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Belief and awareness: reflections on a case of persistent anosognosia

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Abstract

Persisting anosognosia after acute lesions is relatively rare, and no case studies to date have reported functional scanning investigation of this disorder. This is a case report of an 85-year-old right-handed Scottish woman, EN, who showed persistent anosognosia for hemiplegia following a haemorrhagic stroke. Extensive damage in the right hemisphere caused left upper and lower limb flaccid hemiplegia and severe left-sided neglect. Lack of awareness for her deficits was still present 2 years after the stroke, when neurological, neuropsychological, and SPECT examinations were performed. Testing revealed severe left unilateral neglect and poor performance on verbal fluency tasks. EN had age normal memory performance, and her object recognition and praxic abilities were preserved. She showed no global reasoning or language problems apart from her abnormal beliefs. EN believed that she was able to walk and carry out several activities, in a context of other disorders of belief. SPECT scan showed marked hypoperfusion in the right parietotemporal cortex and this extended to the associative cortex in the right frontal regions.

The persistence of anosognosia in this patient cannot be explained by memory impairments or global cognitive decline. A possible account might be that alteration in awareness was maintained by contingent right frontal and/or parietal dysfunction causing a suspension or change in the ability to monitor and check the 'real' and especially to assess the veracity of mental contents.

Keywords: Anosognosia; Hemiplegia; Neglect; SPECT; Persistent; Chronic; Reality monitoring; Delusions

1. Introduction

A failure of awareness of deficit or anosognosia is frequently reported following right hemisphere damage and can also be seen in patients who have global cognitive impairment from degenerative causes like Alzheimer's disease (AD). Lack of awareness of functional decline in AD, when this symptom makes its appearance, is persistent and its severity increases with disease progression. When anosognosia appears following an acute lesion it is usually shortlived, in most cases involves unawareness of hemiplegia, and when the acute or post-acute state is over patients usually gain awareness of their condition. Persisting anosognosia after acute lesions is less frequent, although a few cases have been described (e.g. Berti, Ladavas, & Della Corte, 1996; Berti, Ladavas, Stracciari, Giannarelli, & Ossola, 1998; Cocchini, Beschin, & Della Sala, 2002; Marcel, Tegnér, & Nimmo-Smith, in press). In the context of global cognitive impairments anosognosia can be seen as an inevitable consequence of multiple and increasingly severe cognitive failures, but the need for a theoretical explanation seems both more necessary and potentially achievable where this symptom appears in the absence of global deficits. Several such theoretical accounts have been put forward. These can be fairly divided into psychodynamic or motivational theories (Weinstein & Kahn, 1955) and cognitive theories (Bisiach, Meregalli, & Berti, 1990; McGlynn & Schacter, 1989). Pure psychodynamic theories, involving for example denial or depression, are challenged to some degree as comprehensive explanations of disturbances in awareness by the more frequent link between anosognosia and right hemisphere dysfunction. It is also perhaps hard to understand how mechanisms of denial might be specific to individual aspects of awareness when these disturbances are dissociated in individual patients (Berti et al., 1996). Motivational explanations are also less convincing given the usual parallel resolution of the symptom and the acute phase of brain damage, although it is conceivable that certain forms of brain damage might allow the appearance of otherwise suppressed defence mechanisms.

The existing cognitive theories are numerous and must attempt to account for the various syndromes of

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anosognosia, and the different observed patterns of neurological and cognitive disturbance. No theory appears to account for the phenomenon in all its forms, its association with the right hemisphere, and the appearance of anosognosia as both a transient and occasionally as a persistent symptom.

This paper reports a case of persisting anosognosia for hemiplegia and left-sided neglect in the absence of global mental deterioration. It is suggested that the associated phenomena of false beliefs and misidentifications may be integral components of persistent anosognosia in this case. Similar mechanisms may be of importance, albeit transiently, in association with more acute disturbances of awareness. It is important for our argument, and not available in other published material, that functional as well as structural imaging is available for analysis in this case.

