Cognitive rehabilitation in posterior cortical atrophy

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Cognitive rehabilitation in posterior cortical atrophy

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Posterior cortical atrophy (PCA) is a rare early-onset dementing syndrome presenting with visuo-perceptual deficits. Clinicopathologically, it is most commonly considered a form of Alzheimer’s disease. We present the case of a 64-year-old male patient with posterior cortical atrophy who took part in a cognitive rehabilitation programme that included psychoeducation, compensatory strategies, and cognitive exercises. After the cognitive rehabilitation programme, subtle differences were found in visuoperceptual tasks and in the patient’s subjective perception of difficulties. Cognitive rehabilitation may temporarily improve functioning in patients with posterior cortical atrophy.

**Keywords**: Cognitive rehabilitation; Posterior cortical atrophy.

**INTRODUCTION**

Posterior cortical atrophy (PCA) is a rare early-onset dementing syndrome presenting with visuo-perceptual deficits, which, clinicopathologically, is most commonly considered a form of Alzheimer’s disease (Zakzanis & Boulos, 2001). Unlike typical patients with Alzheimer’s disease (AD), PCA...
patients initially show preserved day-to-day memory, with core symptoms mainly related to visuospatial disorders, including simultagnosia, optic ataxia, oculomotor disorders, visual inattention, topographical disorientation, and deficits in writing, reading and praxis (Caine, 2004). PCA patients may also present with disorders in object and face recognition (De Renzi, 1986) as well as potential reading difficulties that are characterised by letter-by-letter reading and problems in maintaining the focus on a particular line of text (Freedman et al., 1991; Pantel, 1995).

While neuropathological changes in PCA are in most cases similar to those found in AD, the location of said changes seems to differ. In AD patients, early changes occur in the hippocampal formation; however in PCA, these changes tend to affect more posterior parts of the brain (Hof et al., 1993). Most studies report the presence of neurofibrillary tangles and amyloid plaques as typically found in AD, but differing in both their distribution and localisation. In PCA, in particular, primary visual areas and the occipito-temporo-parietal junction are mostly affected, while frontal areas tend to be less affected than in AD (Aharon-Peretz et al., 1999; Ala & Frey, 1996; Hof et al., 1989). On top of AD-like pathology, post-mortem examination of brain tissue of patients with PCA has revealed other neuropathological disorders including diffuse Lewy body disease, subcortical gliosis, corticobasal degeneration, and spongiform changes (Renner et al., 2004).

Regarding neuropsychological performance, patients with this neurodegenerative disorder perform poorly on tasks of visual perception early in the disease (McMonagle, Deering, Berliner, & Kertesz, 2006). Patients present with spared verbal memory with most difficulties arising in visual memory as the disease progresses (Zakzanis and Boulos, 2001). Even if language is considered initially spared, transcortical sensory aphasia has been previously described. Object naming tends to be impaired early in these patients, and comprehension deficits may emerge as the disease progresses. Nonetheless, repetition tends to be spared even in the later stages of the disease (Benson, Davis, & Snyder, 1988; Beversdorf & Heilman, 1998).

Cholinesterase inhibitors represent the standard therapeutic approach to the treatment of early-stage Alzheimer’s disease (AD) and related dementias (i.e., vascular dementia, Lewy body dementia, etc.). However, a proportion of patients experience lack or loss of therapeutic benefit with an initial agent, or discontinue due to safety/tolerability issues. Thus, cognitive training and cognitive rehabilitation constitute methods that aim to help people with early stage dementia.

Despite the fact that studies exploring non-pharmacological interventions in dementia have shown contradictory results, there is growing evidence of its effectiveness in the improvement of at least certain aspects of the disease.
Some methods, such as reality orientation (Spector, Orrell, Davies & Woods, 1998; Woods 2002), spaced retrieval (Camp, 1989), cognitive stimulation (Quayhagen, Quayhagen, Crbeil, Roth, & Rodgers, 1995; Spector, Woods, & Orrell, 2008), face–name association (Clare, Wilson, Carter, Roth, & Hodges, 2002) and errorless learning (Clare et al., 2000) have been shown to be effective in improving cognitive performance.

Most the studies on non-pharmacological treatment have focused on AD and/or vascular dementia, and to our knowledge there are no studies that have investigated the possible benefit of non-pharmacological intervention in patients with PCA.

