Case Reports

Pure topographical disorientation in a patient with right occipito-temporal lesion

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ABSTRACT

نستعرض في هذا المقال حالة مريض مُصاب بالتوهان الطبغرافي الكامل (pure topographical disorientation) وذلك بعد إصابته بجلطة في قشرة الصدغ القذالي الإنسي الأيمن من الدماغ. ولم يكن باستطاعة هذا المريض الإشارة إلى اللوحات الخارجية الغير مرئية أو رسم خريطة للمدينة التي يسكن فيها، إلا أن بإمكانه معرفة اللوحات، وتحديد المسافة، بالإضافة إلى وصف الطريق بين اللوحات المتعاقبة في نفس المدينة. لقد خضع هذا المريض لعدد من الاختبارات الإدراكية، وكان عليه القيام بستة مهام إدراكية من أجل تقييم حالته وإكمال استفتاء تحديد الاتجاهات الطبغرافية على الطريق، وبذلك أثبتت هذه الدراسة أن موضوع التوهان الطبغرافي يمكن أن ينقسم إلى عدد من العناصر المحددة، وأشارت النتائج أن أحد هذه العناصر قد يرجع إلى انفصال وعدم ارتباط استرجاع الذاكرة لتحديد المواقع على الخريطة (allocentric map) بمعرفة الطريق نفسه (route knowlege).

We describe a patient who presented with a pure topographical disorientation after a stroke involving the right mesial occipito-temporal cortex. He could not point to external unseen landmarks or draw a map of his city, while he could recognize landmarks, and judge the distance, and describe the route between pairs of landmarks of the same city. He underwent standardized cognitive tests, and 6 tasks were used to assess a topographical orientation route-survey. This study provides evidence that topographical disorientation can be subdivided into very specific components. The results suggest that one of these components might refer to the processing of an allocentric map separable from the representation of route knowledge.

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Topographical disorientation (TD) is the inability to find one's way in a large-scale environment. A variety of cerebral lesions or a global cognitive impairment can play an important role in topographical orientation.[1] In an influential review, Aguirre and D'Esposito[2] suggested a categorizing of TD into 4 types of disorders: egocentric disorientation, landmark agnosia, anterograde disorientation, and heading disorientation. Egocentric disorientated patients present impaired performance on a variety of small-space spatial tasks such as spatial span, visual distance evaluation, and pointing to an object no longer within vision. According to Aguirre and D'Esposito,[2] this disorder refers to a deficit in building and holding an egocentric short-term map of locations, and in an inability to navigate new and previously familiar large-scale environments. This deficit would be underpinned by brain areas located within the parietal lobe. Patients with landmark agnosia cannot recognize or memorize relevant landmarks present in an environment. Patients with anterograde disorientation show an inability to find their way specific to novel environments; this deficit is hypothesized to be associated with a lesion of the para hippocampus. Finally, heading disorientation refers to the incapacity to derive directional information in a familiar, and in a new environment. Most patients with heading disorientation show a preserved egocentric representation and landmark recognition, but they report having lost their 'sense of direction' within a space that cannot be perceived at one time. Verbal route description, direction estimation, and map drawing are reported to be severely defective. One of the most interesting studies on heading disorientation is patient one reported by Takahashi et al.[3] He presented an unimpaired delayed recall of spatial location of some objects in the examination room, and a preserved ability to recognize the appearance of familiar buildings. In contrast, when asked to indicate the location of some relevant buildings onto a map of his city or to describe a route between a pair of landmarks his performance was severely impaired. In addition, he was completely unable to recall the direction from one landmark to another. According to Aguirre and D'Esposito,[2] and Maguire,[4] this disorder corresponds to a selective deficit in processing long-term allocentric topographical representation. Most of the reported cases present a lesion located in the right posterior cingulated (retrosplenial) cortex. Aguirre and D'Esposito,[2] and Maguire,[4] suggested that these findings are related to the studies that have found in the same area in rats the so-called head-direction cells, cells that fire only when the animal is oriented in a certain direction within the environment. Two different kinds of spatial representations are classically distinguished in behavioral studies of human spatial long-term memory, namely, route and survey.[2] The route representation consists of the memory trace of the sequence of landmarks encountered, and of the turns associated with each landmark performed along a specific route. Specific tasks traditionally devised to test this type of memory are the verbal description of routes and the route-distance estimation between 2 relevant landmarks. The survey representation is a map-like representation centered on an allocentric frame of reference, allowing direct access to the global spatial layout of an environment. Specific tasks usually used to test this type of memory are the drawing of maps and plans from a bird's-eye view, direction estimation, and the Euclidean-distance estimation between 2 relevant landmarks. The aim of the present study was to investigate the specific pattern of deficit presented by a patient with pure TD and a mesial occipito-temporal infarction. As suggested by Aguirre and D'Esposito,[2] the patient was tested with a series of tasks assessing the different components (egocentric short-term representation, long-term representation, landmark processing) involved in large-scale environment navigation. In addition, we used a set of quantitative topographical tasks tapping different route and survey aspects of long-term spatial representations. We devised a verbal route description task and an alternative route planning task to assess the deficit of route knowledge. Moreover, we conceived an Euclidean direction estimation task, an Euclidean-distance estimation task, and a map-drawing task to assess the deficit of survey knowledge. A selective deficit of route or survey knowledge should impair only a particular category of long-term memory topographical tasks.