2. Case report

EN[1] is an 85-year-old right-handed Scottish lady with a history of hypertension, atrial fibrillation and asthma. She was taking aspirin. In February and March 1999 she had two transient ischemic attacks. In the first, she reported mild and shortlived speech disturbance, while in the second, she had left upper limb paraesthesia which resolved spontaneously after 1 h. During the second episode, her case notes record that she felt that her arm was weakened and difficult to move. By the time she was neurologically examined, however, there were no focal signs. Warfarin was prescribed after the first episode. In June 2000 she was admitted to general medical care following a haemorrhagic stroke, with extensive damage in the right hemisphere. The MRI images, 3 days after the stroke, showed a haemorrhage measuring 2 cm × 3 cm × 3 cm involving the right posterior temporal and parietal regions. The lesion was surrounded by a rim of cerebral oedema, and there was some effacement of the posterior horn of the right lateral ventricle. Routine neurological examination indicated a left-sided flaccid paralysis of the upper and lower limb. There was a mild paresis of the left lower facial muscles, which quickly resolved. She also had some difficulty swallowing with impaired tongue movements. She was mildly dysarthric with severe left visuospatial neglect and profound anosognosia for her hemiplegia and unilateral neglect. No visual field defect was elicited. No specialist neurological or neuropsychological assessments were carried out at this time.

The patient had physical rehabilitation but remained unable to walk and was mobilised only with a wheelchair. She was discharged after 4 weeks to permanent nursing home care.

[1] Patient's initials have been changed to preserve anonymity and both the patient and her relatives gave their consent to the present report.

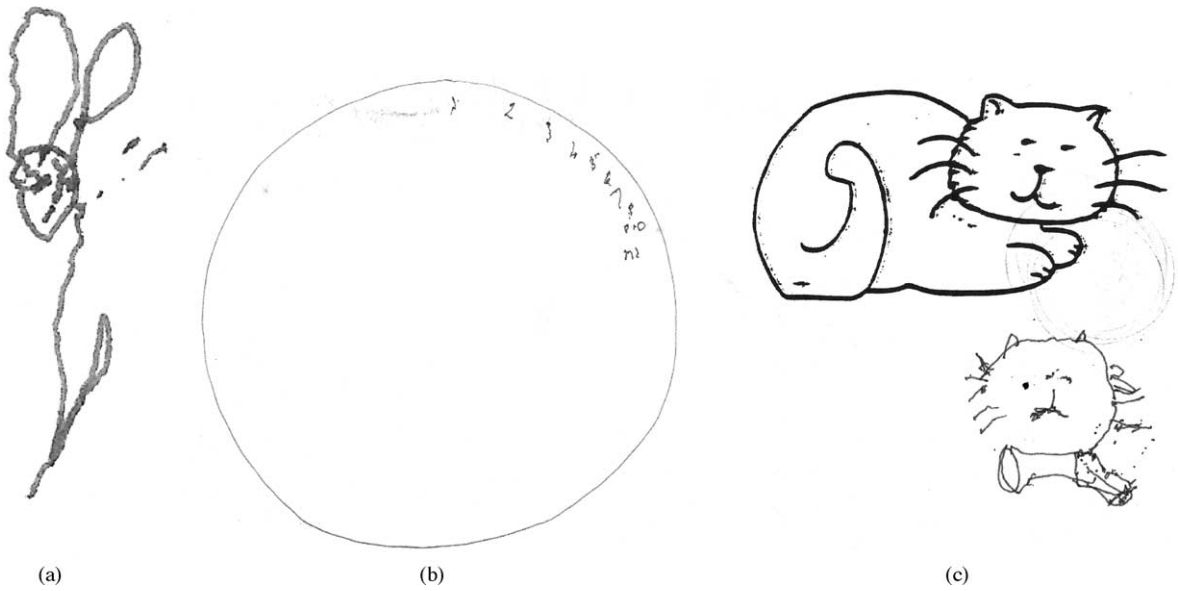
2.1. Neuropsychiatric assessment

In November 2001, 17 months after the stroke, she was referred for psychiatric assessment because of behavioural problems. The nursing home staff reported her spontaneous claims following admission that she was able to stand and walk, and to look after herself. She accused staff of being cruel, torturing and poisoning her and leaving her out in the rain. At times she had requested that her mother be informed of her whereabouts. She had discussed sexual matters with her son in an inappropriate and disinhibited way.