As mentioned by Clare and Woods (2004), several cognition-focused interventions have been developed to address cognitive dysfunction, particularly in AD, and the terms “training”, “stimulation” and “rehabilitation” have been used interchangeably, even though major differences are currently recognised between these approaches. For instance, while cognitive stimulation involves the participation in group activities, hence facilitating the involvement in cognitive and social activities, cognitive training relies on the repeated and standardised practice of tasks that are supposed to enhance affected cognitive function, which sometimes disregards each patient’s individual needs. On the other hand, the cognitive rehabilitation approach for dementia acknowledges that the changes produced by this condition affect the biological, psychological and social levels, and aims to maximise activity and participation in preferred social activities (Clare and Woods, 2004).

Cognitive rehabilitation was originally developed for people with brain injury, and can be defined as a person-centred approach designed to help people with cognitive impairments “in which those affected, and their families, work together with health care professionals to identify personally relevant goals and devise strategies for addressing these” (Wilson, 2002). This approach has been recently applied to help people with dementia (Clare & Woods, 2001), where the goals are selected collaboratively with patients and their families, and aims to tackle the difficulties that they consider most relevant (Clare and Woods, 2004). The cognitive rehabilitation approach involves everyday situations and problems so that interventions are conducted on an individual basis.

The objective of this study is to describe the neuropsychological and cognitive follow-up of a patient with PCA who underwent a cognitive rehabilitation treatment. Because cognitive rehabilitation attempts to improve functioning in everyday contexts rather than to enhance performance on cognitive tasks as such (Clare and Woods, 2004), outcome measures in the present study were both neuropsychological tests and subjective ratings by the patient and his family in order to capture changes observed both in laboratory testing and everyday real-life functioning.
METHODS

Case study

SS is a 64-year-old male who presented to us with difficulties in object localisation, sporadic spatial disorientation, and difficulties in reading. The patient’s insight of his cognitive problems was appropriate; he was able easily and readily to describe his difficulties and concerns. Previous to his consultation, he had visited two different ophthalmologists, both of whom told him that his vision was completely normal. His first consultation at our Institute was for a cognitive evaluation where visuoperceptual and reading (letter-by-letter) skills were predominantly impaired. Given the major visuoperceptual complaints and symptoms, a specific evaluation was conducted to assess said cognitive area.

SS was assessed with the Birmingham Object Recognition Battery (BORB), which follows the Humphreys and Riddoch model of visual object processing (Riddoch and Humphreys, 1993). This model states that visual processing and recognition are hierarchically organised in a series of processes that range from the derivation of information about basic dimensions of the object – such as size, length, orientation and location – to the achievement of the figure and ground segmentation, and to the later accomplishment of viewpoint-invariant visual information. After achieving this form of precategorical visual processing, the viewpoint-invariant information would allow one to access stored knowledge about object shapes, which are crucial for object recognition. According to this model, SS had difficulties in the precategorical dimension of visual perception: (1) he failed in the derivation of proper information about the object, as evidenced by poor performance on the length, size, orientation and position match tasks of the BORB; (2) he also had problems in posterior figure and ground segmentation, evidenced by deficits in the overlapping figures test of the BORB; and finally, (3) he also showed deficits in the minimal feature view task and in the foreshortened view task of the BORB, suggesting deficits in the achievement of an invariant viewpoint, as revealed by his impaired performance.

Besides the aforementioned deficits, and even if on some occasions he did fail to recognise certain objects, SS did not show major object recognition difficulties in real-life settings. Following the second cognitive assessment, it became clear that SS showed what is described as “dorsal simultagnosia” (Bauer, 1993; Farah & Feinberg, 1997), in which an attentional limitation leads to perception of more than one object at the same time, often resulting in the misidentification of objects due to the fact that the patient’s attention is captured by just one part of an object. This was evidenced in SS’s problems in the overlapping figure test and in the recognition of certain objects and line drawings, which were immediately resolved after his attention was redirected.
Optic ataxia, which is an inability visually to direct movements (Holmes, 1918), was also evident, mainly through SS’s difficulties to reach objects. Oculomotor disorders were also noted, presenting as a complete form of Balint’s and Holmes’ syndrome (Husain & Stein, 1988).

The progressive nature of SS’s condition soon became obvious and he underwent neurological, neuroradiological and neuropsychological assessment. A diagnosis of PCA was made by a specialised neurologist (FM). Magnetic resonance imaging revealed marked enlargement of the occipital horn of the left lateral ventricle with atrophy of the left occipito-temporal cortex (Figure 1).

