Pure Topographical Disorientation Following a Right Forceps Major of the Splenium Lesion: A Case Study

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Pure Topographical Disorientation Following a Right Forceps Major of the Splenium Lesion: A Case Study

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A 72-year-old man with pure topographical disorientation following a focal hemorrhage in the right forceps major of splenium was assessed at 2 weeks and 3 months after the onset. Initially, he could identify familiar buildings and landmarks, but noted topographical disorientation, dysfunction in sense of quarters, and in visuo-spatial function. The improvement of topographical disorientation was attained in 3 months, while the inability of the sense of quarters and manipulating visuo-spatial information remained unchanged. These results suggested the heading disorientation was accompanied with impaired sense of quarters, although disabled sense of quarters continued beyond the recovery of heading disorientation.

Keywords: Pure topographical disorientation, forceps major of splenium lesion, heading disorientation, sense of quarters, visuo-spatial function

Introduction

Pure topographical disorientation (PTD), caused by focal brain damage, is characterized by the selective impairment of the ability to find one’s way in a familiar environment. This disorder must not be associated with dementia, acute confusional state, unilateral spatial neglect, visual perceptual disorders, or global amnesia. PTD has simply been divided into two major forms: topographical agnosia or amnesia, however, it is now commonly understood that PTD can be caused by underlying deficits other than agnosia for landmarks and memory impairment for spatial relationships. Indeed, this paper highlights a third mechanism: impaired sense of direction.

The classification of topographical disorientation has recently been revisited. Aguirre and D’Esposito (1999) have proposed a four-part taxonomy: egocentric disorientation, heading disorientation, landmark agnosia, and anterograde disorientation (Aguirre & D’Esposito, 1999) and they selected the patients of Takahashi, Kawamura, Shiota, Kasahata, and Hirayama (1997) as few typical patients with heading disorientation (i.e., loss of sense of direction: the disability to determine the direction from one location to the other.)

The localization of the lesion responsible for heading disorientation is considered the right retrosplenial region. Takahashi and his co-workers’ patients have hemorrhages extending from the right retrosplenial region to the medial parietal lobe (Takahashi et al., 1997). Maguire (2001) has reviewed 14 recent studies of neuroimaging findings of spatial navigation, all of which showed peak activation of the bilateral retrosplenial cortex. Also, the study of rodents has identified a small population of cells related to heading or navigation in the retrosplenial region (Chen, Lin, Green, Barnes, & McNaughton, 1994).

The present report provides three new evidences concerning PTD by heading disorientation: (1) The lesion of our patient is unique and have not reported: the focal forceps major of the splenium region, which is located outside of the retrosplenial area. (2) Existing reports of PTD with retrosplenial lesion stated that the symptoms resolved within the first few months from-onset, however, few studies documented the course of recovery in detail. We examined the patient during 2 weeks and 3 months after the onset of his illness. (3) We examined sense of quarters (the ability to determine directions based on the cardinal coordination in a given location) and visuo-spatial function that have not been mentioned in previous reports.

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Method

Case report

A 72-year-old retired right-handed man (T.H.) with 11 years of education, was born and had always lived in Sapporo, the capital city of Hokkaido Island, where he was an office worker in a transportation company. The office where he worked for over 30 years was located near Sapporo station. On 30 May 2003, while he was driving his car in his neighbourhood, he suddenly lost his understanding of the route to his destination at the intersection near his house. He could recognize his surroundings, but was unable to determine which direction to proceed in. He was admitted to the hospital the same day, and was diagnosed as cerebral haemorrhage. He had a history of hypertension.

His condition was stable when we performed the present experimental investigation on June 9–16 and on August 23–30, 2003. Neurologically he was alert and co-operative. Cranial nerves were all intact including visual acuity and visual field. There were no abnormalities in his motor, sensory, and cerebellar systems.

Brain magnetic resonance imaging (MRI) performed in July (Figure 1A) and September (Figure 1B), 2003 showed a unique right-sided lesion of the focal forceps major of the splenium region, which was located outside of the retrosplenial area.