EN presented as an alert and emotionally serene lady who smiled readily and showed no language disorder. There was no evidence of subacute confusion or alteration in the level of consciousness at any point during the examinations. She claimed that she lived in a new house in the city, although she was presently visiting another city where her sister lived. She had frequent visitors at home and was perfectly able to travel by train between her present residence and that of her sister. She continued to hold the belief that her left side was fully functional and that she was physically independent. At different times the staff reported she had said that either her left hand or both of her hands had disappeared. Neuropsychiatric inventory was completed and the retrospective assessment showed that she had frequent delusions, persistent apathy interspersed with frequent episodes of aggressive behaviour, all of moderate severity. There was evidence of left-sided neglect and inattention. She was referred for functional and structural brain scan and neuropsychological assessment, because of concerns about more global cognitive decline due to progressive degenerative brain disease together with a requirement to counsel and fully inform her family and nursing staff about her symptoms. Risperidone 0.5 mg daily was prescribed with benefit for the hostility and aggressive behaviours associated with paranoid beliefs.

2.2. Neuropsychological assessment

EN was tested in December 2001 and August 2002. On both occasions comprehensive testing as detailed below was carried out, including assessment of the persistent anosognosia. There were only minor differences in psychometric test scores and no changes in her behaviour or mental state between examinations. Mini Mental State Examination (Folstein, Folstein, & McHugh, 1975) showed that EN was partially oriented in time and place. She knew the month and the year, and the city and country where she lived. She could readily understand and follow test instructions and was very cooperative. She had no oral or written comprehension deficits nor grammatical comprehension deficits nor expressive language problems. Her spontaneous speech was well organised with good intonational contour, grammatical and logical structure and rich in information content. On confrontation naming she showed no naming difficulties, although on two occasions she produced visual paraphasias (e.g. thermometer → pen, compasses → stepladder). There



(a) (b) (c)

Fig. 1. EN's spontaneous drawing of (a) a daisy; (b) a clock face and (c) copying a line drawing.

were no object recognition deficits, but her performance was affected by poor eyesight.

Her short term memory was intact and she achieved repetition of seven digits forward and four digits backward. Long term verbal memory was tested with a paired associate word task and she achieved a total score of 11/12, a performance within the range of that achieved by a group of normal Scottish individuals comparable to EN for age and education. Her verbal long term memory was also tested using a prose memory task on which she scored 15/25, a score within the normal range for her age. She was also given a structured questionnaire which assessed susceptibility to provoked confabulations (Cooper, Shanks, Venneri, unpublished). Her performance on this task was within local norms for the memory components, but was within the pathological range for verbal invention. She was able to report personal events, the occupation of family members and their whereabouts. She could identify and name them as well as indicate their relation to her when shown their photographs. She achieved scores well below those of age and education matched controls both on semantic (17 items) and phonemic (8 items) fluency tasks. Reasoning abilities were tested with the similarities subtest of the WAIS III (Wechsler, 1997). She scored 19/28 on this task, a score within the range of age and education matched controls. Her ability to make plausibility judgments was informally evaluated during assessment. EN showed no difficulties in this respect.

2.3. Assessment of neglect

She showed severe left-sided neglect while copying simple geometrical figures with omission of the left side of the figures. Spontaneous drawing was also hampered by severe spatial neglect. She drew only the right part of a daisy and crammed all numbers in the top right quadrant of a clock face (see Fig. 1a and b). She was able to name the objects represented in simple line drawings, but her copying of these drawings was also affected by the presence of neglect (Fig. 1c).

Both in spontaneous drawing and when requested to write a sentence or her name, EN used only the far right side of the sheet which had been positioned in front of her aligned to her trunk midline.

The severity of her left-sided spatial neglect was tested formally using the Albert test (Albert, 1973), the Bells test (Gauthier, Dehaut, & Joanette, 1989), a letter cancellation task and a line bisection task. On the Albert test she made 10 omissions in total, eight of which were in the left half and only two were in the right half. She identified only six target items, all on the far right side, on the Bells test. She also marked two distractors placed in the same spatial region; she was, however, aware of making these errors. Similar findings were obtained on the letter cancellation task where she omitted 38 out of 52 target letters in the right half and 52 out of 52 target letters on the left half. Most of the identified target letters were placed in the bottom part of the right half of the page. Severe neglect was also detected on the line bisection task. She placed the mid point of different lines on the right of the true midpoint and her error was greater with longer lines. Her reading abilities were tested with single words from 4 to 10 letters long. She read all words correctly but one (photograph → monograph).