Case Report. A 68-year-old right-handed man, with 5 years of formal education, suddenly became unable to find his way while driving his car. He was hospitalized, and admission neurological examination revealed a left upper quadrantanopia. A CT showed an ischemic lesion involving the right mesial occipito-temporal region (**Figure 1**). Duplex ultrasonography, transthoracic echocardiography, and MRI-angiography failed to reveal arterial or cardiac sources of embolism. So, he was discharged with the diagnosis of posterior

Figure 1 - Cranial CT. An ischemic lesion involves the right mesial temporo-occipital area in the territory of the posterior cerebral artery.

cerebral artery infarction. Four months later he arrived at our service for a neuropsychological assessment. His wife reported that he could not find his way around the city center, which was extremely familiar to him. Indeed, he had lived in the same city for 30 years, and had been visiting the city center 2 or 3 times a week for a period of 20 years. After the onset of the lesion, he rarely left his own apartment, because he was afraid of getting lost. He reported being able to recognize buildings and places, but being unable to infer in which direction to go.

An MRI revealed a stabilized ischemic lesion involving the mesial occipito-temporal area. The lesion involved the right parahippocampal gyrus and the right lingual gyrus sparing the hippocampus (**Figure 2**). His performance was compared to 5 age-matched healthy male controls with comparable experience of the geography of the city center. They had been living in the same city for at least 30 years, and had visited the city center at least once a week for a period of at least 10 years for education or work reasons. We administered a series of standard neuropsychological tests assessing

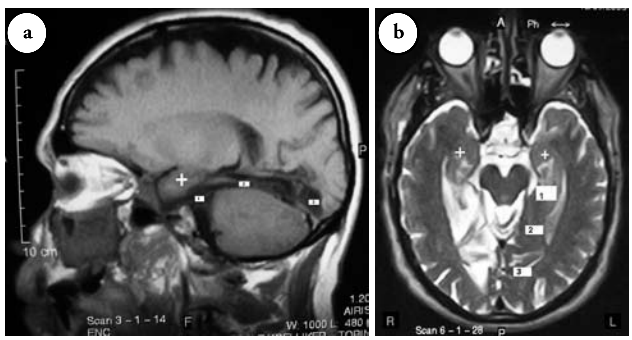

Figure 2 - Brain MRI, T1 sagittal **a**) and T2 axial **b**) scan. The ischemic lesion involves the parahippocampal gyrus [1], the lingual gyrus [2] reaching the cuneus [3]. The retrosplenial cortex as well as the hippocampus [+] are not involved.

Table 1 - Patient neuropsychological test scores.

