Cognitive and motor functioning in a patient with selective infarction of the left basal ganglia: evidence for decreased non-routine response selection and performance

Angela K. Troyer a,∗, Sandra E. Black b, c, d, Maria L. Armilio a, c, e, Morris Moscovitch a, c, e

a Psychology Department, Baycrest Centre for Geriatric Care, 3560 Bathurst Street, Toronto, Ont., Canada M6A 2E1
b Division of Neurology, Sunnybrook and Women’s College Health Sciences Centre, Toronto, Ont., Canada
c Rotman Research Institute, Baycrest Centre for Geriatric Care and University of Toronto, Ont., Canada
d Department of Medicine, University of Toronto, Toronto, Ont., Canada
e Psychology Department, University of Toronto, Toronto, Ont., Canada

Received 1 May 2003; received in revised form 1 December 2003; accepted 3 December 2003

Abstract

Focal damage to the basal ganglia is relatively rare, and little is known about the cognitive effects of damage to specific basal ganglia structures. A 28-year-old, highly educated male (patient RI) sustained a unilateral left ischemic infarction involving primarily the putamen and secondarily the head of the caudate and the anterior internal capsule. Two detailed neuropsychological assessments, at 3 and 16 months post-infarction, revealed that a majority of cognitive abilities were spared. RI’s general intelligence, simple attention, concept formation, cognitive flexibility, and explicit memory were unaffected. Select cognitive abilities were affected, and these appeared to be related to direct involvement of the putamen and/or to indirect disruption of circuits between the basal ganglia and frontal lobes. Consistent with involvement of the left putamen, RI showed micrographia with his right hand. Interestingly, his micrographia was context-dependent, appearing only when verbal expression was involved (e.g., present when writing spontaneously, but not when copying sentences or when drawing). Evidence of disruption to frontal systems included variably decreased sustained attention, mildly decreased ability to generate words and to generate ideas, and significantly impaired abstraction ability in both verbal and visual modalities. Although there are several possible interpretations for these findings, this pattern of cognitive and motor functioning is consistent with neuroimaging research suggesting that the frontal/subcortical circuit between the putamen and frontal motor areas plays a role in non-routine response selection and performance.

Keywords: Putamen; Neuropsychology; Cognition

1. Introduction

The basal ganglia comprise the caudate nucleus, putamen and globus pallidus (i.e., lentiform nucleus), substantia nigra, and subthalamic nucleus. The traditional view of the role of the basal ganglia is that they serve to gather information from widespread cortical regions and channel it to the ventrolateral thalamus and on to motor cortex. However, the delineation of five parallel frontal–subcortical circuits (i.e., skeletomotor, oculomotor, dorsolateral prefrontal, lateral orbitofrontal, and anterior cingulate; Alexander, DeLong, & Strick, 1986) served to extend the spectrum of involvement of the basal ganglia in behavior to include higher cognitive functions (Cummings, 1993).

In this paper, we report the cognitive findings from a patient, RI, who sustained an infarction primarily affecting the left putamen. The putamen is part of the skeletomotor circuit, which projects to the supplementary motor area, premotor cortex, and motor cortex (Alexander et al., 1986; Cummings, 1993). Given the connections between basal ganglia structures and the frontal lobes, one would expect RI’s infarction to be associated with both direct effects due to putaminal dysfunction as well as indirect effects due to dysfunction of the skeletomotor circuit and consequent effects on frontal regions. There are few reports pinpointing the functions of the separate frontal/subcortical circuits, particularly non-motor functions. Our case study demonstrates specific motor and non-motor functions of the skeletomotor circuit.

Previous research has revealed several motor and cognitive difficulties that appear to be related directly to damage...
to the basal ganglia rather than indirect involvement of the frontal lobes. A prime example is micrographia, which results from damage to the basal ganglia (Martinez-Vila, Artieda, & Obeso, 1988; Münchau et al., 2000; Pullicino, Lichter, & Benedict, 1994), but is not associated with focal damage to the frontal lobes. In addition, previous research with the patient described in the present paper (RI) indicated that putaminal damage was associated with intact explicit recollection and impaired implicit habit learning, whereas focal frontal damage was associated with the opposite pattern (Hay, Moscovitch, & Levine, 2002), underscoring a role for the basal ganglia in implicit memory.

There is considerable evidence that damage to the basal ganglia results in indirect effects to frontal brain regions. For example, bilateral lesions restricted to the lentiform nucleus, as documented by structural neuroimaging, are associated with relative hypometabolism in frontal-lobe regions, as seen on functional neuroimaging (Laplane et al., 1989; Strub, 1989). Thus, one would expect basal ganglia damage to be associated with cognitive deficits similar (but not identical, see above) to those found with focal frontal lesions (i.e., impairments in sustained and selective attention, working memory, retrieval of information from long-term memory, memory for source and context, verbal fluency, with phonemic fluency generally worse than semantic fluency, and executive abilities such as concept formation, mental set shifting, inhibition of responses, and susceptibility to interference; summarized in Stuss, Esks, & Foster, 1994). Indeed, neurological disorders that involve degeneration of input nuclei of the basal ganglia circuitry, such as Parkinson’s disease and Huntington’s disease, are associated with impairments in some of these functions (Brandt & Butters, 1996; McPherson & Cummings, 1996).