Neuropsychological assessment (June 2003)

General intelligence was preserved with a score of 30/30 (30/30, August 2003) on the Mini Mental State Examination (MMSE), a verbal intelligence quotient (IQ) of 109, a performance IQ of 97 and a total IQ of 103 on the Wechsler Adult Intelligence Scale – Revised (WAIS–R). Word fluency performance was 15 (17 words, August 2003) words for animals, and 12 (15 August 2003) words for

Fig. 1. A: T2-weighted axial and sagittal images showing a high signal intensity lesion in the focal forceps major of the splenium region, which was located outside of the retrosplenial area (July, 2003). B: T2-weighted axial and sagittal images (September, 2003).
vegetables, in 1 min. The results on the Wechsler Memory Scale – Revised (WMS–R) showed that verbal and visual memory was slightly affected (verbal memory score: 30, index: 81), (visual memory score: 38, index: 80). His delayed recall score was 50, and index was 90 and attention/concentration score was 56 and index was 97. The results of the Visual Perception Test for Agnosia (VPTA) (Japan Society for Higher Brain Dysfunction, 1997), tests of hemispatial neglect, such as copying of a flower, line bisection test, and cancellation test, as well as identification and naming of seven pictures of famous faces, were all normal. He correctly discriminated all seven unknown faces in the VPTA and recognized all three photographs of hospital staff members that he had met after being admitted to hospital. He showed no evidence of aphasia, acalculia, visual agnosia, visuo-spatial agnosia (i.e., Balint’s syndrome), finger agnosia, right–left disorientation, ideomotor apraxia, ideational apraxia, constructional apraxia, or motor impersistence.

**Experimental investigations (June and August 2003)**

**Control subjects**

T.H.’s performance was compared to that of nine normal controls (NC) on several experimental tasks. The control subjects were matched for age (70.8 years; $SD = 2.61$), sex, educational years (12.7 years; $SD = 28.2$), and global cognitive status (MMSE mean score of 29.7/30; $SD = 0.47$). They had all lived in Sapporo for more than 20 years. All subjects gave informed consent to participate in this study.

**Topographical memory of spatial layout**

**Familiar places**

Drawing a layout of his house by the patient and his wife (June 2003) T.H.’s drawing of the layout of his house (Figure 2A) was compared with his wife’s drawing (Figure 2B). On his layout, the perimeter between the house and the flower beds and the garage was not correct. He correctly identified the position of the rooms (kitchen, bathroom, etc.) but made errors concerning the specific location of some pieces of furniture used frequently (television, family Buddhist altar, and the refrigerator near the kitchen).

Drawing a map of the streets around his house (June and August 2003). In the first interview, T.H. said that he could not draw a map of his familiar places in his mind and could not determine the direction at the intersection. He could draw his house and schools near his house correctly, but there were mistakes in the position of the station, and the building around the station. The main errors: the hospital, which is actually in front of the station, appeared behind the station on his map (Figure 3A, June). His drawing of

![Fig. 2. A: Drawing of a layout of his house (T.H.) (June, 2003). B: Drawing of a layout of his house (his wife) (June, 2003).](image-url)
streets, landmarks around the station became more accurate (Figure 3B, August). We believed that he had deficits in topographical memory of spatial layout in June, and that improved in August.

New places

Memory of the hospital to which he was admitted learned after the disease onset (June 2003). In this task, which was conducted in the examination room, 17 days after T.H. had entered and been in the hospital, he was asked to draw a layout of the hospital floor where he had been admitted and to place several items on an outline plan of the floor, he made many critical mistakes: the stairs, toilet and nurse station were all incorrectly located. Moreover, he placed his hospital bedroom on the wrong side of the hospital. Therefore, his impairment of topographical memory of spatial layout in the unfamiliar place was more damaged than that in familiar places.

Sense of direction in new places

Identification of the direction of places in the hospital from examination room (June 2003). We administered this task in the same room and day as the task mentioned above (New places), T.H. was asked to point by finger in the direction of his bedroom, the nurse station, the toilet, and the lobby. He was correct with only one of the four places. Therefore, he had severe dysfunction in sense of direction in new places as well as learned direction.