Test	Score
General	
MMSE	27/30
Raven colour matrices	26/36
Visual perception	
Benton visual form discrimination	28/32
Benton face discrimination	26/27
Line bisection task (mean error)	+ 2.0 mm
Diller letter H cancellation test	0 left omissions 0 right omissions
Verbal memory	
Digit span	4*
Paired associates learning	10
Short story recall	9.3/16
Visual memory	
Corsi's span	4
Corsi's supraspan	impaired*
Geographical knowledge	
Map of Italy	9/15

*Abnormal results. The performance of the patient was assessed with reference to Spinnler and Tognoni[6] for the standardized tests.
MMSE - Mini mental state examination

general cognitive status, visual perception, verbal and visual memory[5] (**Table 1**). All tests offer normative data for the Italian population.

Egocentric orientation. A test of left/right disorientation was administered. He was asked to declare the side of different segments of his body indicated by the experimenter while keeping his eyes closed and/or open. Furthermore, a test of memory of locations, comparable to the one described by Takahashi et al,[3] was performed. In our study, he sat on a chair located on a large sheet of paper spread on the floor reproducing a goniometer. Initially, he was asked to point with his arm to 7 objects (a television, a bookshelf, a door, a desk, a window, a lamp, a picture) indicated one at time by an experimenter and positioned around him. He was asked to memorize their position. Then, he was blindfolded and asked to perform a distractor activity (an interview concerning the problems of his memory). After 5 minutes he was asked to point again to the 7 objects. Performance was measured as the angle between the direction of the arm projected on the goniometer and the correct direction (to the center of the object). As Takahashi et al[3] reported only the number of mistakes without indicating the angular error above which the performance was considered wrong, we adopted in an arbitrary way the following criterion: an absolute angular error superior to 30 degrees was considered as an error. Furthermore, a simplified version of the redrawn Mental Rotation Test (MRT) was administered, and he was requested to find 2 out of 4 figures of a 3D object identical to a model but rotated on the vertical axis. Differently from the original procedure of MRT, only the first 12 items were presented and no time limit was set.

Recognition and identification of landmarks. He was presented with 16 photographs of buildings. The photographs were taken from a position in front of the main entrance of the buildings. Half of these were relevant landmarks located in the city center of the patient; the others were buildings located in other towns of Northern Italy matched with reference to size, architectural style, and functional relevance. He was asked to indicate whether each landmark was located in his city and to name it.

Pointing to landmarks. Only the items correctly identified in the Landmark Recognition task were used (7 out of 8). Ten pairs of landmarks were chosen with the criterion of using all the landmarks at least once, and of avoiding pairs of landmarks visible from each other or the inversion of the same pair (that is, AB and BA). He was asked to imagine being located near the main entrance of a landmark and to point to another landmark. The estimate was made by moving a pointer rotating around the center of a goniometer located on the table in front of him. On this goniometer, 0 degrees corresponded to straight ahead, 180 degrees to backwards, negative values leftwards and positive values rightwards. A correct response varied between -120 degrees and 86 degrees. After a familiarization phase, performance was measured as the error in degrees between the correct direction and the direction to which he pointed. No feedback was given.

Euclidean distance estimation. The same 10 pairs of landmarks used in the previous task were presented. He was asked to verbally estimate the straight-line distance (in meters) between 2 landmarks. Correct responses varied between 440-2520 meters. To familiarize the patient with the task, at the start 2 practice items were presented and he received feedback of the correct distance. No feedback was given for the following items. Pointing to landmarks and Euclidean distance estimations were interleaved in a random order.

Route description and alternative route description. He was asked to describe a possible route between the 10 different pairs of starting-target landmarks used in the previous tasks. He was asked to indicate the street and place names and the directions of turn. If he were unable to give the names of the streets, he was allowed to indicate the presence of a relevant landmark (a monument, a police station…). The descriptions were recorded. Afterwards, he was also asked to find an alternative route for 5 routes, assuming that one of the streets that he had previously named was now blocked off. The 5 controls performed the same tests in the same order and with the same pairs of starting-target landmarks. Although 2 controls had failed to identify a landmark correctly identified by the patient, this procedure was adopted to minimize the difference in testing between patient and controls.