Left-sided spatial neglect was observable on both testing occasions. She also showed a marked bias to the right in both her trunk posture and face orientation. There were signs of personal neglect for the left upper limb. A cushion had been positioned by the nurses to support her left arm, which had a severe flaccid paralysis. However, her left arm would

often slip from its original position and she would leave it dangling, with no attempt to make it resume its original position, no matter what activity she was engaged in and no matter how awkward her position was.

2.4. Assessment of sensation and extinction

Visual, auditory and tactile extinction was tested. Extinction in the visual modality was severe, and only mild in the auditory modality. During the assessment of tactile extinction it became apparent that EN had severe sensory deficits in both her left upper and lower limbs. Sensory assessment by routine neurological examination showed that she was insensitive for touch over the upper and lower left limbs although she was able to feel pinprick, and she spontaneously complained of discomfort in her left foot.

2.5. Assessment of anosognosia

EN showed severe anosognosia for her hemiplegia on both occasions, confirming the observation and reports by staff in the nursing home. Hemiplegia was assessed by routine neurological examination of the limbs. When asked to lift her hands, she only lifted her right hand. When asked if she could move her left arm she replied 'Yes, I can move my arm, but it is better if it is rested. The doctor told me to rest it'. Similar behaviour was observed when asked whether she could walk. She claimed she could walk. When challenged by the examiner to go to the door to greet her visitors, she replied 'Yes, I could get up to meet them, but the doctor says it would be better if I rested'. A formal questionnaire (adapted from Feinberg, Roane, & Ali, 2000) was then used to explore her awareness of her hemiplegia for the left upper and lower limbs separately. This instrument allows a qualitative assessment of anosognosic symptomatology, rather than a quantitative estimate. When asked whether she had any weakness in her arms she acknowledged that her left arm was weak but denied that it was causing any problems, except when she had to be put in a hoist. Asked why she had to be put in hoist, she replied that this was to avoid tiring the staff when they were helping her. When challenged, she acknowledged that she could not use her left arm as well as before and that she was fearful about losing her ability to use her arm properly. She claimed that sensation in her left arm was normal. She acknowledged that her left arm had been paralysed and when her left arm was lifted she agreed that the limb appeared to be rather weak.

A similar interview was completed in reference to her left lower limb. If asked about weakness in her legs she claimed that there was none. EN maintained that her left leg was not causing her any trouble. She claimed that sensation in her left leg was normal and that the leg felt normal. She acknowledged that she could not use it as well as before and she was fearful that something might prevent her from using it properly in the future. When asked whether her doctor's account of her leg being paralysed was correct, she denied paralysis and asserted that only the functions of her left wrist had been impaired. When her left leg was lifted and she was invited to look at the limb and to see that she had no control over it, she acknowledged this fact but could not explain why she had lost control.

Her ability to use both unimanual as well as bimanual objects was also investigated. She was requested to lift and open a book. She took it with her right hand by its spine, holding the book as if her left hand might contribute to the action, but then failed to complete the task. She said that she was unable to act as requested. Asked why, she could not say. Similar behaviour was observed for all actions requiring bimanual involvement (harmonica, stethoscope, etc.). Even when repeatedly asked, she would insist that she did not know why she was unable to execute the requested action. She completed all unimanual activities successfully with her right hand.

2.6. Neuroimaging investigation

Three-dimensional MRI scan and SPECT scan were acquired in August 2002. The MRI images showed the right parietotemporal lesion demonstrated in the acute phase post-stroke with resolution of oedema, and no extension of the stroke. T2-weighted MRI images showed hyperintensities in deep and periventricular white matter in both hemispheres (Fig. 2).

Functional brain scanning with ^{99}Tc HMPAO SPECT showed that a large area of hypoperfusion in the right hemisphere involved not only the overtly damaged right parietotemporal structures, but also a significant part of the right frontal lobe. In detail, the dorsolateral and ventral right frontal cortex showed a marked reduction in blood flow affecting the primary motor cortex and the posterior part of the middle and inferior frontal gyri (Fig. 3). These investigations were carried out while EN still showed anosognosia and this symptom still persisted in December 2002 when the patient's carers were last contacted.

