Remission of anosognosia for right hemiplegia and neglect after caloric vestibular stimulation

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Abstract. Neglect and related phenomena, as anosognosia for hemiplegia and somatoparaphrenia, are often associated to right-hemisphere lesions. These deficits can be alleviated by caloric vestibular stimulation, but little is known about the efficacy of this physiological intervention on neglect following left-hemisphere lesions. Here we report the case of an ambidextrous left brain-damaged patient with severe right personal and extrapersonal neglect, anosognosia for right hemiplegia and somatoparaphrenia. These symptoms co-occurred with more typical manifestations of left-brain damage, such as aphasia and apraxia. Neurological examination revealed right hemiplegia, hemianesthesia and hemianopia. Visuo-spatial tests for personal and extrapersonal neglect, as well as an anosognosia questionnaire, were submitted before and after caloric vestibular stimulation. Results showed a dramatic improvement of anosognosia for hemiplegia and neglect; no change was observed for the remaining deficits. The results confirm the notion of the selectivity of vestibular stimulation for neglect and related disorders and extend this notion by showing that similar effects can be obtained after lesion of the left hemisphere, suggesting that similar mechanisms are responsible for left- and right-sided neglect. Such a peculiar association of language and visuo-spatial disorders jointly present after a left-sided lesion opens the question of the link between handedness and lateralization of cognitive functions.

Keywords: Anosognosia for right hemiplegia, right neglect, left-sided lesion, caloric vestibular stimulation

1. Introduction

The term anosognosia (Babinski, 1914) refers to the lack of awareness of neurological or cognitive deficits following brain injury. In case of neurological diseases, anosognosia for hemiplegia is the most extensively examined phenomenon. The term anosognosia is also used to indicate the impaired awareness of cognitive disorders, such as memory disease (Akai et al., 2009). The prevalence of anosognosia for motor deficit is asymmetric, the majority of patients exhibiting this symptom following a lesion of the right hemisphere (see Orfei et al., 2007 for a review). In right-brain damaged patients, anosognosia for left hemiplegia is frequently (but not always) associated with left unilateral spatial neglect (USN), a disorder characterized
of defective attention, perception and representation of the contralesional side of the space (Vallar and Ronchi, 2006). The relationship between the two disorders is reinforced by the finding that caloric vestibular stimulation, a physiological technique that may temporarily improve the pathological manifestations of USN, can alleviate both deficits (Rode et al., 1992).

Few data are reported in the literature about the recovery of anosognosia and right neglect following a left-brain lesion. Even if contralesional spatial neglect is more frequent after a right cerebral damage, evidence of patients affected by right USN after left-hemisphere lesions are on record (see an example in Bultitude and Rafal, 2010). Vallar et al. (1995) have reported the case of a right-handed patient with left fronto-parietal damage who exhibited both a severe fluent aphasia and right-sided spatial neglect. Caloric vestibular stimulation (CVS) temporarily improved right visuo-spatial neglect, without any detectable effects on the patient’s dyphasic deficit, suggesting the selectivity of CVS. In such cases, anosognosia for hemiplegia, which is typically assessed by a verbal questionnaire, is not evaluated because of the importance and severity of the linguistic dysfunction. In a subsequent study, Schiff and Pulver (1999) reported that caloric vestibular stimulation can provoke the transient remission of right USN and the associated disownership of the right hand in a patient with a left hemispheric damage, but no information about anosognosia were included in that report. Although less frequently documented in the literature, anosognosia for right motor deficits has been reported in patients with left-brain damage (Orfei et al., 2007). The frequency of this disorder may actually be underestimated, as the verbal evaluation, usually employed to estimate the presence of awareness, could not allow for a correct assessment in patients with important linguistic deficits (Cocchini et al., 2009).

In this study, we document the case of an ambidextrous patient who exhibited both right neglect and anosognosia for right hemiplegia, following a unilateral left-sided lesion due to a stroke. Caloric vestibular stimulation was performed to assess the potential improvement of the deficits and the selectivity of the intervention in this peculiar syndromic association.

2. Case report

G.B. was a 73 year-old man who sustained a left hemisphere ischaemic cerebrovascular disease on October 25th, 2007. He was first admitted to the Emergency Department at the Hospital of Lyon and, 25 days after the onset, was moved to a Neurorehabilitation Department for the persistence of right hemiplegia. A MRI scan performed four months after the stroke (see Fig. 1), showed a large cortical and sub-cortical hypodense lesion in the left hemisphere including the temporo-parietal junction, without damage of the right hemisphere.