The most direct way of determining the role of the basal ganglia in cognition is to study the effects of circumscribed damage to these structures, although there are few opportunities to do this because lesions restricted to selected regions of the basal ganglia are relatively rare. As such, little is known about the impairments associated with damage to these structures alone, and even less is known about damage to individual basal ganglia structures. A range of deficits associated with isolated basal ganglia damage has been documented, but for the most part, these studies report deficits on gross measures of cognition, sensorimotor function, and affect (Caplan et al., 1990; Pardal, Micheli, Ascanape, & Paradiso, 1985; Stein et al., 1984). In general, caudate lesions are associated with a broad range of cognitive deficits, including decreased intelligence, memory, verbal fluency, and language abilities (Crosson, 1992; Mendez, Adams, & Skoog Lewandowski, 1989; Petty, Bonner, Mourtagalou, & Silverman, 1996; Pozzilli, Passafiume, Bastianello, D’Antona, & Lenzi, 1987; Richfield, Twyman, & Berent, 1987).

In contrast to the caudate nucleus, lentiform structures are more closely involved in motor functioning. Lesions to lentiform structures in general and the putamen in particular are associated with motor disorders such as dystonia (Bhatia & Marsden, 1994) and micrographia. Although many reports do not specify whether the micrographia is unilateral or bilateral (Martinez-Vila et al., 1988; Pullicino et al., 1994), one recent case indicated contralateral micrographia following unilateral lesions in the globus pallidus (Münchau et al., 2000). Lentiform structures are also involved in the cognitive aspects of motor activities, such as learning and planning new sequences of actions (Jueptner, Frith, Brooks, Frackowiak, & Passingham, 1997; Jueptner et al., 1997). Similarly, functional imaging studies using the Wisconsin Card Sorting Test show that the putamen is involved during performance of an action under a new response set following the reception of negative feedback (Monchi, Petrides, Pette, Worsley, & Dagher, 2001). This suggests a role for the putamen in guiding actions (especially those that are novel) according to behavioural rules.

There are few reports of other cognitive effects of damage to lentiform structures. The most prominent cognitive impairments associated with bilateral or unilateral lentiform lesions tend to be related to the motor aspects of language (e.g., fluency and naming; Laplane et al., 1989; Pullicino et al., 1994; Warren, Smith, Denson, & Waddy, 2000). Bilateral lentiform lesions are associated with greater deficits in phonemic fluency than semantic fluency (Haaxma et al., 1993; Laplane et al., 1989), similar to the pattern of patients with focal frontal lesions. The effect of lentiform damage on explicit memory is inconsistent, with some patients showing intact explicit memory (Hay et al., 2002; Laplane et al., 1989; Pickett, Kuniholm, Protopapas, Friedman, & Lieberman, 1998; Warren et al., 2000) and others showing memory deficits ranging from mild to severe (Haaxma et al., 1993; Laplane et al., 1989; Pullicino et al., 1994; Strub, 1989), regardless of whether the damage was unilateral or bilateral. When explicit memory deficits are present, they involve poor acquisition and normal retention (Haaxma et al., 1993), although it has not been determined at which stage deficits occur on implicit memory tests (Hay et al., 2002). Surprisingly, detailed assessment of frontal/executive abilities has been completed in only a few cases of focal damage to the lentiform nucleus. In one case study, damage to the left lentiform nucleus was associated with deficits in verbal abstraction on a test of similarities, although unspecified “frontal executive and motor functions” were reported to be normal (Warren et al., 2000). Other studies of frontal/executive abilities have shown impaired reasoning and shifting on various clinical tests, including the Wisconsin Card Sorting Test, the Tower of London, and the Stroop task (Haaxma et al., 1993; Strub, 1989). Given the significant connections between the basal ganglia and the frontal lobes, more detailed studies are needed of frontal/executive deficits following damage to the distinct structures in the basal ganglia.

Because little information has been reported about the cognitive consequences of damage restricted to the...
lentiform nucleus, the purpose of the present study is to report and discuss the findings of a detailed neuropsychological evaluation of a patient with a focal lesion affecting primarily the left putamen with some extension to the anterior internal capsule, a small portion of the caudate, and external globus pallidus. Repeated testing, at both 3 and 16 months post-infarction, allowed us to determine the acute and long-term effects. The comprehensive testing included assessment of abilities sensitive to lesions in both frontal and non-frontal regions, which allowed us to examine the indirect and direct effects of his basal ganglia lesion.