While SS had no formal higher education, he still showed elevated premorbid intellectual functioning (Word Accentuation Test – Buenos Aires: 41 out of 44 points, 70th percentile). He had been a businessman for most of his life and had retired at the age of 60. His difficulties had become more apparent to himself and his family. On physical examination and neuroimaging it was noted that his posterior brain structures were affected.

SS could not tolerate the side effects of the cholinesterase inhibitors and was referred to the neuropsychology department to undergo the cognitive rehabilitation programme that is the focus of the present investigation. Due to the specificity of his difficulties, an individualised approach was designed to meet the particular needs of the patient.

Assessment

*Neuropsychological assessment.* Neuropsychological assessment was carried out at baseline and after the 10-session programme. SS’s performance was compared with available age-, education-, and gender-matched normative data. Tasks for which baseline vs. after-treatment changes surpassed 1.5 SD based on normative data, were considered to exceed the cognitive
changes expected by pure chance. The neuropsychological assessment included the following measures:

- **MMSE**
- **Addenbrookes Cognitive Examination (Mathuranath, Nestor, Berrios, Rakowicz, & Hodges, 2000).** This simple screening test is designed to detect mild dementia through six cognitive domains: orientation, attention, memory, verbal fluency, language, and visuospatial ability.
- **Rey Auditory Verbal Learning Test (RAVLT; Rey, 1941).** This classical verbal memory test instructs the patient to learn a structured list of words through repetition. Alternative stimuli were used upon re-assessment to avoid practice effects.
- **Complex Rey Figure (Rey, 1941).** This is a classical visuospatial (copy phase) and non-verbal memory (delayed phase) test. An alternative version of the test was used on post-intervention assessment to avoid practice effects.
- **Abbreviated version of the Boston Naming Test (Kaplan, Goodglass, & Weintraub, 1983).** The subject was confronted with 20 of the pictures used in the Boston Naming Test. The pictures were selected based on their frequency of use, ranging from common to rare words. The purpose of the test is to assess the ability to name pictured objects.
- **Token Test (Spreen & Benton, 1977).** We selected the more difficult items of the Token Test, which is part of the Neurosensory Center Comprehensive Examination for Aphasia. The purpose of this test is to assess verbal comprehension of commands of increasing complexity.
- **Semantic Fluency.** This test instructs the patient to produce as many words as possible in a one-minute interval under a semantic rule, in this case: animals.
- **Phonological fluency.** This test instructs the patient to produce as many words as possible in a one-minute interval following a phonological rule, in this case: words starting with the letter “P”.
- **Trail Making Test Part A (Partington & Leiter, 1949).** In this test, patients must connect numbered circles in an ascending order using straight lines, and without raising their pencil. This test is thought to measure sustained attention, graphomotor capabilities, and speed of processing.
- **Trail Making Test Part B (Partington & Leiter, 1949).** This version of the test instructs patients to connect circles containing numbers and letters, in an alternating order (e.g., 1-A-2-B). Because this ability depends more strongly on executive functions, the test is also thought to measure set shifting and flexibility.
- **Digit Span Forward and Backwards (Wechsler, 1991).** Digit Span consisted of two tasks: Forward span requires the subject to repeat
sequences of three to nine digits in the same order as they are presented; backward span sequences are two to eight digits long, and the subject must repeat them in the reverse order.

Subjective rating of everyday function. Private meetings were held with SS and his wife. During these meetings, they were asked separately to rate on a scale from 1 to 10 their own perception of SS’s everyday functioning at baseline and after the treatment.

Procedure

Establishment of treatment goals. Following baseline assessment, a meeting with SS and his wife was arranged in order to determine treatment goals. The nature of the disorder was explained to SS and his wife, strongly emphasising the progressive nature of this disorder and the limited evidence of cognitive rehabilitation as a remediation in dementia. The findings of the neuropsychological examination were discussed with both of them and they were asked to identify SS’s main everyday life problems. A difficulty in locating objects in space was one of the most salient symptoms. SS had trouble finding things around him, which was really embarrassing to him. He also recalled an argument with his wife about a misunderstanding caused by not being able to read a written message left by her. Many concerns raised by SS stem from the fact that he is a very sociable man who enjoys hosting gatherings and dinner parties for his friends and family. For this reason, SS was really disturbed by his inability to serve drinks, mainly because he would miss the glass due to his optic ataxia. After this initial session, the objectives of the treatment were outlined together with SS and his wife.