In summary, the case description shows that EN remained anosognosic for left hemiplegia and hemianaesthesia 26 months after a right sided haemorrhagic stroke in the territory of the middle cerebral artery. Her neuropsychological profile demonstrated a preservation of language, short term and long term episodic memory, abstract reasoning, praxis and object recognition. Semantic and autobiographical aspects of memory were also spared, as was face recognition. She had no visuoconstructive deficits, although her actual performance showed the influence of left unilateral visuospatial neglect. EN had reduced verbal fluency with more pronounced impairment in the phonemic task. Her reading and writing abilities were affected by mild neglect dyslexia and dysgraphia. There was visual and auditory left extinction. Motor and sensory functions of the left upper and lower limbs were lost.

The disturbances of awareness were to a degree dissociated in that she was unaware of her motor disability and how

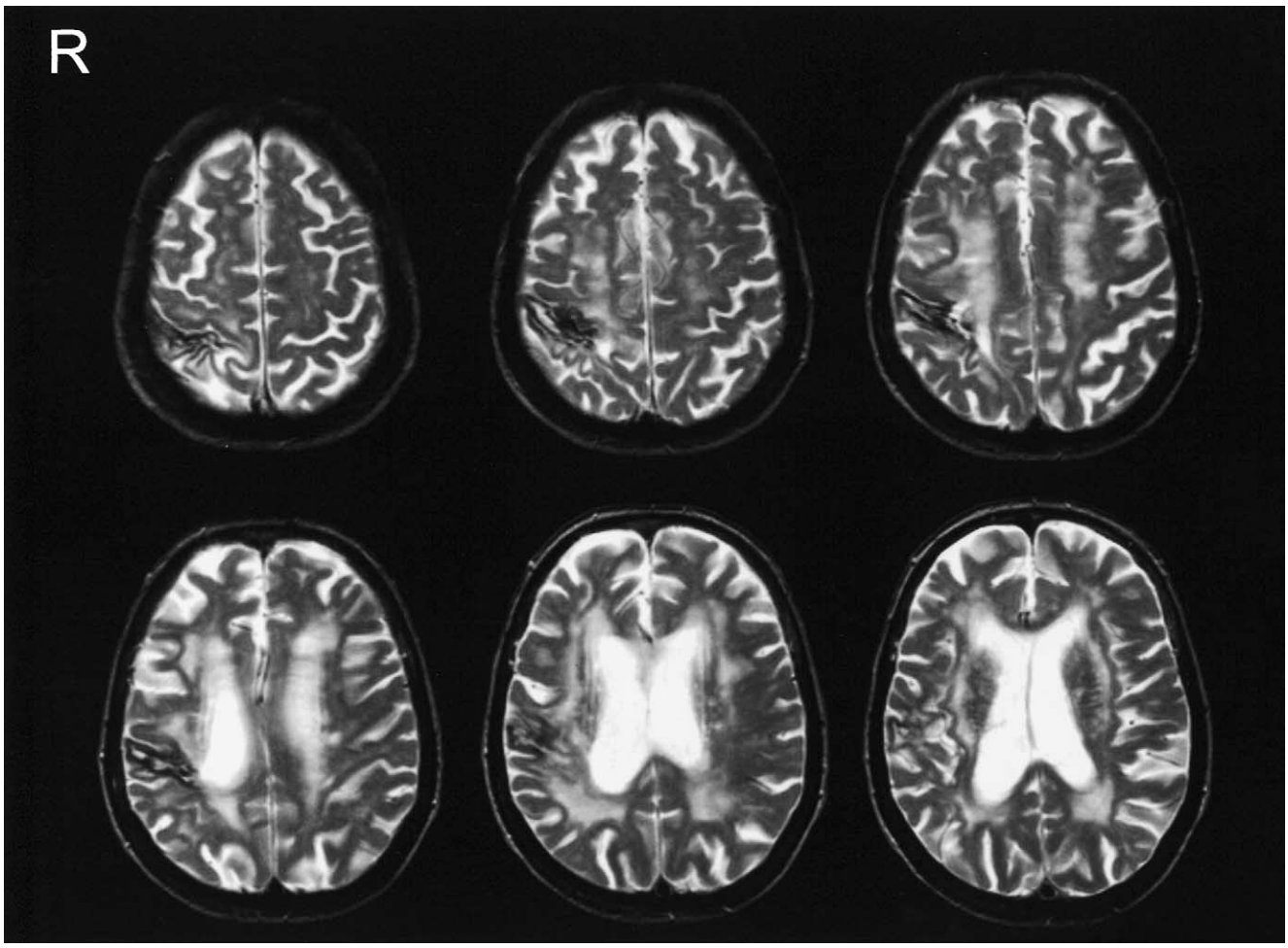


Fig. 2. Axial T2-weighted MRI images showing the residual damage to the right parieto-temporal structures.