2. Case report

2.1. Background

RI is a 28-year-old male who had recently completed a professional degree in a health-related field and was pursuing post-graduate training at the time of the infarction. He had no prior known medical problems. The acute signs of the infarction were right hemiparesis of the arm and leg and inability to speak upon awakening in the morning. There was also evidence of confusion at onset, for example, despite awareness of these problems, rather than seeking help, he frantically began packing a suitcase. He was able to understand and to gesture responses to questions asked of him by paramedics who arrived shortly after the onset of these problems. He experienced nausea and had patchy memories for 2–3 days following the infarction. By the second day post-infarction, he regained some ability to speak, and by the third day, he was able to walk. His symptoms improved rapidly, and he returned to school within 3 months. Two years after his stroke, he had completed a post-graduate fellowship that included a master’s degree and planned to start professional degree in a health-related field and was pursuing post-graduate training at the time of the infarction. He had no prior known medical problems. The acute signs of the infarction were right hemiparesis of the arm and leg and inability to speak upon awakening in the morning. There was also evidence of confusion at onset, for example, despite awareness of these problems, rather than seeking help, he frantically began packing a suitcase. He was able to understand and to gesture responses to questions asked of him by paramedics who arrived shortly after the onset of these problems. He experienced nausea and had patchy memories for 2–3 days following the infarction. By the second day post-infarction, he regained some ability to speak, and by the third day, he was able to walk. His symptoms improved rapidly, and he returned to school within 3 months. Two years after his stroke, he had completed a post-graduate fellowship that included a master’s degree and planned to start professional practice.

Detailed neurological, neuroradiological, and neuropsychological evaluations were undertaken. RI’s consent to participate in these clinical and research procedures was obtained, consistent with the institutional research ethics committees. In this report, RI’s actual initials have been changed to protect his confidentiality.

2.2. Neurological findings

Neurological examination 6 weeks post-infarction revealed a mild amnesia and decreased finger dexterity in the right hand as assessed by finger/thumb opposition. Neurological function was otherwise normal. At 6 months post-infarction, he had persisting mildly decreased finger dexterity and, over the previous few months, had developed micrographia when trying to write or take notes with his right hand. There were no apparent increased tone in the right arm or bradykinesia of other arm movements. At 1-year post-infarction, neurological function was unchanged.

An ischemic infarct with some hemorrhagic transformation was diagnosed and was attributed to a paradoxical embolism through an atrial septal defect after extensive investigation for stroke etiology. He was reluctant to take warfarin and was treated with aspirin for stroke prophylaxis.

2.3. Neuroradiology

Magnetic resonance imaging, obtained on a 1.5 T MR unit (Sigma, General Electric Medical Systems, Milwaukee, WI), included a spin echo sequence (TR/TE 2000/30-0), which provided forty-eight 3 mm thick interleaved slices, and a T1-weighted three-dimensional volumetric sequence (TR/TE 5/24/1, 5° flip angle and 1.3 mm slice thickness). The three-dimensional images were realigned parallel to the anterior–posterior commissure line (Talairach & Tournoux, 1988) using ANALYZE AVR™ software (Biomedical Imaging Resource, Mayo Foundation, Rochester, MN) on a Sun workstation (Sun Microsystems, Mountain View, CA).

Axial T1-weighted and spin echo images taken about 1-year post-infarction showed a well-demarcated lesion in the left putamen consistent with encephalomalacia (Fig. 1). The lesion also involved the external segment of the left globus pallidus, whereas the internal segment of the globus pallidus medial to the putamen was spared. The lesion extended across the anterior internal capsule into the ventral head of the left caudate resulting in enlargement of the left frontal horn of the lateral ventricle (Fig. 1a and c).

2.4. Neuropsychological findings

A comprehensive neuropsychological assessment was performed 3 months post-infarction and was repeated about 1 year later, at 16 months post-infarction. Subjective cognitive complaints at the initial assessment included slowed speech, word-finding difficulty, and slowed and illegible handwriting. One year later, RI continued to complain of word-finding difficulty. At that time, he also described difficulties with formulation of ideas, in addition to expression. Although he felt the legibility of his handwriting had improved, he was aware of his micrographia, and he reported having to exert great effort in order to form sufficiently large letters.

Neuropsychological findings from the first and second assessment are presented in Table 1. The z scores were tabulated based on available normative data (as referenced in the subsequent text) from RI’s age-matched peers. In standard clinical neuropsychological practice, test scores obtained by an individual are interpreted using normative data, but the determination of impairment which is significant for the individual is based on a consideration of his or her presumed premorbid level of performance (e.g., Lezak, 1983; Spreen & Strauss, 1998). For example, “average” test scores may represent a significant impairment for an individual who is expected to obtain “superior” scores in relation to his or her peers. Patient RI’s estimated premorbid level of general cog-
Fig. 1. Axial T1-weighted (a), T2-weighted (b), and Coronal T1-weighted (c) MR images demonstrated a well-demarcated lesion in the left putamen (arrows), which also involved the external segment of the globus pallidus, head of the left caudate nucleus (arrowhead in (a)), and the anterior internal capsule. The internal segment of the globus pallidus appears to be spared.

...utive function, based on his demographic information (e.g., age, education, occupation; Barona, Reynolds, & Chastain, 1984) is in the high average range (i.e., IQ score of 117; z score or S.D. of +1.1). His test scores were interpreted in the context of his expected premorbid functioning as measured by this index; that is, he should generally obtain z scores of approximately +1.0. Any score equivalent to −1.0 or lower represents a decline of 2S.D. from the presumed previous level, which would be significant at the P = 0.05 level. Note that, in some cases, this level of performance may not represent impaired performance in the general population.