Ability to learn new heading in the hospital after the disease onset. During his hospitalisation, T.H. failed to find his way from his bedroom to the neuropsychological examination

Fig. 3. A: Drawing of a map of the streets around his house (June, 2003). The arrow indicates correct position of the station. B: Drawing of a map of the streets around his house (August, 2003).
room on five consecutive occasions, which is located on the same floor, approximately 20 m in distance, and requiring three turns from his bedroom.

In August, he came to the hospital correctly by driving his car, but he failed to find his way from the reception desk near the entrance to the neuropsychological examination room, which was located at the second floor, being from 30 m away, going upstairs to the second floor, and requiring three turns from the entrance. However, his wife could find her way in the hospital easily. So his heading disorientation in new places was more damaged than that in familiar environments.

**Geographical memory**

Identification of cities in Japan and in Hokkaido Island on an outline map (June 2003). T.H. correctly placed seven major cities in Japan (NC = 6.44, SD = 0.68) and five in Hokkaido (NC = 4.78, SD = 0.42) on an outline map, because his geographical memory was preserved.

**Learned sense of quarters**

Identification of the quarters from Sapporo to the main cities of Hokkaido Island (June and August 2003). In this task, T.H. had to indicate verbally the quarters (north, north-west, north-east, etc.) from Sapporo to five main cities in Hokkaido that he had correctly placed on an outline map. Two of the five responses (June also August) were accurate whereas NC results were 4.67 (SD = 0.66) out of five. We therefore considered his performance to be impaired and the inability has not changed.

**Indication of the quarters from Sapporo station to several buildings or landmarks (June and August 2003).** This task consisted of indicating verbally the quarters (north, north-west, north-east, etc.) from Sapporo station to several buildings or landmarks. He responded correctly to one (June), two (August) of the five items. Normal controls’ performance of this task was 4.27 (SD = 0.47) out of five. Since, in Sapporo, the streets form a grid, similar to that of chess board, in the central region of the city, and each block is named like as “north 2 west 3”, it can be considered that the patient had a severe impairment on the sense of quarters.

**Identification of the four quarters (north, south, east, and west) (June and August 2003).** In this task, T.H. was asked to indicate the four directions by pointing from the examination room of the hospital, his 69-year-old wife responded correctly for all four directions. In addition, the NC subjects were each tested at home and all responded correctly for all four directions. This part of examinations is unique, since the streets are arranged in four directions in Sapporo, probably being difficult to observe these finding in the street situations like Paris.

**Visuo-spatial function**

**Maze test (June 2003).** T.H. was also given the visual maze task of the Wechsler Intelligence Scale for Children – III (WISC–III). He was asked to trace the path to the exit as quickly as possible. He performed as well as NC subjects in this task (15 and 14.7 points (SD = 1.49), respectively).

**Spatial span task [Visual memory span (WMS–R)] (June and August 2003).** In the visual memory span, T.H. performed well in the test for forward span (9 points), (June, also August) but he showed obvious defect in the test for backward span (5 points) (June, also August) in comparison with NC subjects [forward span, 9.56 points (SD = 1.34); backward span 9.33 points (SD = 1.24)]. The low score in the test for backward span was mainly caused by the mistakes in order; mixed responses of forward and backward span were seen. The inability of manipulation with memorized visual information in the reverse order has not improved.

**Outlining of the letter F (June and August 2003).** In this task, modified from the one designed by Brooks (1968) (Figure 4), T.H. was asked to recall the image of the letter F and describe the outline of the figure without moving his finger or head. This task requires a spoken explanation for each move from exocentric point of view: “go up, turn to the right, down, turn to the left, down, turn to the right, down, turn to the left, down, and turn to the left”. Each correct answer for each line was given one point; the maximum score was therefore 10. He could read and write the letter F, however, his score was only one (June) or two (August) out of 10 for this task. As NC subjects obtained 9.44 points (SD = 0.68) in this task, we concluded that he had a severe impairment to determine the direction on imaged-representation.

