
clearly presented the major syndromes usually found
associated with left hemisphere pathology (such as
aphasia, alexia, agraphia, acalculia, and ideomotor
apraxia), no major defects usually observed in cases of
right hemisphere damage were evident (such as general
spatial disturbances, prosopagnosia, and topographic
agnosia). Nonetheless, some decline in visuoconstructive abilities was documented between
the first and the second evaluation. The Semicomplex
Figure Copy score decreased approximately 20% (12/12 and 9.5/12), the Block Design score decreased
by more than one standard deviation (scaled score [SS] = 9 and 4), and the Picture Arrangement score de-
creased in one standard deviation (SS = 9 and 6). This
observation may support the hypothesis that in cases of
progressive cognitive syndromes, brain dysfunction ex-
tends from a focus to surrounding areas, eventually in-
volveing the whole brain and resulting in a global cogni-
tive deterioration. During the second testing, our
patient presented with significantly severe defects in
some cognitive domains, whereas other domains were
relatively well preserved. It may be assumed that, in the
future, brain atrophy will become even more extended,
and cognitive deterioration more severe, although some
nonverbal abilities will be the last to become disturbed.

Reading and writing difficulties observed in A.R.A.
have some similarities with other cases of progressive
deterioration of specific cognitive functions previously
reported. Writing difficulties documented in Ardila et
al. (1997) of an English-speaking patient partially cor-
responded to a lexical agraphia, even though alexia was
significantly more severe than agraphia. The inability
to correctly use orthographic rules in Spanish writing
has been interpreted as a lexical agraphia (Ardila et al.,
1996), and, in consequence, the initial writing distur-
bance in A.R.A. can be interpreted as a lexical
agraphia. Further, a severe agraphia was found, even
though the ability to write individual letters was better
preserved. Regardless of the severity of the agraphia

Cognitive deterioration progressed significantly be-
tween the two evaluations. According to the Naming
test and the WAIS Vocabulary subtest, anomia was evi-
dent during the first evaluation and even more severe
during the second evaluation. Thus, since the initial
evaluation, an anomic aphasia was evident. During the
second evaluation, concept-formation difficulties,
ideomotor apraxia, right-left disorientation, and finger
agnosia were observed. Memory abilities during the
second evaluation corresponded to an abnormal range.

Interestingly, copying ability remained relatively
well preserved, and no evident spatial or geographical
orientation difficulties were found. The patient con-
tinued driving and going around the city without any sig-
nificant difficulty. She correctly recognized places and
people, and in consequence, no topographic agnosia or
prosopagnosia were seen. A significant dissociation be-
tween some verbal and some nonverbal abilities was
quite evident.

Several important dissociations were observed in the
second evaluation. They included (a) a dissociation be-
tween reading and writing ability, (b) a dissociation be-
tween spontaneous writing and writing by copy, (c) a
dissociation between ability to read words and pseudo-
words, (d) a dissociation between ability to repeat and to
write, and, most significantly, (e) a dissociation be-
tween verbal and spatial abilities. Some dissociation
between ability to write letters and ability to write
words was also found: Word writing was notoriously
more difficult than letter writing. During the second
evaluation, she wrote to dictation only 2 of 12 words
(the shortest ones, ir and sol) but correctly wrote two (D
and Z) of six letters (D, J, L, V, and Z). When writing
the alphabet, she wrote A, B, C, H, E, D, I, G, J, L, M,
N, Ñ, O, P, S, Y, T, Z.

It is interesting to note that using Spanish orthogra-
phy was the initial writing difficulty noted in A.R.A.

found in the second evaluation, letters were in general clearly recognizable. In a case, Grossman et al. (2001) pointed out that graphemes were typically legible, although letters were written in an inconsistent and heterogeneous manner. We did not note any inconsistency in writing the letters regardless of the task (spontaneous, copying, dictation), but in the second evaluation it was evident that her handwriting had dramatically changed. During the first evaluation, although an evident writing defect (lexical agraphia) was documented, no changes in handwriting were noted. Changes in handwriting from cursive to block letters, as observed in A.R.A., have been reported from cases of right hemisphere pathology (Ardila & Rosselli, 1993). It can be conjectured that the dramatic change in handwriting observed during the second evaluation might be associated with some extension of the brain pathology to the right hemisphere. This hypothesis is congruent with the moderate deterioration of some nonverbal abilities found in the second evaluation.

This case clearly illustrates the progression of focal cognitive defects over time and the spreading of abnormalities to other domains. Progressive focal neuropsychological syndromes can represent an excellent model for the study of brain organization of cognition.

References


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