2.5. Intelligence

RI’s measured level of overall intellectual ability was average, and it improved slightly between the first and second assessment (Wechsler Adult Intelligence Scale-Revised [WAIS-R] Full Scale IQ = 100 and 108, respectively; Wechsler, 1981). This overall level of performance is somewhat lower than the expected high average range (i.e., IQ of 117) based on the previously reported demographic information. At the first assessment, there was no significant difference between intellectual tests tapping verbal abilities such as expressive vocabulary and general fund of knowledge (verbal IQ = 99) versus those tapping visual abilities such as the ability to arrange blocks into specified designs and to arrange pictures in sequence (performance IQ = 104). Both of these scores were average. At the second assessment, verbal abilities remained in the average range (verbal IQ = 102), and visual abilities improved to the high average range (performance IQ = 116), resulting in a notable verbal-visual difference. This pattern of lower-than-expected verbal abilities is consistent with the left-hemisphere localization of the infarct.

2.6. Abstraction

RI showed considerable difficulty with abstract thinking at the initial assessment. Three WAIS-R subtest scores thought to require abstraction (Lezak, 1983) involve the ability to recognize social norms (comprehension), to formulate similarities between words and ideas (similarities), and to detect missing visual details (picture completion). RI’s scores on these tests of abstraction were low average (reported in Table 1) which are not impaired in comparison to the average person but are lower than expected based on RI’s high premorbid general ability. In comparison to these subtests requiring abstraction, his performance was at or above average on the remaining subtests, which are less dependent on abstraction. Consistent with this finding of decreased abstraction, RI had significant difficulties interpreting abstract proverbs such as “A rolling stone gathers no moss” on Gorham’s Proverbs test (Gorham, 1956). That is, he was severely impaired on the standard version requiring him to generate interpretations of the proverbs, and he was moderately impaired on the multiple-choice version requiring him to recognize the correct interpretations (according to normative data provided by Albert, Wolfe, & Lafleche, 1990). RI had similar problems on two tests of abstraction that are predominantly visual. (a) He was mildly impaired on a task requiring him to examine four pictures and identify two different ways that three of the four items are alike (e.g., same shape, orientation, or concept; the Visual–Verbal Test; Feldman & Drasgow, 1981). (b) On a complex-design copying task, his ability to adopt a logical and organized approach was lower than expected (Organization score on the Rey–Osterrieth Complex Figure Test; Stern et al., 1999).

At the second assessment, RI’s abstract thinking improved in some areas, but several significant difficulties remained.
Table 1  
RI’s performance on select neuropsychological tests at 3 and 16 months post-infarction.