**Fig. 4.** Block diagram of the letter F. The arrow indicates the starting point.
Table 1. Results of experimental investigations

<table>
<thead>
<tr>
<th>Tests for geographical memory</th>
<th>Patient June (August)</th>
<th>NC (SD)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Identification of cities on an outline map of Hokkaido and Japan (maximum: Hokkaido, 5; Japan, 7 points)</td>
<td>5</td>
<td>4.78 (0.42)</td>
</tr>
<tr>
<td>From Sapporo to other major cities (maximum 5 points)</td>
<td>2 (2)</td>
<td>4.67 (0.66)</td>
</tr>
<tr>
<td>In Sapporo, from Sapporo station to other buildings (maximum 5 points)</td>
<td>1 (2)</td>
<td>4.27 (0.47)</td>
</tr>
<tr>
<td>Indication of four quarters (maximum 4 points)</td>
<td>0 (2)</td>
<td>4.0</td>
</tr>
</tbody>
</table>

Tests for sense of quarters

<table>
<thead>
<tr>
<th>Tests for visuo-spatial function</th>
<th>Patient June (August)</th>
<th>NC (SD)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Maze test (WISC-III) (points)</td>
<td>15</td>
<td>14.67 (1.49)</td>
</tr>
<tr>
<td>Visual memory span (forward) (WMS-R) (points)</td>
<td>9 (9)</td>
<td>9.56 (1.34)</td>
</tr>
<tr>
<td>Visual memory span (backward) (WMS-R) (points)</td>
<td>5 (5)</td>
<td>9.33 (1.24)</td>
</tr>
<tr>
<td>Outlining of letter F (points) (maximum 10 points)</td>
<td>1 (2)</td>
<td>9.44 (0.68)</td>
</tr>
</tbody>
</table>

WISC III = Wechsler Intelligence Scale for Children – III; NC = normal controls; ( ) indicates the results of tests administered in August 23–30.

Discussion

Experiments 2 weeks after onset

T.H.’s syndrome was characterized by a difficulty in directional orientation in the real world, concerning places that were well-known before the onset, and in newly learned environments after the onset. His pure topographical disorientation (PTD) was caused by the dysfunction of sense of direction and impairment of topographical memory of spatial layout that is more damaged in new places than that in familiar environments, whereas the recognition of familiar landmarks or buildings and geographical memory were preserved. In addition, egocentric and exocentric senses of quarters were severely impaired both in the real world and in map-like representations. He also displayed a deficit in manipulation of visuo-spatial information: visual backward memory span task, and in describing the outline of the letter F, whereas he performed well in visual forward memory span task and in the maze test.

The main characteristics of the topographical disability of T.H. were similar to those of the patients reported by Takahashi et al. (1997), relating in an inability to derive directional information from prominent landmarks, falling into the category of heading disorientation classified by Aguirre and D’Esposito (1999). However, we disclosed additional symptoms of heading disorientation on T.H.: deficits with sense of quarters and with manipulation of visuo-spatial information, such as visual backward memory span and outlining of the letter F. T.H.’s focal lesion was localized to the right forceps major of the splenium region, which was located outside of the retrosplenial area. Other cases of topographical disorientation caused by a lesion located in around the right retrosplenial cortex extending to the medial parietal lobe have been reported (Alsaadi, Binder, Lazar, Doorani, & Mohr, 2000; Bottini, Cappa, Geminiani, & Sterzi, 1990; Cammalleri et al., 1996; Suzuki, Yamadori, Hayakawa, & Fujii, 1998; Takahashi et al., 1997). In all of these cases, patients could recognize landmarks, but had difficulty in navigating in both familiar and new environments.

However, the patient reported by Katayama, Takahashi, Ogawara, and Hattori (1999) who had a lesion in focal retrosplenial cortex (i.e., the isthmus of the right posterior cingulum) (Brodman’s area 29) showed an inability to memorize new routes in contrast to mild impairment in finding the way in the streets around her home. Also, the patients with tumours involving the splenium of the corpus callosum reported by Rudge and Warrington (1991) showed selective impairment of memory and visual perception. There is only one report by Suzuki et al. (1998) that examined exocentric sense of quarters, the patient with subcortical hemorrhage in the right parietal lobe located close to the precuneus showed topographical disorientation with a selective inability to identify the view of a particular building, in contrast to T.H., the patient performed perfectly the task of indicating which direction was a city with respect to another, and could draw an almost complete plan of her house and the buildings surrounding it.