The patient was ambidextrous, as assessed by the Edinburgh Handedness Inventory (Oldfield, 1971). G.B. was submitted to a neuropsychological evaluation on December 6th 2007, one month and half after the stroke. At this time, the neurological exam showed the persistence of a complete right hemiplegia affecting both the upper and lower limbs, associated to a right severe hemianesthesia affecting all modalities and to an apparently severe right homonymous hemianopia. The patient was quite alert and well oriented in time and space. At first, linguistic and praxic functions were assessed. The patient presented a severe apraxia in both ideational and ideomotor domains when asked to perform gestures with his left hand (De Renzi et al., 1980; De Renzi and Lucchelli, 1988). A standard language examination was performed (BDAE: Mazaux and Orgogozo, 1982). The Oral expression assessment revealed a spontaneous fluent speech without phonological, morphological and syntactic deficits, but with anomalies. Word repetition was globally preserved (8/10, cut-off: 8). The visual confrontation task was severely compromised (23/30, cut-off: 75) and characterized by frequent anomias and rare perseverations. Responsive naming revealed a poor performance (6/30, cut-off: 22). In the body-part naming test the patient obtained a compromised performance (14/30, cut-off: 20). The Auditory comprehension was preserved in functional situations and simple conversation, more difficult in case of more complex context or long sentences. In the word discrimination test the patient scored 34/72 (cut-off: 56), with a major impairment for colors and numbers. The body-part identification task was in the normal range (15/20, cut-off: 15). Complex ideational material was severely impaired (4/12, cut-off: 6). The Written expression was constrained by associated deficits, in particular by praxic and visual ones. The production of the patient’s forename has been correctly obtained with moving letters. Words’ spelling to dictation was impossible to assess. The Reading comprehension showed a partial impairment, with few possibilities in word recognition (2/8,
For the stroke, the patient presented with eye and head oriented towards the left, revealing a defective exploration of the contralateral right hemispace. By consequence, the presence of extrapersonal and personal right neglect was evaluated. Two target cancellation tasks were administered: because the patient was unable to cross out the targets using a pencil (due to his severe associated apraxia), the task was modified asking the patient to point at targets. The performance was pathological both in the line (modified version of Albert, 1973, 5/40 lines found) and in the circle (9/17 circular targets found) cancellation tasks. The line bisection task (6 lines, 20 cm each in length) performance showed a large leftward deviation (−36.65% on average). Right personal neglect was also assessed. During the body exploration task (modified version of the Fluff test: Cocchini et al., 2001), the patient had to catch some targets disposed on the right half of the body: G.B. correctly found only 11 out of 21 targets with visual control (eyes opened), and 8 out of 21 targets without visual control (eyes closed). In the hand reaching test (Bisiach et al., 1986), G.B. started the movement towards the right, but he reached his right shoulder (score: 2/3). The battery of tests administered indicated the presence of severe right personal and extrapersonal spatial neglect. During the neuropsychological evaluation the examiner recorded also the presence of left-right disorientation.

Finally, anosognosia for right hemiplegia was assessed with a short interview with increasingly specific questions (Bisiach et al., 1986). This test indicated that G.B. was unable to admit his motor deficits, even after a neurological demonstration (score for both upper and lower limbs: 3/3). During the interview, some somatoparaphrenic delusional elements (Vallar and Ronchi, 2009) were recorded: the patient claimed he had only the left hand and, when his right paralyzed hand was put close to the left one, he said that it was “apparently” his hand.

3. Materials and methods

The caloric vestibular stimulation (CVS) was performed by irrigating the external right ear canal with 60 cc of cold (20°C) water for 30 seconds. During stimulation the room illumination was decreased; the patient laid down on his bed with his head tilted of approximately 30° forward. The vestibular activation produced a clear horizontal nystagmus, with a rightward slow phase.

The following neurological and neuropsychological indexes, evaluated before the CVS (see the case report description), were also assessed immediately after the stimulation: 1) motor and somatosensory functions; 2) anosognosia for right hemiplegia; 3) right personal neglect, assessed by the body exploration (eyes opened) and hand reaching tests; 4) right extrapersonal neglect, evaluated using the circle cancellation and line bisection tasks; 5) spontaneous speech. The whole experimental session lasted about 20 minutes.

4. Results

The performance of G.B. before and after CVS is reported in detail in Table 1. Results of the neurological exam showed no change, with the persistence of
complete right hemiplegia and hemianesthesia. On the contrary, the awareness of motor deficits was significantly modified: after the vestibular simulation the patient recognized the presence of hemiplegia. The delusional ideas about the feeling to have only one (the left) hand were no longer present.

Regarding the spatial attention deficit, after the vestibular stimulation the clinical observation of G.B. revealed that the head and gaze deviation towards the left was markedly reduced. The performance in the circle cancellation task indicated an amelioration of the right extrapersonal neglect (pre-post CVS difference: 4 targets). In the line bisection test, the leftward deviation was importantly reduced through vestibular stimulation (pre-post CVS difference: +13.84%). With respect to the personal neglect, the performance of G.B. in the Fluff test indicated a better exploration of his right hemibody (pre-post CVS difference: 6 targets). The reaching task performed after the CVS indicated the complete accuracy of the performance, with the correct reaching of the right hand.

Lastly, regarding the linguistic functions, spontaneous speech revealed that the deficit was not modified, with persistence of fluent aphasia. Two days after the caloric vestibular stimulation, the clinical examination of G.B. revealed the reappearance of an important head and gaze deviation towards the left hemi-space, with severe right hemi-spatial neglect for extrapersonal and personal right spaces. Remarkably, the patient was able to recognize the presence of right hemiplegia, but he minimized the severity of his illness, consistent with an anosodiaphoria.

