Progressive impairment of constructional abilities: a visuospatial sketchpad deficit?

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Abstract

The case of a patient, PC, with an impairment of constructional abilities due to a progressive degenerative disease is described. Ideomotor apraxia was also present. PC showed difficulties in all tasks requiring a “manipulation” of a visual model or requiring a visual model to address a specific movement or a choice among alternatives. The spatial component of mental imagery was also severely impaired. The conclusion was that the patient presented a deficit of the rehearsal component of the visuospatial sketchpad (VSSP). This produced an impairment of spatial learning, but spatial retrograde memory was preserved, as well as object recognition. The most prominent sites of atrophy were located in the parietal lobe bilaterally. A 3-year follow-up is reported, showing a progressive impairment of verbal abilities.

Keywords: Constructional apraxia; Perceptual abilities; Imagery; Visuospatial sketchpad

1. Introduction

In recent years, there have been a large number of case reports of patients showing a progressive breakdown in language abilities, with sparing of other cognitive functions [40]. There are also descriptions of progressive dementia in which the prominent features are visuo-perceptual and spatial impairments [12,18,31], as well as two reports of patients with progressive degeneration of the right temporal lobe and progressive prosopagnosia [15,36]; finally, two cases of progressive visual agnosia have been described [9].

Visuo-perceptual impairment is reported in patients with Lewy bodies dementia [26]: these patients are particularly impaired in size and form discrimination, overlapping figure identification and visual counting.

Disorders of constructional abilities are very common in the course of degenerative dementia [17] and they may also be the first symptom [12], or the most severe [21]. However, no attempt has been made in these cases to understand the “functional” locus of the impairment. In contrast, cases with selective cognitive deficits may provide an interesting contribution to the development of cognitive models of visuo-spatial processing. In one of such cases, it has been shown that mental representation of horizontal and vertical spatial relations in an egocentric co-ordinate system are functionally dissociated [21].

There are two main problems regarding constructional apraxia (CA). The first concerns the fact that CA can, indeed, be caused by a variety of cognitive deficits: visuo-perceptual, mental representation, motor implementation or executive disorders. The second problem is related to methodological issues, because CA has been assessed in different ways: copying designs; drawing from memory; arranging sticks in a pattern; building three-dimensional models; block design. Even if all of these tasks require some basic common abilities, they nevertheless differ in other important aspects, such as the load of general intelligence or the involvement of grapho-motor abilities. Recently, the study of CA has become the study of drawing, more precisely of copying. Copying requires different stages to be carried out. Several models have been proposed [20], but essentially, they all include three main stages. During the preliminary stage, a search for an interpretative hypothesis of the model is performed: on the one hand, the individual tries to identify in the stimulus objects that have already been drawn in the past, and at the same time he/she analyses the spatial relationships between elements of the picture and those between the picture and the paper on which it is drawn. During this stage a visual representation of the model is stored in a visual short-term memory system. This stage requires visuo-perceptual abilities and short-term memory. In the second stage, the elements identified by this analysis...
are processed in order to formulate a drawing plan. It has also been proposed that a subject can retrieve separate parts of the model, which are known configurations (such as squares, circles, etc.), stored in a sort of "constructional lexicon" [1]. This stage requires mental imagery and retrieval from long-term memory. Then, in the third, executive stage, the plan is preserved in a short-term memory system for as long as is necessary to complete its translation onto paper, due to activation of motor programmes [20].

Visuospatial short-term memory in constructional tasks has not received much attention, but from the description above, it is evident that a short-term memory system plays a role. I will refer to the visuospatial sketchpad (VSSP) component of working memory (WM) [2]. The VSSP is assumed to maintain and manipulate visual information and to be involved in visual imagery. Logie [23] suggests that the VSSP can be divided in two sub-components (in analogy with the phonological loop): the visual cache and the inner scribe, working in partnership. The visual cache is thought to deal with information that is visual in nature, such as form and colour, and to be closely linked to the activities of the visual perceptual system. The inner scribe, in contrast, is proposed to handle information about movement sequences and, in a manner somewhat analogous to the sub-vocal rehearsal process of the phonological loop, to refresh the contents of the visual cache. It is obvious, therefore, that in case of a damage of the visual cache or the inner scribe, a constructional task would not be performed. Baddeley and Andrade [4] suggest that the maintenance of information by active rehearsal is necessary for imagery. They assume that the regeneration process is part of the function of the slave systems.