The treatment goals that emerged from the initial interview included: (1) improving understanding of the disease and its symptoms, (2) diminishing SS’s problems in finding things around him, (3) diminishing difficulties in pouring drinks, and (4) being able to understand written messages left by his family.

Rehabilitation procedure. The cognitive rehabilitation programme consisted of weekly individual sessions lasting 45 minutes. Rehabilitation strategies selected for each objective are displayed in Table 1 and the total amount of weeks dedicated to each is also shown. Besides the weekly session, activities to do at home were also a part of the treatment, and included repeating activities that were being practised at the time. Each rehabilitation strategy was explained to SS in relationship to its goals. SS’s wife was contacted when necessary.
<table>
<thead>
<tr>
<th>Treatment goals</th>
<th>Rehabilitation strategies</th>
<th>Weeks dedicated to each strategy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Improve disease and symptom understanding</td>
<td>• Psychoeducation: These two sessions consisted of a more detailed explanation of SS’s condition. The difference between vision, perception and recognition was discussed including a description of the steps needed to adequately perceive and recognise an object. SS’s test results were discussed in order to show which steps of recognition were affected more prominently. Daily living problems caused by those deficits were then identified. Information about the visual direction of movements was also provided in order to explain to SS his optic ataxia.</td>
<td>2</td>
</tr>
<tr>
<td>Diminishing his problems in finding things around him</td>
<td>• Training object recognition with salient features: SS was trained in the scanning of unrecognised objects and in the localisation and identification of those features that were unique for that object in particular, in order to rely on that feature for recognition. • Visual scanning training: Visual scanning and searching was trained both with computerised and real object stimulus. The main steps of the tasks included (1) fixation on a point, (2) searching for a target embedded among distracters, (3) reaching for the target. Tasks varied on the amount of distracters and targets and size of the scanning area.</td>
<td>5</td>
</tr>
<tr>
<td>Diminishing his difficulties in pouring drinks</td>
<td>• Training in the tactile direction of movements: SS was trained in the use of tactile rather than visual information to direct his movements. This method was first practised during the rehabilitation session and then at home using bottles and cups. • Compensatory strategy: To direct his movements using tactile and not visual information to serve drinks</td>
<td>2</td>
</tr>
<tr>
<td>Being able to understand written messages left by his wife</td>
<td>• Training in letter recognition using salient features: After being trained in object recognition with the use of salient features the same method was applied for letter recognition. • Compensatory strategy: Those important notes must be written in print.</td>
<td>1</td>
</tr>
</tbody>
</table>
RESULTS

Neuropsychological performance

As shown in Table 2, baseline performance revealed severe impairments on tasks of visual attention (Trail A) but within-normal performance on auditory attention (digit span). Visuoperceptual deficits were also present, as observed on the Rey Complex Figure copy score. Language showed no abnormalities at baseline, including tasks of naming (abbreviated version of the Boston Naming Test), comprehension (abbreviated version of the Token Test), and verbal fluency. Initial storage and recall of verbal material was within normal ranges, with more noticeable difficulties emerging when visual stimuli were to be recalled.

As shown in Table 2, tasks that initially tackled visuoperceptual problems showed an improvement demonstrated by better performance on both the Complex Rey figure copy and on the Trail Making test A. However, prominent anomias were detected in the short version of the Boston Naming Test that were not present at baseline. Memory performance remained within the normal ranges in delayed recall and recognition phase throughout the course of treatment.

Regarding the subjective perception of SS’s cognitive state at baseline, SS rated his deficits as less severe than did his wife and the neuropsychologist.

### TABLE 2
SS’s neuropsychological performance at baseline and after treatment

<table>
<thead>
<tr>
<th>Neuropsychological assessment</th>
<th>Baseline</th>
<th>After treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Raw score</td>
<td>Z score</td>
</tr>
<tr>
<td>MMSE (max. 30)</td>
<td>28</td>
<td>-1.38</td>
</tr>
<tr>
<td>ACE (max. 100)</td>
<td>88</td>
<td>0.57</td>
</tr>
<tr>
<td>RAVLT immediate recall (max. 75)</td>
<td>24</td>
<td>-0.48</td>
</tr>
<tr>
<td>RAVLT delayed recall (max. 15)</td>
<td>5</td>
<td>0.57</td>
</tr>
<tr>
<td>Complex Rey Figure: immediate (max. 36)</td>
<td>13.5</td>
<td>-4.10</td>
</tr>
<tr>
<td>Complex Rey Figure: delayed recall (max. 60)</td>
<td>4.5</td>
<td>-1.29</td>
</tr>
<tr>
<td>Abbreviated version of BNT (max. 20)</td>
<td>17</td>
<td>-1.00</td>
</tr>
<tr>
<td>Abbreviated version of the Token Test (max. 26)</td>
<td>26</td>
<td>1.00</td>
</tr>
<tr>
<td>Semantic fluency</td>
<td>12</td>
<td>-1.02</td>
</tr>
<tr>
<td>Phonological fluency</td>
<td>18</td>
<td>1.28</td>
</tr>
<tr>
<td>Trail Making A</td>
<td>124</td>
<td>-7.41</td>
</tr>
<tr>
<td>Trail Making B</td>
<td>135</td>
<td>-1.39</td>
</tr>
<tr>
<td>Digit span</td>
<td>7</td>
<td>0.50</td>
</tr>
</tbody>
</table>