Map drawing. He was asked to draw a map of his apartment on an A4 sheet of paper. No time limit was set. Afterwards, he was presented with photographs of the correctly identified landmarks and asked to draw a map of the city center and to locate the landmarks onto it.

The neuropsychological test results are summarized in **Table 1**. Confrontational testing to assess the presence of visual field defects confirmed a left upper quadrantanopia. Visual perception, verbal memory, and geographical knowledge were relatively preserved. There was no sign of visuomotor ataxia, apraxia, visual neglect, or visual extinction. His spatial span was low but normal. He could not learn the sequence of the spatial supraspan.

Egocentric orientation. As far as egocentric orientation was concerned, mild left/right confusion was not found (3 errors out of 24 responses on the test). His performance for memory of locations of 7 objects (5 correct responses) was within the controls' range (5-7 correct responses). As far as mental rotation was concerned, this test proved to be extremely difficult, also for normal controls. Our patient's score (3 correct responses out of 12) was low but within the controls' range (3-11).

Recognition and identification of landmarks. He correctly classified 7 landmarks of his city and 7 distractors. All the identified landmarks were correctly named. His performance was in the normal range (6-8 for both).

Pointing to landmarks. If we consider the normal range as the mean value for the control group ±2 standard deviations (controls' mean (SD) = 35.7 (10.69), max-min = 21.3-49.1 degrees), his performance (mean absolute pointing error = 80.1 degrees) was clearly impaired (**Figure 3**).

Euclidean distance estimation. The correlation (Pearson's r) between his evaluations and the corresponding actual Euclidean distance was 0.79 ($p<0.006$). If we consider the normal range as the mean value for the control group ± 2 standard deviations (controls' mean correlation (SD) = 0.74 (0.10), max-min = 0.60-0.87), his performance was normal. Interestingly, the correlation (Pearson's r) between his evaluations and the corresponding actual route distance, computed on the basis of the routes described by the patient himself, was even higher and equal to 0.89 ($p<0.0001$) (**Figure 4**).

Route description and alternative route description. Surprisingly, the descriptions were always accurate and detailed. He made only a few mistakes regarding the direction of 4 turns (out of 40). He was rarely unable to recall the street names (3 streets out of 45). In those cases, he described accurately the appearance and function of some landmarks present on the route. In addition, he could find an alternative way without any problem or hesitation.

Map drawing. When asked to draw a map of his apartment, he misplaced a balcony, but the drawing seemed to be globally coherent and adequately preserves the spatial relations between the different rooms. In addition, the analysis of the map confirms that his visuoconstructional ability is largely unimpaired. When asked to draw a map of the city center and to locate the landmarks on it, he placed one landmark, and then he became confused trying to locate the position of a second one and refused to continue with the task (data not shown).

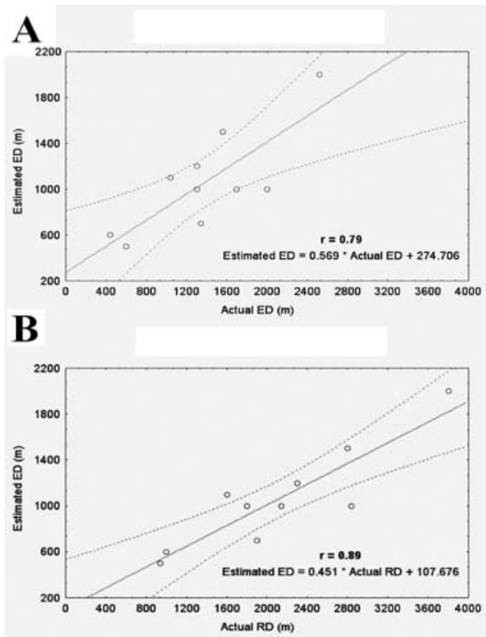

Figure 3 - Distance estimation showing: **A**) Correlation between actual Euclidean Distance (actual ED) and Euclidean Distance estimated by patient. **B**) Correlation between actual Route Distance (actual RD) and Euclidean distance estimated by patient.