A number of studies, both psychological and neuropsychological, now exist to suggest separable subsystems of visuospatial memory for dealing with spatial information (such as the location of an object) and visual information (such as appearance) [13,16,32]. Recent research has hypothesised that WM is not a form of transit lounge that acts to hold perceptual input on its way to long-term memory, but as a workspace [24].

Here, the case of a woman who showed a severe CA, due to a progressive degenerative disease, is described. A mild degree of ideomotor apraxia was also present, while oral apraxia was absent. The functional locus of impairment is discussed in the frame of the WM model [2].

With regard to neural correlates, all data from the literature seem to confirm that CA is generally due to lesions encroaching upon the posterior (parieto-temporo-occipital) regions [17]. In the past, the qualitative differences observed between the constructional performance of right and left brain-damaged subjects led to advance the hypothesis that an executive, planning defect subsumed constructional disturbances in left brain-damaged patients, while visuospatial deficits could be responsible for the disorder in right brain-damaged patients [17]. This distinction, however, has proved to be inconsistent [19].

Recently, four patients have been described with a progressive degenerative pathology, showing early visuospatial problems, agraphia of a predominantly peripheral type and difficulty with bimanual tasks [34]: neuroimaging disclosed bilateral parietal lobe atrophy and hypoperfusion, which was out of proportion to that seen elsewhere in the brain.

Given that CA can result from a variety of deficits (visuo-perceptual, visuospatial short-term memory, mental imagery, motor translation and execution), it seems obvious that lesions in different sites—causing a functional damage in different loci along the processing system can be associated with CA.

A 3-year follow-up is reported, showing in the third year, an initial involvement of verbal abilities. A brief discussion of the clinical aspects of the disease is also given, together with an attempt to clarify the neural substrates of it.

2. Case report

PC was a 52-year-old right-handed woman with 8 years of education, who, in 1998, when she owned a sport shop, started to find difficulty in hanging up suits and in giving change. This last problem was due to a difficulty in grasping the money. In that period, getting dressed also became difficult: for example, she was no longer able to tie her shoelaces. She also misreached when opening the doors. Progressively, she realised that she was unable to cut cakes, to properly dress the table and to read the time on the clock. Finally, an impairment in writing appeared (she was only able to write very slowly in print) and in reading prose passages. In this last task, she was unable to follow the line and to read a word longer than five letters.

2.1. March 1999

In March 1999 PC was submitted to her first neurological examination. She showed no sign of parkinsonism or gaze movement anomalies, including apraxia of gaze or optic ataxia, and the neurological examination was absolutely normal. PC also carried out a formal neuropsychological assessment (see Table 1). She was oriented in time and place, her speech was fluent and her comprehension was normal. In contrast, she showed marked difficulties in all spatial tasks, such as Corsi Block span and supraspan learning, Elithorn’s Perceptual Maze Test and CA. Furthermore, she refused to attempt calculation tasks, because she was unable to follow the right procedures.

She also underwent a unilateral spatial neglect examination, including line bisection, line extension and judgement of length of coloured lines: a moderate degree of spatial neglect of a perceptive type was diagnosed.

Imaging: A CT-scan and a SPECT scan were performed: the first revealed a very mild atrophy, while the SPECT showed a hypoperfusion in the left temporal and parietal lobes and in the ipsilateral caudatus and putamen.
Table 1
General neuropsychological examination