Patterns of cortical activation showed by neuroimaging studies with topographical tasks are consistent with results based on lesion studies (Aguirre, Zarahn, & D’Esposito, 1998; Barrash, 1998; Maguire, 2001; Maguire, Frackowiak, & Frith, 1997; Maguire et al., 1998). The activation of the bilateral retrosplenial cortex, the wide-spread occipitotemporal regions, bilateral parahippocampal gyrus, and right medial parietal regions have been shown in neuroimaging studies of topographical orientation (Aguirre et al., 1998; Barrash, 1998; Maguire et al., 1998). Aguirre et al. (1998) suggested that activation of the superior and inferior parietal lobule could be established by using position (target location) tasks, and that the activation of the parietal, retrosplenial and lingual cortices could be revealed in both recognition and position tasks (Aguirre et al., 1998). Barrash (1998) suggested that retrosplenial cortex is very likely important for integrating
spatial-topographical with information about topographical features. However, neuroimaging study on sense of quarters has not been examined yet.

Taking the findings of this research into consideration, the complexity of topographical orientation, which requires interaction among identification of landmarks, retrieval of relational maps of landmarks from long-term memory, determining one’s current location, planning a route to a new destination, updating one’s current position relative to the goal, and deciding on direction to proceed based on local landmarks, which involves a variety of posterior medial locations of bilateral hemispheres, including the parahippocampus, splenium, retrosplenial cortex and forceps major of the splenium region, may make it vulnerable to disruption at many levels of processing in a widely distributed network of brain regions.

Therefore, the lesion of T.H. was strictly centered to the forceps major of the splenium region, that is involved in the function of the region in the regulation of the sense of direction, sense of quarters in the real world and in map-like representations, and relating to topographical memory of spatial layout in familiar and newly places, and the manipulation of visuo-spatial information at 2 weeks post-onset.

Experiments 3 months after onset

The topographical orientation ability of T.H. improved in the experiment 3 months after the onset. T.H. stated that he had no difficulty walking and to driving a car around well-known places.

It has been suggested that topographical disorientation caused by the right retrosplenial cortex and parietal lobe resolved within the first few months after the onset (Maguire, 2001), and that the persistence of topographical disorientation over long period is unknown (Barrash, 1998). The time till topographical disorientation disappeared in the reports were as follows: Takahashi et al.’s (1997) case 1: 3 weeks; case 2: 1 month; case 3: 2 months; Suzuki et al.’s (1998) case, 2 months; and Alsaadi et al.’s (2000) case 2, 3 months; case 3, 2 months. In T.H., actual navigation had improved after 3 months like reported by others. We administered examination by interview, we found that, in June, the patient could not drive a car in familiar places, however, he could come to the hospital or go to his familiar golf course by car without hesitation in August. This fact was also confirmed by his wife. Also, the description of the route from his house to the hospital was accurate. We believed that the recovery of his topographical disorientation was caused by not only the recovery of topographical memory of spatial layout but also that of sense of directions, that is, in real route-based navigation, the patient was able to imagine the route map and determine the direction with the aid of a view that includes several well-known landmarks. However, the impairment of sense of quarters has not improved. We believed that the sense of quarters requires higher cognitive functions such as the manipulation of visuo-spatial information: some sort of compass mechanism to provide directional infor-

mation (Farrell, 1996), and ability to correspond topographical information with a coordinate axis.

In conclusion, the topographical disorientation caused by the right forceps major of the splenium lesion improved in 3 months, however, a selective inability in the sense of quarters and manipulation of visuoso-spatial information continued beyond 3 months after the onset in this patient. We speculated that the sense of quarters might be distinct mechanism from the sense of direction and the topographical memory of spatial layout.

References