<table>
<thead>
<tr>
<th>Test</th>
<th>Months post-infarction</th>
<th>Reference score&lt;sup&gt;a&lt;/sup&gt;</th>
</tr>
</thead>
<tbody>
<tr>
<td>Intelligence</td>
<td></td>
<td></td>
</tr>
<tr>
<td>WAIS-R Full Scale IQ score</td>
<td>100</td>
<td>100 ± 15</td>
</tr>
<tr>
<td>WAIS-R Verbal IQ score</td>
<td>102</td>
<td>100 ± 15</td>
</tr>
<tr>
<td>WAIS-R Performance IQ score</td>
<td>104</td>
<td>100 ± 15</td>
</tr>
<tr>
<td>Abstraction</td>
<td></td>
<td></td>
</tr>
<tr>
<td>WAIS-R comprehension, similarities, picture completion (scaled scores)</td>
<td>6–8&lt;sup&gt;b&lt;/sup&gt;</td>
<td>9–11</td>
</tr>
<tr>
<td>Gorham’s Proverbs standard version (raw score/24)</td>
<td>7&lt;sup&gt;c&lt;/sup&gt;</td>
<td>7&lt;sup&gt;c&lt;/sup&gt;</td>
</tr>
<tr>
<td>Gorham’s Proverbs multiple-choice (raw score/40)</td>
<td>24&lt;sup&gt;d&lt;/sup&gt;</td>
<td>25&lt;sup&gt;d&lt;/sup&gt;</td>
</tr>
<tr>
<td>Visual–Verbal Test error&lt;sup&gt;e&lt;/sup&gt;</td>
<td>6&lt;sup&gt;f&lt;/sup&gt;</td>
<td>6&lt;sup&gt;f&lt;/sup&gt;</td>
</tr>
<tr>
<td>ROCF Organization (score/8)</td>
<td>4&lt;sup&gt;g&lt;/sup&gt;</td>
<td>8</td>
</tr>
<tr>
<td>Matrix Reasoning (scaled score)</td>
<td>na</td>
<td>14</td>
</tr>
<tr>
<td>Conceptualization and flexibility</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Wisconsin Card Sorting Test categories (raw score/8)</td>
<td>6</td>
<td>6</td>
</tr>
<tr>
<td>Wisconsin Card Sorting Test perseverative responses&lt;sup&gt;b&lt;/sup&gt;</td>
<td>10</td>
<td>6</td>
</tr>
<tr>
<td>Stroop interference score&lt;sup&gt;b&lt;/sup&gt;</td>
<td>1.7</td>
<td>1.3</td>
</tr>
<tr>
<td>Trail Making Test Part B (s)&lt;sup&gt;b&lt;/sup&gt;</td>
<td>52</td>
<td>54</td>
</tr>
<tr>
<td>Attention and working memory</td>
<td></td>
<td></td>
</tr>
<tr>
<td>WMS-R Attention/Concentration Index score</td>
<td>125</td>
<td>120</td>
</tr>
<tr>
<td>Consonant Trigrams, 18 s delay (raw score/15)</td>
<td>13</td>
<td>15</td>
</tr>
<tr>
<td>Stroop colour naming (s)&lt;sup&gt;b&lt;/sup&gt;</td>
<td>11</td>
<td>12</td>
</tr>
<tr>
<td>Trail Making Test Part A (s)&lt;sup&gt;b&lt;/sup&gt;</td>
<td>19</td>
<td>22</td>
</tr>
<tr>
<td>Paced Auditory Serial Addition Test, trial 1 (score/80)</td>
<td>39</td>
<td>51</td>
</tr>
<tr>
<td>Memory</td>
<td></td>
<td></td>
</tr>
<tr>
<td>CVLT List A total (raw score/90)</td>
<td>68</td>
<td>64</td>
</tr>
<tr>
<td>CVLT List A Long-Delay Recall (raw score/16)</td>
<td>16</td>
<td>15</td>
</tr>
<tr>
<td>ROCF Immediate Presence/Accuracy (score/20)</td>
<td>16</td>
<td>16</td>
</tr>
<tr>
<td>ROCF Delayed Presence/Accuracy (score/20)</td>
<td>16</td>
<td>16</td>
</tr>
<tr>
<td>Expressive language</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Boston Naming Test (raw score/80)</td>
<td>53&lt;sup&gt;d&lt;/sup&gt;</td>
<td>49&lt;sup&gt;d&lt;/sup&gt;</td>
</tr>
<tr>
<td>Fluency total words generated</td>
<td></td>
<td></td>
</tr>
<tr>
<td>FAS</td>
<td>30&lt;sup&gt;g&lt;/sup&gt;</td>
<td>31&lt;sup&gt;g&lt;/sup&gt;</td>
</tr>
<tr>
<td>Animals</td>
<td>25</td>
<td>27</td>
</tr>
<tr>
<td>Fluency clustering score</td>
<td></td>
<td></td>
</tr>
<tr>
<td>FAS</td>
<td>0.6</td>
<td>0.2&lt;sup&gt;i&lt;/sup&gt;</td>
</tr>
<tr>
<td>Animals</td>
<td>1.8</td>
<td>1.5</td>
</tr>
<tr>
<td>Fluency switching score</td>
<td></td>
<td></td>
</tr>
<tr>
<td>FAS</td>
<td>18&lt;sup&gt;g&lt;/sup&gt;</td>
<td>28&lt;sup&gt;g&lt;/sup&gt;</td>
</tr>
<tr>
<td>Animals</td>
<td>18&lt;sup&gt;g&lt;/sup&gt;</td>
<td>10</td>
</tr>
<tr>
<td>Alternate Uses (raw score correct)</td>
<td>na</td>
<td>17</td>
</tr>
<tr>
<td>Motor function</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Finger Tapping Test, left and right hands (raw scores)</td>
<td>na</td>
<td>48, 40&lt;sup&gt;j&lt;/sup&gt;</td>
</tr>
<tr>
<td>Grooved Pegboard, left and right hands (s)&lt;sup&gt;k&lt;/sup&gt;</td>
<td>na</td>
<td>65, 70&lt;sup&gt;k&lt;/sup&gt;</td>
</tr>
<tr>
<td>Grip strength dynamometer, left and right hands (kg)</td>
<td>na</td>
<td>51, 42&lt;sup&gt;k&lt;/sup&gt;</td>
</tr>
<tr>
<td>Paragraph writing speed, left and right hands (s)&lt;sup&gt;k&lt;/sup&gt;</td>
<td>na</td>
<td>232, 150&lt;sup&gt;k&lt;/sup&gt;</td>
</tr>
</tbody>
</table>

Note: WAIS, Wechsler Adult Intelligence Scale; WMS-R, Wechsler Memory Scale-Revised; ROCF, Rey-Osterrieth Complex Figure; CVLT, California Verbal Learning Test; na, not administered.

<sup>a</sup>Reference scores are presented as mean ± standard deviation.

<sup>b</sup>On these tests, lower scores represent better performance.

<sup>c</sup>RI’s scores were 1 or more S.D. poorer than reference scores (i.e., at least 2S.D. below expected levels).

His performance on the WAIS-R abstraction subs tests improved to average. He performed at or above average on additional abstract thinking tasks that were not administered during the first assessment. (a) He was able to complete patterns of numbers, letters, or words (e.g., “1, 3, 6, 10, ??”; Abstraction Subtest of the Shipley Institute of Living Scale; Zachary, 1986). (b) His ability to consider multiple aspects of stimuli in order to complete visual patterns in two dimensions was good (Matrix Reasoning from the WAIS-III; Wechsler, 1997). His approach to copying a complex figure
was more organized. In contrast, there was no improvement on proverb interpretation (moderately to severely impaired on Gorham’s Proverbs) or finding similarities between visual designs (mildly impaired on the Visual–Verbal Test).