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<tr>
<td>Milan overall dementia assessment (MODA; n.v. ≥ 89.1)</td>
<td>76.7</td>
<td>80.9</td>
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<tr>
<td>Language</td>
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<td>Token Test (n.v. ≥ 29)</td>
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<tr>
<td>Picture naming (n.v. &gt; 80% for the 80-item test)</td>
<td>75/80</td>
<td>72/80</td>
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<tr>
<td>Naming by description (n.v. ≥ 33.5) [28]</td>
<td>37.5</td>
<td>55.8</td>
<td>33.3</td>
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<tr>
<td>Proper names (n.v. ≥ 14.5)</td>
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<td>6.3</td>
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<td>Verbal fluency (phonemic cue; n.v. ≥ 37) [28]</td>
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<tr>
<td>Verbal fluency (semantic cue; n.v. ≥ 25) [28]</td>
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<td>Memory</td>
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<td>Digit span (n.v. ≥ 3.75) [29]</td>
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<td>Corsi span (n.v. ≥ 3.50) [28]</td>
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<td>Visual perception</td>
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<td>Line orientation (n.v. ≥ 17) [6]</td>
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<td>Overlapping figures</td>
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<td>Unknown faces recognition (n.v. ≥ 41) [6]</td>
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<td>Constructional apraxia</td>
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<td>Rey figure (n.v. ≥ 33.24) [7]</td>
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<td>Executive functions</td>
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<tr>
<td>Raven coloured progressive matrices (n.v. ≥ 18)</td>
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<td>16.5</td>
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<tr>
<td>Digit cancellation test (n.v. ≥ 31) [33]</td>
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<td>23</td>
<td>n.a.</td>
<td>n.a.</td>
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</tbody>
</table>

Pathological scores are underlined.

2.2. December 1999–January 2000

Because the impairment of constructional abilities was progressively increasing, the patient was submitted to a second general neuropsychological evaluation as an outpatient, where she came to the author’s attention. Her speech was normal; she was perfectly oriented in time and place and could give correct autobiographical information. PC was also able to cook and to go out shopping alone with no difficulties. She could describe which route she had to follow to go from her town to another. In doing this, she followed a “lexical” strategy. For example, she explained that she had to reach the grocery and then enter the road where a particular tree was, etc., in other words she did not make use of spatial co-ordinates, such as left right. Indeed, the patient reported an increasing difficulty in writing numbers such as 5 or 2, because she could not “remember which way they faced.” Furthermore, when writing, she mixed print and cursive, while oral spelling was normal. A neurological examination did not show any pathological sign, such as rigidity or eye movement deficits.

Neuropsychological examination: During an informal examination, the patient was able to correctly answer questions about her life. Her speech was fluent, without aphasic type errors or articulatory difficulties. In contrast, it took several attempts to hold a pen or sit down.

The patient was administered an extensive battery, including tests of language production and comprehension, executive functions, memory and visuo-perceptual abilities. Results are shown in Table 1.

- **Object picture naming:** In object picture naming [30], PC was able to name 75 out of 80 pictures, 30 belonging to non-living categories and 30 to living categories, 10 pictures of body parts and 10 pictures of musical instruments (mean value for 60 normal subjects 75.4, S.D. 4.32).
- **Fame judgement of people:** When asked to decide whether people were famous or not, PC correctly discriminated 43 famous faces. The errors concerned the pictures of Cosiaga (previous Italian president), Kevin Costner, Einstein, Freud, Muti (Italian director), Scalfari (Italian journalist) and Gheddafi. Five matched control subjects obtained a mean score of 45.20 (range 38–48, S.D. 4.20). PC incorrectly judged, seven unknown people as being famous, reporting that they could be actors, but she could not remember their name.
- **Picture naming of famous people:** She correctly named 28 people out of 50 (adjusted score 31.25). The errors were
either an inability to retrieve the name or semantic errors (for example Verdi was named Beethoven). The score was in the normal range (normal value ≥ 14.51).

- **Token Test** [10]: PC did not show any difficulty in performing the task and her score was in the normal range.