5. Discussion

To our knowledge, the results of this report represent the first demonstration that caloric vestibular stimulation can improve not only right neglect but also the associated disorder of awareness for right motor deficits due to a left-sided hemispheric lesion. A dramatic regression of neglect, in both personal (reaching and body exploration tests) and extra-personal domains (line bisection and cancellation tasks), and a recovery of anosognosia for hemiplegia (assessed by a questionnaire) were found consecutive to CVS. In sharp contrast, no changes were observed for the remaining neurological and neuropsychological deficits. The temporary amelioration of spatial neglect symptoms is compatible with the corpus of knowledge about the short lasting effects of vestibular stimulation (see a review in Rossetti and Rode, 2002). Interestingly, although not quantitatively assessed during the follow up session (48 hours after CVS), the positive effect of CVS on anosognosia for right hemiplegia did not completely vanish at the following examination: two days...
after the stimulation, the patient clinically presented with only a slight degree of unawareness, compatible with the presence of anosognosia (Babinski, 1923).

The modulation of neglect and related disorders after CVS reveals that effects similar to those obtained in case of classical right-brain lesions can be obtained after damage of the left hemisphere, suggesting that left and right neglect may share not only similar neural substrates (Vallar, 1993), but also analogous functional mechanisms. In support to this view, recent studies have shown that right spatial neglect symptoms, similar to the left neglect ones, can be reduced by prism adaptation (Buititude and Raffard, 2010).

Furthermore, here we have confirmed the selectivity of CVS on the spatial symptoms. G.B.’s performance was unchanged with respect to linguistic functions, as previously reported by Vallar et al. (1995); in addition, the neurological deficits due to the left hemisphere damage were similarly left unchanged. Unlike previous studies reporting regression of anosognosia and motor deficit after left CVS in right-brain damaged patients (Rode et al., 1992), no similar improvement of right hemibody force was observed in the present case. This difference may be due to the size of the left hemisphere damage and the shorter post-onset delay. The specificity of the CVS effect indicates that the amelioration of right spatial neglect and, for the first time, of anosognosia for right hemiplegia, is not triggered by a general hemispheric activation or increased arousal. This finding is corroborated also by the null effect previously found on USN symptoms after a bilateral vestibular stimulation (Rode et al., 2002), which produces stronger cerebral activation than the unilateral one. Interestingly, recent anatomo-clinical data (Vocat et al., 2010) suggest that, at a sub-acute phase, the severity of anosognosia for hemiplegia is correlated to visuo-spatial neglect. However, double dissociations between spatial neglect and anosognosia for hemiplegia have been documented (Jeitkomen et al., 2000).

Finally, the present report provides important information about the hemispheric cognitive specialization of an ambidextrous patient. For right-handed subjects, the principal role of the left and right hemispheres respectively in linguistic and spatial functions is well known. However, previous studies have reported exceptions to this classical hemispheric dominance, such as right-handed patients with crossed aphasia (Faglia and Vignolo, 1990) or crossed right hemisphere syndrome (Marchetti et al., 2005). Also USN in right-handed patients after left-brain lesions is on record (Kleinman et al., 2007). Regarding the relationship between handedness and lateralization of cognitive functions, while the cerebral organization of language in left-handed subjects has been extensively studied, a minor corpus of data is available about the hemispheric dominance of visuo-spatial functions in non-right-handed people. No difference between right- and non-right-handers is found in visuo-spatial judgment (Masure and Benton, 1983) and spatial reasoning (Sheehan and Smith 1986) tests. On the contrary, a different cerebral specialization in right- and left-handed people is recorded during a mental rotation task (Shimoda et al., 2008). With respect to spatial attention, in the study of Maeshima et al. (1992) right- and non-right-handed patients with left hemisphere lesions and USN were evaluated: the majority of the nine non-right-handed (one left-handed and eight ambidextrous) patients also presented with different degrees of aphasia, suggesting that in these cases the left hemisphere has the principal role for both linguistic and visuo-spatial functions. In that study, the USN of right-handed subjects was present, but temporary. Overall these findings indicate that sometimes the lateralization of visuo-spatial functions in the right hemisphere may be incomplete, especially for non-right-handed people.

In the non-right-handed patient reported here, the left hemisphere appears to be the site of multiple cognitive functions: after a single extensive stroke, severe praxic and visuo-spatial deficits were observed: the aphasic symptoms were mild, revealing that linguistic functions could be partially lateralized in the left hemisphere with a possible involvement of the right, intact, hemisphere in verbal production and comprehension. To conclude, the case of G.B. here reported contributes to extend our knowledge about the lateralization of cognitive functions in ambidextrous subjects. Moreover, the effect of caloric vestibular stimulation on the spatial deficits confirms the notion of specificity of this intervention and provides the first demonstration of its effectiveness on the remission of the unawareness of motor disease after left-sided lesions.

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References