2.7. Conceptualization and flexibility

RI had no difficulty in this area on either assessment. He performed at or above the average range on a test of his ability to deduce simple concepts (i.e., color, shape, and number) based on feedback from the examiner (Wisconsin Card Sorting Test; Heaton, Chelune, Talley, Kay, & Curtiss, 1993). Cognitive flexibility was at or above average on tests of his ability to inhibit a dominant response (i.e., name the color of a word rather than reading the word) on a Stroop of his ability to inhibit a dominant response (i.e., name the color of a word rather than reading the word) on a Stroop Test (Spreen & Strauss, 1998) and to quickly follow an alternating number/letter sequence (i.e., 1, A, 2, B, 3, C, . . . ) on the Trail Making Test Part B (Spreen & Strauss, 1998; normative data provided by Stuss, Stethem, & Pelchat, 1998).

2.8. Attention and working memory

At both assessments, RI’s simple attention ability (i.e., counting and reciting letters of the alphabet, counting by 3’s) was above average (Wechsler Memory Scale-Revised Attention/Concentration Index; Wechsler, 1987). His ability to hold information in memory (i.e., retain three letters) while performing a distracting task (i.e., count backwards by 7’s) on the Consonant Trigrams test (Stuss et al., 1988) was also average at both assessments. Cognitive speed was average on tests of his ability to perform simple tasks quickly, such as naming the colors of dots (Stroop color naming; Spreen & Strauss, 1998) and following a simple numerical sequence on paper (Trail Making Test Part A; Spreen & Strauss, 1998; normative data provided by Stuss et al., 1988).

He had more difficulty sustaining attention and manipulating information at a rapid pace. On the Paced Auditory Serial Addition Test (Gronwall, 1977), RI was asked to add pairs of orally presented digits. At the first assessment, his performance was low-average in comparison to normative data (Stuss et al., 1988), which is below expected levels in comparison to his presumed premorbid level of ability. This improved to average performance at the time of the second assessment.

2.9. Memory

A thorough assessment of multiple types and processes within explicit memory indicated that RI’s levels of performance were consistently at or above average at both assessment times. This included his initial acquisition, learning, retention, immediate and delayed free recall, and immediate and delayed recognition on memory tests involving word lists (California Verbal Learning Test; Delis, Kramer, Kaplan, & Ober, 1984; Recognition Memory Test-Words; Warrington, 1984), paragraphs (Wechsler Memory Scale-Revised Logical Memory; Wechsler, 1987), a geometric figure (Rey–Osterrieth Complex Figure; Stern et al., 1999), and faces (Recognition Memory Test-Faces; Warrington, 1984).

In comparison to his intact explicit memory on these clinical tasks, a previous report (Hay et al., 2002) showed that RI was selectively impaired in implicit memory. That is, on a word-pair fragment-completion task, using equations from Jacoby (1991) process-dissociation procedures, RI’s habit learning (or implicit memory) was decreased relative to controls whereas recollection (or explicit memory) of the word pairs was normal.

2.10. Expressive language

In general, RI’s expressive language was slightly worse than expected based on his background and his high-average general verbal intelligence. At both assessments, confrontation naming was tested by showing drawing of common and uncommon objects (e.g., house, acorn, abacus) and requiring him to produce their names (Boston Naming Test; Kaplan, Goodglass, & Weintraub, 1983). His spontaneous naming ability was low average (t scores of −0.6 to −1.3). Provision of phonemic cues on this test (e.g., “It starts with the sound ‘ab’”) did not significantly improve his naming during the first assessment, but during the second assessment it resulted in successful retrieval for all items not spontaneously named. This pattern during the latter assessment suggests difficulties at the level of retrieving words. Although his overall level of performance is not significantly impaired in comparison to the general population, this is a considerable decline from presumed premorbid ability levels, estimated to be in the high average range (t score of +1.0).

Phonemic fluency on a test of his ability to generate words beginning with the letters F, A, and S as quickly as possible within 60’s (FAS test; Spreen & Strauss, 1998) was borderline impaired at both assessments. Again, this is a considerable decline from presumed previous performance levels which would be expected to be 1.S.D. above the mean. Phonemic fluency performance contrasts with his semantic fluency on a test of his ability to generate animal names as quickly as possible within 60’s (Spreen & Strauss, 1998), which was average at both assessments. To pinpoint the nature of his difficulty, his responses generated on these fluency tasks were examined to determine the cognitive processes contributing to his performance (i.e., clustering and switching; Troyer, 2000). Clustering is defined as the ability to generate words within phonemic subcategories on phonemic fluency (e.g., words starting with the same first two letters or words that rhyme) and within semantic subcategories on semantic fluency (i.e., related subgroups, such as jungle animals and farm animals on animal fluency); this ability is generally unaffected by lesions in the frontal lobe (Troyer, Moscovitch, Winocur, Alexander, & Stuss, 1998).
Switching, on the other hand, is defined as the ability to switch from one subcategory to another, and is impaired in patients with frontal-lobe lesions (Troyer et al., 1998). In comparison to age-corrected normative data (Troyer, 2000), RI’s clustering was generally at or above average, whereas his switching was generally below average. Thus, he demonstrated measurable difficulties with switching, regardless of whether his total fluency output was impaired (i.e., on phonemic fluency) or intact (on semantic fluency).