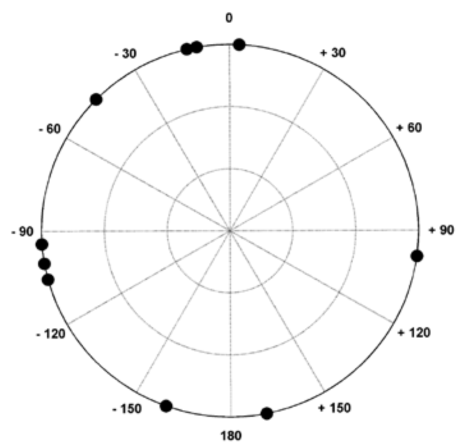

Figure 4 - Circular scatter diagram of the pointing errors produced by the patient. The value 0 corresponds to a performance optimal.

Discussion. Our patient presented a pure TD with normal egocentric orientation, landmark recognition, but severely impaired heading disorientation. However, his disorientation was characterized by a peculiar pattern of deficits: he could not indicate the direction of distant salient landmarks or to locate their position on a map, while he could estimate their distance and describe direct and alternative routes between them.

A comparison between our patient and the other cases of heading disorientation is extremely interesting. Most of the published cases have not been tested using all the tests we administered. Indeed, none tested on a distance estimation task and only 2 used a direction estimation task. Patient one of Takahashi et al[3] failed to draw a map of the neighborhoods of their home, could not describe the route between 2 landmarks nor recall the direction from a landmark to another. Patient 2 and 3 of Takahashi et al,[3] and the patient of Luzzi et al,[6] showed a deficit both on map drawing and on route description, while the patient reported by Katayama et al,[7] revealed a deficit on route description. When tested, all these patients failed on all tests tapping long-term topographical memory. In other words, none of these cases presented dissociation between different aspects of long-term topographical memory. The only exception present in the literature is represented by the case reported by Van der Linden and Seron.[8] Although the lack of a control group makes their findings difficult to evaluate, their patient seems to present a good performance on the route description task and Euclidean distance estimation task, while his performance seems to be severely defective on a direction estimation task and on a map drawing task. The performance of their subject exactly mirrors the performance of our patient.

Our interpretation of the results refers to the theoretical distinction between route and survey knowledge. This dissociation in memory retrieval between different tests of topographical knowledge found in our patient suggests that long-term topographical memory may be divided into different subcomponents. One of these components could encode an allocentric spatial map (survey system). Damage to this module could prevent deriving directional information and severely impair map-drawing. Another system, however, could be sufficient to describe and to plan routes (route system). Furthermore, in our view the latter system would also accomplish our distance estimation task. In other words, in the absence of the survey system, the route system might approximate aerial distance estimation in terms of route distances (or travel time) between 2 landmarks. This hypothesis is supported by the strong correlation between estimated distances and the actual route distances. That could explain the different performances of our patient in drawing the map of his apartment or of the more complex center of his city. It is important to note that our interpretation is completely consistent with that of Van der Linden and Seron.[8] In fact, the main merit of our work is to clarify further the functional features of this kind of topographical deficit with reference to the other kinds of topographical disorders. In addition, the use of a control group leads us to strengthen the evidence for their original hypothesis.

Our hypothesis is consistent with a recent human brain imaging study investigating the retrieval of route and survey knowledge. Before scanning, Hartley et al[9] asked a group of normal subjects to learn a fixed route in one virtual environment and the spatial configuration of another equally complex environment. During fMRI scanning the subjects were required to reproduce the well-learned route during some blocks and to navigate to a target during other blocks. The comparison between the 2 tasks shows major activation of the right hippocampal region for the survey task and of the right head of caudate for the route task. According to these authors, these findings prove that navigation may be sub served by 2 different memory systems. A system localized in the basal ganglia would be involved in the sequential aspect of a route, while another system would be involved in a map-like representation of the environment. The latter would be localized in the mesial temporal lobe and based on the interaction between hippocampal formation and parahippocampal cortex. The authors suggest also that the existence of an alternative (namely, route) system could explain at least partially the good performance of some reported cases with lesions of structures of the hippocampal region.