Therefore, PC could correctly perceive and name objects and people’s pictures; she could also correctly point at different colours. In contrast, all tests involving a spatial component were extremely impaired: she was unable to score on tests of visuospatial learning and line orientation judgement. On the Corsi Block task PC could only correctly reproduce a sequence of two, and in a matrix task, she could only recall a $2 \times 2$ matrix in which two of the squares were filled black and two were unfilled. Her ability to copy simple figures, such as a square or a rhombus, was very poor; copying a complex figure was almost impossible: after 10 min all she had drawn was a rectangle, with two lines, one bisecting and another emerging from the rectangle. Spontaneous drawing was also severely impaired; her drawings lacked essential elements or had distorted components: for example, a flower was depicted as a circle, with a vertical line as a stem. No petals or leaves were added. A house was drawn without doors, and with the windows “linked” to the roof. The patient correctly stated whether a line was “short” or “long,” “vertical” or “horizontal,” but then she could not choose the right alternative.

- **Line orientation** [1]: A line stimulus is presented to the subject, who has to choose among four alternatives, which line is the same length as the stimulus. PC was correct two times over 20 trials. The mean score of 14 subjects with the same educational level and approximately the same age (60 ± 14) was 15.7 ± 2.0. The patient correctly stated whether a line was “short” or “long,” “vertical” or “horizontal,” but then she could not choose the right alternative.

- **Position in gap match task** [35]: This is version B of test 5 of the BORB. The subject is shown 40 pairs of circles, which contain a gap in the same position in half cases and in a different one in the other half. The task is to judge whether the gap is in the same or in a different position within the two circles. PC performed correctly on the first two items, but then refused to continue, finding the task too difficult.

- **Discrimination of length** [1]: A line stimulus is presented to the subject, who has to choose, between four alternatives, which line is the same length as the stimulus. PC was correct two times over 20 trials. The mean score of 14 subjects with the same educational level and approximately the same age (60 ± 5.82) was 15.7 ± 2.0. The patient correctly stated whether a line was “short” or “long,” “vertical” or “horizontal,” but then she could not choose the right alternative.

3. Experimental study

3.1. Visuo-perceptual abilities

- **Visual exploration** [1]: (i) The patient had to count all the dots on three different sheets of paper, containing, respectively 14, 34 and 36 dots. PC scored, respectively 11, 13 and 21. Omissions occurred in different positions, even centrally, and were not due to hemispatial neglect. (ii) In a second task, the patient was presented with three sheets of paper, each containing eight lines of letters. The task was to count all the A, H, and F, respectively, on each sheet. Scores were: 11 out of 21 for A; 8 out of 15 for H; 5 out of 8 in case of F. Again, omissions were observed in any position on the sheet.

- **Form comparison** [35]: PC was presented with 32 pairs of scribbles (16 identical and 16 different) and asked to decide whether they were identical or different. She scored 21/32, which is at the lower limit in the normal range. Errors were made on identical items, which were considered as different.

- **Length comparison** [35]: The patient was presented with 32 pairs of vertical lines, 16 “same” and 16 “different.” As in the previous test, the task is to judge whether the segments of each pair are “same” or “different.” PCs score was 21 (normal values ≥ 18), which corresponds to a normal performance.

- **Position in gap match task** [35]: This is version B of test 5 of the BORB. The subject is shown 40 pairs of circles, which contain a gap in the same position in half cases and in a different one in the other half. The task is to judge whether the gap is in the same or in a different position within the two circles. PC performed correctly on the first two items, but then refused to continue, finding the task too difficult.

- **Identification of letters** [33]: The letters of test 6 of the BORB, presented as single stimuli, were used. PC was able to read all letters per sheet in 10 s. The worst of eight control subjects did it in 11.2 s. Thus, PCs performance was considered to be normal.

- **Identification of overlapping stimuli** [33]: This was assessed relative to non-overlapping stimuli. This test corresponds to test 6 of the BORB: only pictures were used. The patient was asked to name the items as quickly as possible in three conditions: presented as single stimuli, as paired non-overlapping and paired overlapping. Performance was timed in all cases. PC took 20 s per sheet to name single stimuli, 30 s per sheet to name paired
non-overlapping stimuli and 2 min to name each item of paired overlapping stimuli. Moreover, in this last condition, the patient was able to correctly name only 12 pairs, as she misinterpreted the second stimulus of a pair; for five pairs, she identified and named only one stimulus, and for two pairs, none. Eight controls (mean age 69.8) took 14.5 s per sheet to name single stimuli; 21.5 s per sheet to name paired non-overlapping stimuli and 23.9 s per sheet (1 s per item) to name paired overlapping stimuli. The lowest control score for single stimuli was 40.9 s per sheet, 49.9 s per sheet for paired non-overlapping stimuli and 2.8 s per item for paired overlapping stimuli.