Because of his self-reported difficulties generating ideas, a test of his ability to generate unusual uses for common objects (Alternate Uses; Guilford, Christensen, Merrifield, & Wilson, 1978) was administered during the second assessment. Six items (e.g., shoe, button, key) were presented individually, and 60’s was allowed to generate unusual uses for each item (e.g., a key could be used to open a can). Overall, RI’s ability to generate unique responses was at the low end of the average range (z score of −0.5), which is not significantly impaired but is lower than expected (z score of +1.0).

2.11. Motor function

We were unable to test RI’s manual motor functions during the first neuropsychological assessment because he broke his right arm in a skiing accident before the assessment was completed. One year later, simple motor speed on a finger-tapping task, manual dexterity on a pegboard task, and hand strength as measured with a grip dynamometer (Heaton, Grant, & Matthews, 1991) were worse in his right hand than in his left hand. These right-sided motor difficulties are consistent with the left-hemisphere localization of the infarct.

A brief assessment of RI’s handwriting and copying was completed during the second assessment. Handwriting speed was examined by asking him to copy short paragraphs (each containing several sentences) as quickly and legibly as possible with both his right and left hands. The right-hand speed advantage demonstrated by RI (i.e., 22%) was smaller than that showed by six right-handed control subjects (mean 54%, range 48–59%) tested in our department. This is consistent with the findings from the simple motor tests. Regarding writing size, there was evidence of micrographia with the right hand, but not with the left hand, when RI was asked to generate and write sentences describing pictures that depicted complex scenarios. There was no evidence of micrographia with either the left or right hand when the demands on expressive language ability were decreased by asking him to copy sentences. Similarly, there was no evidence of micrographia with his right hand on a test of design copying (i.e., Rey–Osterrieth Complex figure).

2.12. Mood

Mood was not depressed at either assessment by self-report. That is, he obtained scores less than 4 on the Beck Depression Inventory (Beck, 1987), a multiple-choice mood questionnaire.

3. Discussion

This case study of RI, a 28-year-old man, demonstrates specific cognitive deficits associated with focal infarction of the left basal ganglia affecting the entire putamen and extending to include small portions of the anterior internal capsule, head of the caudate, and external globus pallidus. Neuropsychological assessment revealed general sparing of many cognitive abilities, including intelligence, simple concept formation, cognitive flexibility, simple attention ability, and explicit memory. In contrast to these spared abilities, specific effects of the infarction were noted on sustained attention and expressive language and abstraction 3 months post-infarction. Approximately 1 year later, sustained attention was improved, but moderate abstraction impairments and mild language involvement remained. Implicit memory was decreased, as previously reported (Hay et al., 2002). Right-hand strength and finger-tapping speed were decreased, consistent with the lateralized left-hemisphere lesion. An interesting pattern of context-dependent micrographia was present, with micrographia evident during expressive writing with his right hand but not during sentence or figure copying with either hand.

Via its frontal–subcortical circuit, the putamen projects primarily to supplementary motor and premotor cortex in the frontal lobes (Alexander et al., 1986). As previously reviewed, neuroimaging studies implicate these areas in non-routine response selection and performance (Monchi et al., 2001). Given that RI’s lesion primarily involved the putamen, we would expect persisting difficulties on tasks with high demands on this type of ability. Consistent with impaired non-practiced response selection, RI had difficulties switching between subcategories (but not generating words within subcategories) on fluency tasks, difficulties recognizing the correct interpretation of abstract proverbs, and a decreased ability to generate novel or unusual uses of objects. Similarly, his micrographia was present during expressive writing (which would require non-practiced responding) but not while copying prose passages or figures. These difficulties are presumed to be related to specific involvement of the skeletomotor circuit including the putamen and frontal motor areas, as implicated in neuroimaging studies.

There is evidence that the basal ganglia are involved in language processes, such as fluency, sentence-level comprehension, and grammar (D’Esposito & Alexander, 1995; Grossman, 1999; Ullman, 2001). An alternate interpretation of RI’s cognitive pattern, therefore, is that it is related to his language deficits. Because we did not perform a comprehensive assessment of RI’s language processes, our ability to comment on the role of language processing on his cognitive pattern is limited. Nevertheless, several of the present
findings do implicate difficulties with language processes, namely his subjective complaints of difficulties expressing himself verbally, and the objective findings of decreased naming and fluency, impaired proverb interpretation, and micrographia when generating sentences. Consistent with previous research, these findings could indicate difficulties with sentence-level comprehension and production. RI’s other findings, however, suggest that some of his language abilities are spared (i.e., intact performance on an expressive vocabulary test requiring him to generate sentences and on a paragraph recall test requiring him to listen to and later recall sentences), and some of his difficulties appear unrelated to language (e.g., decreased attention to visual detail, disorganized approach to copying a complex visual design, and deficit in memory based on habit but not recollection). Thus, although decreased language processing may well play a role in RI’s cognitive pattern and deserves further investigation, we do not think it accounts for the constellation of deficits he showed.