this affected her performance on spatial and motor tasks, but she was aware of poor performance in some areas of cognitive functioning. For example, she was not aware of the neglect component in any graphic or lexical production but recognised her lack of artistic ability and poor handwriting. She was also fully aware of poor performance on the fluency tasks. There was no awareness of the profound anaesthesia in the upper and lower limbs, but she sometimes showed awareness of the left arm paresis. This deficit was acknowledged in response to specific questions, but in practice and during object use she was anosognosic for upper limb function. Her avoidance of certain actions with a rationalisation and her attempts to perform certain actions with her right hand rather than left hand, however, raise the possibility that EN might have had some implicit awareness of her inability although she resisted full explicit acknowledgment.

These neuropsychological deficits were associated with equally striking and persistent neuropsychiatric symptoms. EN confabulated extensively on themes of active excursions and bizarre acts of persecution. Some of these themes were relatively stable including a conviction that her mother was alive and concerned about the alleged transgressions against her daughter by the staff. An equally stable belief was that her surroundings were "home", and she seemed able to entertain apparently conflicting versions of her current reality as both persecuting institution and her familiar former home.

3. Discussion

To what extent do the existing theoretical models for anosognosia offer a plausible explanation for EN's lack of awareness and the persistence of this symptom? What significance, if any, should be attached to the other abnormalities in her mental state? Any explanation should try to account for transient and persistent loss of awareness, the prevalence of right hemisphere lesions and the patterns of dissociated awareness described in the literature.

EN's emotional reactivity without evidence of clinical depression, her alertness and the absence of global cognitive decline sufficiently refute any argument for reduced awareness based on disturbance of these elements in her mental state. There are formulations of anosognosic symptoms which point to the probability of profound sensory deficits