*tasks for which baseline vs. after-treatment changes surpassed 1.5 SD based on normative data.
As shown in Table 2, SS and his wife both noticed the improvement found at the post-treatment neuropsychological assessment.

Cognitive follow up

It has to be noted that after initial improvement, it was decided to continue with the treatment for another 12 months. However, after the initial improvement described above, difficulties increased over time and were evidenced on follow-up neuropsychological assessment. However, SS and his wife continued to perceive subtle but persistent improvements.

DISCUSSION

To our knowledge, this is the first description of cognitive rehabilitation in a patient with PCA. The substantial improvement perceived by the patient, his wife and the neuropsychologist in charge of the cognitive rehabilitation programme suggests the intriguing possibility that cognitive rehabilitation may improve, at least temporarily, everyday problems caused by this disease. This improvement was also shown by a better performance on laboratory testing including a better performance on tests of visual attention and copy of complex figures.

The goal of this study was to describe the treatment of a patient with a diagnosis of PCA who underwent a 10-session cognitive rehabilitation treatment programme. Treatment goals were selected collaboratively with the patient and his family, and aimed to improve performance in the everyday activities that they thought were important. The cognitive rehabilitation programme included psychoeducation, compensatory strategies, and cognitive exercises that were selected to strengthen preserved functions in order to facilitate the compensation of SS’s deficits. As mentioned by Clare and Woods (2004), even if the progressive nature of dementia implies that cognitive rehabilitation goals will change over time, cognitive functioning may be a valuable focus for rehabilitation in the early stages of this condition.

At baseline, SS showed initial deficits in visuoperceptual tasks that were the most concerning to him and were sufficient to affect his daily activities. The difficulties at baseline included deficits in the Complex Rey Figure Test and in the Trail Making Test part A, suggesting visuoperceptual and visual attention difficulties.

After treatment, both SS and his wife observed an improvement in the patient’s everyday functioning. The patient also showed better results on cognitive tests that were initially affected. This benefit cannot be attributed to practice effects because alternative versions of the tasks were always used when appropriate.
It must be noted, however, that the differences found across evaluations were subtle, and a worsening on other areas that were not the focus of the treatment was also found. Anomias that were not evident at baseline rapidly appeared. Moreover, when the treatment was prolonged for another 12 months, difficulties continued to increase over time in the neuropsychological assessment, as one would expect based on the progressive nature of dementia. However, since the objective of cognitive rehabilitation is to improve everyday functioning rather than enhance performance on cognitive tasks (Clare & Woods, 2004), it must be pointed out that SS and his wife continued to perceive a subtle yet persistent improvement during the entire duration of the treatment.

Even though this study illustrates an individual case, this is the first empirical study to examine cognitive rehabilitation in posterior cortical atrophy and our results suggest that improvements can be achieved through cognitive rehabilitation, using psychoeducation, compensatory strategies, and training of preserved cognitive functions in order to reduce excess disability. The use of a combination of cognitive rehabilitation and pharmacological treatments that have shown at least some efficacy in other forms of dementia must be the focus of further studies.

It is not within the reach of the present study to demonstrate the effectiveness of any particular technique of training in PCA, but instead to validate a possible line of approach to rehabilitation that takes into account individual needs of patients with PCA and their families and which may be applicable to this and other forms of dementia. As a single case study, this investigation can only attempt to introduce a possible line of approach on patients suffering from PCA. Further studies including multiple patients, combining cognitive rehabilitation with pharmacological treatments and controlling for the efficiency of the rehabilitation programme will be necessary to establish the role of cognitive rehabilitation in this disease.

**REFERENCES**


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