3.1. Discussion

PC presented with a severely impaired performance in visuo-perceptual tasks involving a “spatial” component. Occasionally she seemed to show simultanagnosia, because increasing the number of stimuli affected performance. However, when informally tested with a picture representing a meaningful scene, she was perfectly able to describe it. Moreover, PC recognised and correctly used real objects, and watched television. When presented with the stimulus and the four alternatives, she was able to count and to state that they were different. In contrast, she was unable to “hold” the stimulus in order to allow a comparison with more than one single item. Even in identifying stimuli in the overlapping condition, she was able to detect the first one, but often not the second, as if she could not abstract the first shape and hold it in a visuospatial store, to differentiate the other one. The deficit was not due to hemineglect, because, as already mentioned, in visual exploration she failed to count dots or letters in any position. In contrast, a type of inattention, the patient was able to correctly name only 12 pairs, and 2.8 s per item for paired overlapping stimuli. PC was completely unable to perform this task and she answered at chance, considering as wrong correctly rotated letters and as correct wrongly rotated ones. As already mentioned, one of the first problems she presented with was the performance of five matched controls. However, she was definitely impaired in both mental rotation tasks. It seems that the patient cannot perform tasks used to argue for the spatial nature of the imagery. Letter and Form Rotation Tests require retention and manipulation of visual material in an imagery condition. This assumption will be discussed later in detail.

3.2. Mental imagery

The patient was then submitted to a battery of tests of mental imagery including:

- **Letter rotation** [16]: Five asymmetrical capital letters (F, G, B, K and R) were presented four times each, twice normal and twice mirror reversed, for a total of 20 trials. The letters were oriented at 45, 90, 135 and 180° of angular displacement from the upright and the task was to say whether each letter was normal or mirror reversed. PC was completely unable to perform this task and she answered at chance, considering as wrong correctly rotated letters and as correct wrongly rotated ones. As already mentioned, one of the first problems she presented with was the performance of five matched controls. However, she was definitely impaired in both mental rotation tasks. It seems that the patient cannot perform tasks used to argue for the spatial nature of the imagery. Letter and Form Rotation Tests require retention and manipulation of visual material in an imagery condition. This assumption will be discussed later in detail.

- **Figure rotation** [1]: The subject had to select among four alternatives (meaningless figures), the item, which, if rotated, was identical to the test stimulus. In this task, PC was unable to give a single answer. Fourteen normal subjects of the same educational level had a mean score of 5.29 out of 10.

- **Mental Colour Comparison Test** [14]: Fifteen pairs of colour-specific objects were read aloud, with the respondent being instructed to say whether or not they were chromatically similar (for example: whipped cream/white or red). The patient’s performance was errorless. The average score of five controls was 14.4.

- **Naming colours of objects**: Twenty objects (such as blood, sky, grass, snow, etc.) were proposed verbally, with the respondent being instructed to name the colour. PC’s performance was errorless, as was that of five matched controls.

- **Size comparison** [16]: This task involved judging which of two similar-sized items was bigger. Sixteen pairs were selected (for example: egg/olive) and their names were read aloud to the patient. Again her performance was errorless, as was the performance of five matched controls.

- **Animal tails** [16]: Eight pairs of animals were selected and PC was asked which of the two had a longer tail (for example: cow/sheep). Her performance was errorless, as was the performance of five matched controls.

3.2.1. Discussion

PC showed a preserved performance in a task assessing colour imagery and in the size and tail comparison tasks. Therefore, her ability to retrieve visuospatial information from long-term memory is clearly unimpaired. However, she was definitely impaired in both mental rotation tasks. It seems that the patient cannot perform tasks used to argue for the spatial nature of the imagery and can perform tasks used to argue for the visual nature of the imagery. Letter and Form Rotation Tests require retention and manipulation of visual material in an imagery condition. This assumption will be discussed later in detail.