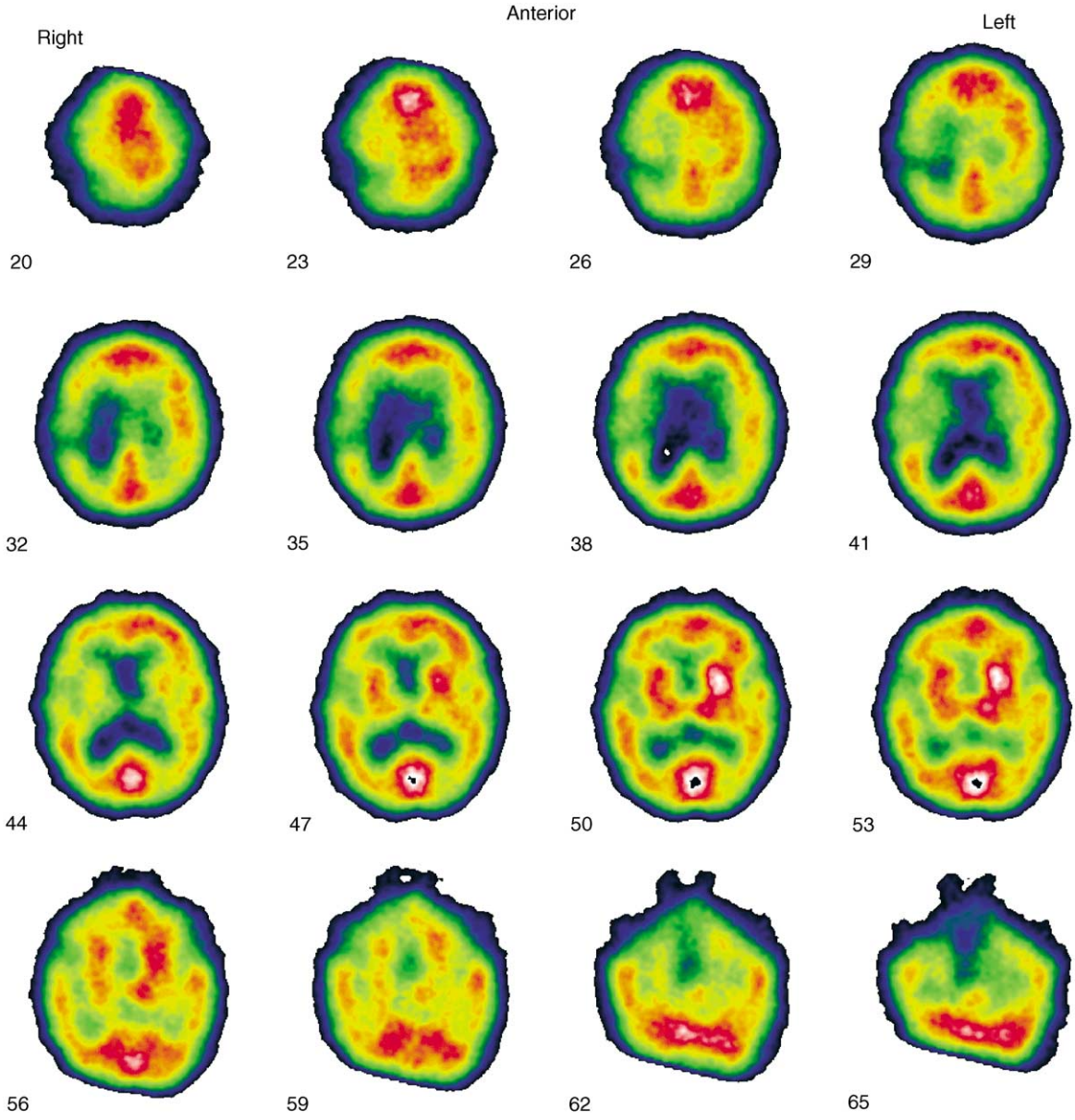


Fig. 3. T99 HMPAO SPECT images showing marked hypoperfusion in the area of the stroke, including the right parieto-temporal cortex and extending anteriorly to involve the right middle and inferior frontal gyri.

and sensory inattention leading to lack of central feedback and therefore an inability to experience the paretic limbs (Heilman, Barrett, & Adair, 1998). While this might contribute to explanation in some cases, EN indicated discomfort from a tight fitting shoe on her left foot while engaged in a drawing task. Neurological examination demonstrated a lack of acknowledged epicritic skin sensation in the left limbs, but she was sensitive to pinprick and so there may have been sparing of elements of the protopathic sensory pathways related to the perception of pain or discomfort. There are no published case studies bearing on the incidence of such sensory dissociations in cases of anosognosia. In a wider sense, the argument from impaired sensory feedback finds difficulty in accounting for lack of awareness in other cognitive domains of, for example, memory or reasoning.

EN's persistent anosognosia was associated with concurrent phenomena of lifestyle as well as examination triggered confabulations and with other reality dysfunctions. These included delusions based on autobiographical memory failures, place misidentification with double orientation and persecutory delusional beliefs about staff members and their intentions. Similar neuropsychiatric symptoms have been argued, on the basis of case and cohort studies in AD, to arise from regional neuropsychological failures in a context of disturbances in reality monitoring due to right hemisphere dysfunction (Feinberg & Shapiro, 1989; Förstl, Almeida, Owen, Burns, & Howard, 1991; Mizukami, Yamakawa, Yokoyama,

Shiraishi, & Kobayashi, 1999; Shanks & Venneri, 2002; Staff et al., 1999). Such symptoms are seen with highest frequency in patients with right frontoparietal dysfunction. EN did demonstrate some of the other behavioural changes which have been associated with frontal lobe dysfunction including occasional disinhibition and socially inappropriate behaviours together with apathy and loss of realistic motivation. Reduced verbal fluency was also recorded, but no other components of a formal dysexecutive syndrome.

SPECT examination showed an area of hypoperfusion in the right frontal lobe extending beyond the precentral cortex into lateral and orbital associative cortex. It is difficult of course to be certain in someone of this age whether the extended regional deficit demonstrated by rCBF measurement is wholly related to the stroke event. There is always the possibility that asymptomatic microvascular and/or degenerative brain disease (even developmental deficits) might contribute to the functional impairments following stroke. Her MRI also showed high grade white matter hyperintensities in both hemispheres. These white matter changes might have contributed, speculatively, to an overall reduction in cerebral reserve, but there was no indication of global cognitive decline, as might be expected if diffuse vascular brain disease were an important aetiological factor. Brain ageing may well be a contributory factor in this patient, but instances of confabulation and somatoparaphrenia can also be observed in younger populations.