Moreover, our findings highlight that the right mesial occipito-temporal area plays a key role in the retrieval of allocentric information, and are in agreement with some cases of TD with retrieval deficit of topographical knowledge associated with a lesion confined to the right parahippocampal gyrus. Habib and Sirigu[10] reported 4 patients with TD and lesions involving the right parahippocampal gyrus. Within Aguirre and D'Esposito's[2] framework, these patients are the model cases of anterograde disorientation. Although the main deficit of these patients applied to the learning of new topographical information, 3 out of 4 showed problems in navigating in a familiar environment and, then retrieving previously acquired topographical knowledge. Interestingly, Habib and Sirigu[10] reported briefly that one of these cases (case 4) could describe a route and draw a map despite his deficit on navigation. In addition, the patient reported by Luzzi et al[6] with clear signs of heading disorientation presented right parahippocampal damage. These findings support the hypothesis that the para hippocampus plays an important role in the storing and/or retrieval of allocentric knowledge and not only in its acquisition. Thus, our findings suggest that both the right retrosplenial cortex and parahippocampal region participate in the recall of allocentric knowledge, but they might play different roles in achieving this. It is important to note that the lesion site corresponding to the patient of Van der Linden and Seron[8] differs from our patient's. They reported a right cortico-subcortical fronto-parietal lesion. The lack of more precise brain data does not allow us to better define the extension of the lesion. We do not have a convincing explanation for this difference.

Interestingly, our case shares another characteristic with the case reported by Luzzi et al,[6] the only previous case of heading disorientation with a parahippocampal lesion. While most patients with heading disorientation presented resolution in 3-8 weeks, our subject and that of Luzzi et al[6] presented an enduring TD after a longer period. Our patient was tested 4 months after the onset of the lesion; the topographical deficits of the patient studied by Luzzi et al[6] were still present at a 6-month follow-up. As stressed by Maguire,[4] the deficit following damage of the retrosplenial cortex seems to be short-lived. In contrast, our findings suggest that the deficit of spatial navigation following damage of the parahippocampal gyrus is more lasting. This difference could be extremely interesting in understanding the role played by the retrosplenial cortex and parahippocampal gyrus in the processing of topographical information. Interestingly, the patient of Van der Linden and Seron[8] presented his deficits a year after the onset of the lesion. However, available data are insufficient to speculate further on this issue.

In addition, the analysis of our case raises a stimulating question. If the route knowledge of our patient is relatively unimpaired, it is not clear why the subject cannot use it to navigate. Unfortunately, we did not test the subject in the real situation but only in a simulated one, and we cannot directly compare his performance on the laboratory tests and in real life. However, real navigation is more complicated and difficult than the simple recall of topographical knowledge. The presence of distractors and the need to process information quickly might interfere with the memory retrieval. For instance, our patient reported being dazed by the velocity of the flow of spatial information experienced as a passenger during a journey by car from his home to the neuropsychological service. In addition, the deregulated input from the damaged survey memory system might interfere with the route system resulting in a situation of confusion and affecting the performance.

To conclude, this study provides evidence that TD can be subdivided into specific components. It suggests that one of these components might refer to the processing of an allocentric map separable from the representation of route knowledge. However, a note of caution is necessary as we only report a case of simple dissociation. As suggested by Van der Linden and Seron,[8] we cannot rule out an explanation in terms of a difference in complexity between the route and survey knowledge processing. So, the different levels of difficulty of the tasks might explain our dissociation. Although the data from neuroimaging and experimental psychology seem to be in line with our hypothesis, only an opposite dissociation could really clarify this issue.

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