3.3. Central organisation of the task

None of the tasks of this section (complex figure identification, embedded meaningless figure detection, and mental construction) could be completed. The patient did not give any answer and refused to continue the test.

3.3.1. Discussion

These tasks are designed to verify how information is integrated in order to plan a procedural strategy to solve the task. PC cannot analyse and search for configurations inside a picture, and therefore she cannot achieve any information to use for planning. This procedure needs the information to be maintained in a WM system.
3.4. Motor execution

The patient was asked to draw a line joining two dots, arranged vertically, obliquely, horizontally at different distance from each other. PC correctly performed the task, which initially seemed to be easier when the dots were at a considerable distance from each other, relative to closer dots. However, this finding was not constant in repeated sessions, and therefore will not be further discussed. PC was also able to connect a third dot.

3.4.1. Discussion

This proved to be the least impaired component in processing constructional tasks. PC was trying some rehabilitation techniques, and she seemed to be able to perform satisfactorily only the task of connecting dots on a sheet.

3.5. Reaching

One of the patient’s earliest symptoms had been the inability to grasp the right money, to tie shoelaces or to hang clothes. This suggested a misreaching problem. Therefore, PC was submitted to a formal examination of reaching. She was asked to touch, with her right (and subsequently left) index, the index finger of the examiner, put in different points of her visual field. Twenty trials for each hand were performed. She never missed the target. Subsequently, she was asked to point to 20 dots (presented one at a time) on a sheet of paper A3, both with the right and the left index finger. Again, her performance was errorless. This ruled out the possibility that some of the deficits in PC’s performance could be explained with a response-related disorder.

3.6. Visuospatial memory

As already reported, Corsi Block span and Brooks matrix were severely impaired. The patient could only reproduce sequences of two blocks and recall a $2 \times 2$ matrix.

As far as long-term memory was concerned, PC was able to find her way in the town where she lived (and had always lived) and in her house. She went shopping and also visiting her friends alone. However, she could not remember her way back from the testing room, even if she perfectly recognised the examiner and she distinguished the same from a different room. Her spatial difficulties occurred only with locations with which she was unfamiliar, similarly to patient ELD [22]. Perhaps in the case of new, unfamiliar locations, PC had not yet developed the “lexical” strategy described above and she had to rely on spatial co-ordinates, which were severely impaired.

3.7. Reading

PC was asked to read 20 single letters, 20 high-frequency short (such as CANE, Italian for dog) and 20 high-frequency long words (such as ZUCCHERO, Italian for sugar). These were in printed black capital letters on a white board. The patient was able to correctly read all of them, but while the task was performed at a normal speed for letters and short words, time for long words varied between 20 and 30 s each. For example, it took about 20 s to read a word such as BICCHIERE (Italian for glass). Finally, she could not read a passage of prose.

Imaging: A perfusion MRI revealed a mild-moderate degree of biparietal hypoperfusion.

4. Follow-up


On 10 July the patient was reassessed on a selection of tests in order to verify the progression of her disease. A further examination continued in September. The results of the two sessions are reported together. At informal examination, PC was able to describe the major events in her life and the evolution of her disease. She was still completely autonomous. Her daily activity had not changed and she was accomplishing the same tasks as before. In particular, she was still shopping alone and had no problem in finding her way in her town or in her house. Her speech was fluent, but the same words were used far often, and many passe-partout sentences were present. However, PC did not complain about word-finding difficulties and, even when her attention was focused on it, she denied that her language somehow become poorer. No articulatory problems or phonological errors were produced.

The patient performed the following tests (see Table 1):

4.1.1. Visual perceptual abilities

- Street Completion Test [35]: PC was able to identify only one stimulus out of 14.
- Line Orientation Test [6]: She did not produce any correct answer.
- Face Recognition Test [6]: The patient’s performance had further deteriorated on this task.
- Overlapping figure identification [33]: Again her performance on this task had dramatically worsened. The patient distinguished only one of two overlapped stimuli, while she was still able to name paired non-overlapping ones.