Several of the cognitive difficulties exhibited by RI are similar to those seen in patients with focal frontal lesions, including poor abstraction on Gorham’s Proverbs Test and the Visual–Verbal Test, initial difficulty with sustained attention on the Paced Auditory Serial Addition Test, mild difficulties with confrontation naming on the Boston Naming Test (which improved with phonemic cueing), and a pattern of poorer phonemic fluency than semantic fluency. In RI’s case, these difficulties are presumed to be related to disruption of subcortical/frontal circuits resulting in indirect effects to frontal regions. In contrast to these difficulties reflecting frontal dysfunction, he showed intact conceptualization, no tendency toward perseverative behavior, and good cognitive flexibility on the Stroop Test, Trail Making Test, and Wisconsin Card Sorting Test. Thus, although some aspects of his cognitive profile are consistent with disruption of frontal processes, a number of related abilities were intact, reflecting the focal involvement of frontal regions.

In addition to impairments reflecting frontal dysfunction, RI showed difficulties that are presumably related directly to damage to the basal ganglia. As previously reported, RI’s implicit memory for habit formation was affected, similar to the pattern seen in other patients with basal ganglia damage but not patients with focal frontal damage (Hay et al., 2002). Another direct effect of RI’s damage to the basal ganglia is his micrographia, which can be associated with lesions of the contralateral lentiform nucleus (Muncuçu et al., 2000). Our case study serves to extend previous findings on micrographia to suggest that a unilateral lesion primarily in the putamen and external segment of the globus pallidus disrupts only contralateral production of motor-based programs used for writing. Moreover, our findings suggest an interaction between micrographia and expressive language deficits or task difficulty, as micrographia was evident during spontaneous writing (which requires the generation and expression of ideas, a difficult task for RI) but was absent during paragraph copying (which has no demands on generation or expressive language).

Given that RI’s lesion affected the entire putamen, this case report adds several pieces of information to the current literature on the cognitive effects of damage to the putamen and skeleton motor circuit. Although RI’s lesion extended to other basal ganglia structures, including the caudate nucleus, which may have contributed to some of the cognitive effects, most findings are consistent with damage to the putamen. For example, there has been some previous indication of a link between lentiform lesions and abstract thinking (Warren et al., 2000). RI’s pattern of poor abstraction in both verbal and visual modalities extends this general finding and suggests that the left putamen and its associated frontal circuit may be involved in abstract thinking in general, regardless of modality. This difficulty remained past the acute phase of the infarction, indicating a relatively persistent deficit. RI did not have difficulty on all tests requiring abstract thinking, however, and one might speculate a link between response generation and abstraction thinking. That is, the tests of abstract thinking that RI was able to perform normally (i.e., Matrix Reasoning, Shipley’s Abstraction, and Wisconsin Card Sorting Test) generally had few possible responses from which to choose and a single response that was clearly correct. In contrast, tests that were particularly difficult for him (i.e., Gorham’s Proverbs and the Visual–Verbal Test) were more open-ended, with possible responses not clearly evident, and more than one response that could be considered correct. The latter tests likely make greater demands on response generation and selection. Interestingly, it is apparent that RI was able to compensate for his decreased abstract thinking in his daily life, as evidenced in his subsequent successful completion of a post-graduate degree. Perhaps his significant previous knowledge of his area of study benefited his ability to think abstractly in his daily life.

Another finding not previously reported in the literature is RI’s general difficulty on tasks requiring generation, and this may also be related to decreased non-routine response selection and performance. We found evidence not only of difficulty generating words (e.g., decreased confrontation naming and verbal fluency), but also a decreased ability to generate ideas. Thus, RI’s self-reported difficulty expressing himself is likely related to difficulties not only with finding words to express his ideas, but also difficulties coming up with the ideas themselves. It is possible that his abstraction and generation difficulties are related. Perhaps when faced with concrete materials such as letters or objects, it is difficult for him to generate responses that are more abstract (e.g., exemplars on Fluency tasks or novel uses on Alternate Uses).

In summary, this case study provides further support for the integral role of the basal ganglia in higher cognitive functions as a result of direct involvement of the basal ganglia and indirect involvement of frontal regions. We demonstrated a relatively circumscribed impairment in non-routine response selection and performance that was consistent with a lesion
of the putamen. RI’s lesion, although primarily affecting the putamen, also included small portions of other basal ganglia structures, and it is possible that these also contributed to the pattern of cognitive and motor difficulties described in this report. Further case studies with detailed assessment of frontal/executive abilities, including both traditional clinical and more novel experimental tasks, would be useful in determining the precise correspondence between specific cognitive impairments and individual structures within the basal ganglia.

Acknowledgements

The authors would like to acknowledge F.Q. Gao for assistance with the figures and interpretation of the MRI. Grant support for this work was obtained from Canadian Institutes of Health Research grant number MT1329 to Sandra E. Black.

References