4.1.2. Constructional apraxia

PC was again evaluated on a battery for CA [1]. On this occasion, however, considering that the patient had been unable to perform most of the battery during the previous session, a simplified version was prepared. Only two alternatives, instead of four, were shown to the patient. PC obtained the following scores: line size: 12 out of 20; line orientation: 0 out of 20; angle width: 3 out of 10; dots position: 3 out of 12. No other tasks could be accomplished, because...
the patient reported that she had no idea on how to proceed. During the test, she described the size, orientation, or position of the stimulus, but then she was unable to find the corresponding one, between two alternatives. For example, she described the stimulus as long and she reported that she had to choose the longer one, but then, in front of the two alternatives, she chose the shorter pretending that it was the one identical to the stimulus. In the dot position task, she was able to perform the task with only a single dot inside the square, but when there were two dots, she protested that she was getting “confused.”

4.1.3. Visuospatial memory
PC could not remember even a sequence of two blocks. She was able to tap a single block. Therefore, she was not submitted to visuospatial learning. Also in a matrix task she could not remember any pattern. This contrasted with her ability to remember familiar routes and faces.

4.1.4. Language and verbal memory
Other cognitive domains, such as verbal memory and language, were explored, to verify whether the impairment was still circumscribed to constructional abilities. While verbal memory, both short- and long-term, proved to be in the normal range, as well as controlled association by phonemic cue, controlled association by semantic cue was now mildly impaired (see Table 1). In a semantic battery (Miceli and Capasso, unpublished data), PC produced 10 visual errors. This happened with lower-frequency objects, such as organ or piers. This battery, which contains pictures larger with respect to the previous version, was used in order to reduce perceptual difficulties. Also increasing the number of stimuli seemed to provide a more sensitive test for the suspected initial language impairment.

4.1.4.1. Discussion. It was now evident that visuo-perceptual and constructional abilities had further worsened and they were now at a floor level. In addition an initial impairment of language abilities, and particularly of its lexical-semantic aspects, was appearing. Other linguistic features were preserved, such as phonology and syntax.

Imaging: A SPECT showed a biparietal hypoperfusion, more evident on the right side, without a significant change from the previous one (March 1999).

4.2. May 2001
The patient was seen again 8 months later. Both, the patient herself and her husband, reported a serious progression of the disease and PC was now aware of word-finding difficulties. Moreover, she could not remember the day of the month and, occasionally, also her birthday.

Her speech was indeed very poor as far as communicative content was considered, but was still produced with normal prosody and without articulatory deficits. No phonological or semantic paraphasias were found in spontaneous speech. PC took care of the house, cooked and solved cross-words; she went shopping alone. She did not show anterograde or retrograde memory deficits; she recognised people and common objects and did not show apraxia of use. She correctly chose dresses according to the season (she had only a mild difficulty with some wearing, such as bra or shoelaces), she did not suffer behavioural changes or depression.

PC was re-submitted to a neuropsychological examination, including tests of language, apraxia, and verbal memory. Results are presented in Table 1. PC was now severely impaired in naming famous people: she recognised, but she could not name, many famous faces. Also picture naming was mildly impaired: she produced not only visual errors, but also five semantic paraphasias (such as JAGUAR for TIGER; HEEL for ANKLE) and one circumlocation. Phonological errors were absent. Naming by description was still in the normal range, but her performance had slightly worsened since the first examination. Verbal long-term memory was also impaired, probably due to her language difficulties. Oral apraxia testing was errorless, while the patient showed ideomotor apraxia, with performance on the left being mildly poorer than on the right (35/72 and 40/72, respectively). She was able, however, to describe what the examiner was doing (for example: touching her front with her hand), but she could not imitate. Visuo-perceptual tests, such as the Street Completion Test or tests from the CA battery, even in the shortened version with only two alternatives, were impossible to perform and the patient, after a few attempts, asked to stop. For that reason, tests of screening for dementia were not performed, due to the amount of visual information shared by almost all the sub-tests. By contrast, connecting two dots, in different positions, with a vertical, horizontal or oblique line was still possible. Imaging: An MRI showed an enlargement of both lateral ventricles and atrophy of the parietal cortex bilaterally.

5. General discussion
The case of a 52-year-old woman with a progressive impairment of visuo-perceptual and constructional abilities is reported. Later in the progression of the disease, a lexical-semantic deficit appeared. PC does not show the same pattern of impairment as the patients reported by Delia Sala et al. [12] she identifies objects and finds her way in the streets and in her house, because she recognises buildings, furniture and other possible visual cues. For the same reason, she did not present with a typical biparietal syndrome, as the one described by Petersen [31]. Single aspects of her performance will be now examined.

5.1. Visual perception
The patient could not judge line orientation, nor decide which of the four squares contained two dots in the same position as in a target stimulus, even if she was able to give a
description of the stimuli. She needed an incredible amount of
time to distinguish the second figure of paired overlapping
ones. However, she could hold other perceptual tasks, such as
perceiving and naming single line drawings of ob-
jects as quickly as controls; naming people’s pictures; com-
paring pairs of scribbles and lines, reading letters and short
words; distinguishing colours, such as in the Token Test.

In other words, it seems that PC could correctly perceive
the stimulus, especially if this could also be retrieved from
long-term memory (as in case of objects and people), but
was impaired when she had to hold and manipulate its re-
presentation in a visuospatial short-term memory system, in
order to make a comparison, or to program a response or a
movement.

5.2. Mental imagery

PC could not draw from memory and tasks requiring men-
tal manipulation were dramatically impaired. In contrast,
mental imagery involving long-term knowledge and a ver-
bal answer, such as naming colours of objects, comparing
brightness of colours or size of objects was very good. A ver-
bal report does not need the involvement of visual short-term
memory (cherry red-blood red → same, olive small-egg big-
ger → egg). This can be achieved directly from a long-term
visual store, bypassing the VSSP. Patients with dissociation
between visual and spatial mental imagery have been already
described. For example, Luzzatti et al. [25] reported the case
of a patient with a dissociation between preserved visual and
impaired spatial processing: this subject was able to repre-
sent objects, but she failed in any task requiring mental rota-
tion, recall of spatial position, or execution of spatially based
imagery operation, as PC did. The opposite pattern (deficit
of visual imagery with intact spatial processing) is also re-
ported [16]. There is also evidence that visual perception
and visual imagery can dissociate. Bartolomeo et al. [5] re-
ported a patient who, following a bilateral temporo-parietal
lesion, showed a severe visual perception impairment, but a
preserved visual mental imagery.

5.3. Motor implementation

The patient was able to connect dots, regardless of their
position and reaching proved to be normal. In contrast, she
could not perform a task when she had to hold a visual im-
age throughout the execution, such as in copying or repro-
ducing motor patterns, as in ideomotor apraxia. It could be
the case that with a simple movement, she could rely on
kinaesthetic information. Alternatively, if the spatial image
is very simple, it can be retained in order to control the
movement, without the need of rehearsal.

5.4. Visual and spatial short-term memory

PCs performance on visuospatial short-term memory
tasks proved to have deteriorated since March 1999, and

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to parieto-occipital lesions: this component was damaged in the patient.

5.4.2. Clinical aspects

It is not clear which degenerative disease was at the basis of PC's impairment. The age of onset argues against a diagnosis of probable AD, as does the absence of memory deficits. Moreover, the presence of an impairment of constructional abilities is uncharacteristic of Pick's disease. Neurological examination ruled out the possibility of cortico-basal degeneration.

6. Conclusions

In conclusion, PC seems to show, at least at the beginning of the disease, an impairment of the VSSP, in particular of the process of rehearsal, the so-called inner scribe. This can explain the impairment of all tasks, such as visual judgements, spatial mental imagery and control of movements, associated with unimpaired verbal description of stimuli and preserved simple perceptive tasks. PCs CA, which involves both, copying and spontaneous drawing, could then be attributed to a functional deficit at this level of processing.

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