When these neuropsychological, neuropsychiatric and neuroimaging findings are taken into account, it is reasonable to consider whether existing cognitive theories or modifications of them might offer some explanatory hypothesis accounting for this patient's symptoms. Most cognitive theories invoke higher order systems whose damage might lead to anosognosia. These conjectured systems might either be integral with the functional domain for which awareness has been lost (i.e. modular Bisiach et al., 1990) or superordinate and centralised, able to influence awareness across a range of sensorimotor and cognitive functions (McGlynn & Schacter, 1989). In the latter case, even the invocation of partial disconnection cannot account for the clinical observation of patients with loss of awareness for some symptoms but not for others. Neither modular nor centralised hypotheses seem to explain why the greatest proportion of left hemiplegic patients do not show anosognosia.

An attempted hypothesis at this level of cognitive functioning might be informed by theories of self monitoring and reality monitoring. The argument would likely find support from findings made in studies assessing the role of the right frontal lobe in the retrieval and monitoring of self related memories, as well as those evaluating the function of the right hemisphere in verifying the truthfulness of recollections (Craik et al., 1999; Fletcher, Shallice, Frith, Frackowiak, & Dolan, 1998; Metcalfe, Funnell, & Gazzaniga, 1995; Schacter, Curran, Galluccio, Milberg, & Bates, 1996; Shanks & Venneri, 2002; Venneri, Shanks, Staff, & Della Sala, 2000). It is worth considering in the first place, therefore, whether cognitive disturbances suggesting a wider dysfunction of frontal lobe functions are observed either transiently or chronically in association with neurologically defined lack of awareness. Similarly, it may be relevant if the brain damage and dysfunction identified in cases of anosognosia might in some instances extend beyond the lesion in the motor cortex to compromise frontal lobe function and contribute to related disturbances of normal consciousness, particularly when the disorder of awareness is persistent.

The most straightforward point to make is that in all cases of anosognosia for hemiplegia there will be direct or indirect damage to the motor cortex in the frontal lobe. This must, from neurological reasoning, involve a significant part of the precentral gyrus and/or its subcortical connections and associated subcortical structures that even if structurally spared must be functionally damaged. In the acute phase following stroke, there is likely to be, at the least, a more anterior and ventral region of oedema and parenchymal reaction encroaching on the associative cortex of the frontal lobe. This raises the possibility that in transient cases of anosognosia for hemiplegia, there might be a more extensive or regionally significant frontal dysfunction dependent on individual variation in the local consequences of acute brain lesion either directly or via disconnection. The extent of any frontal lobe dysfunction has rarely been assessed in acute stroke patients with anosognosia for hemiplegia either by functional imaging, or by the appropriate detailed neuropsychological assessment. Evaluation of frontal lobe function following stroke has been problematic because of lack of test specificity, conflicting findings and the challenge of determining which frontal function might be most closely involved in the development of anosognosia. Conventional psychometric measures may not be appropriate for the detection of subtle deficits related to abnormal reality judgements. In this respect single case studies are perhaps of limited value without the kind of neuroimaging evidence available in this case, but the detailed study of individual patients may generate working hypotheses for future research. One report records anosognosia and confabulation in a patient with right frontal hypometabolism after a stroke in the area of the right anterior choroidal artery (de la Sayette et al., 1995). Functional neuroimaging findings are available in degenerative brain disease, but these conditions are clearly different in their clinical presentation and determinants. In AD the appearance of anosognosia has been associated with a reduction in right frontal regional cerebral blood flow (Starkstein et al., 1995) and with neuropsychiatric symptoms (Migliorelli et al., 1995).

The association between loss of awareness of hemiplegia and right hemisphere dysfunction has also been examined using the Wada technique. Some authors have found that unawareness of hemiplegia is preferentially associated with anaesthesia of the right hemisphere while others claim that anosognosia can be observed with induced dysfunction of either hemisphere (see Heilman et al. (1998) for a

review). Most of the subjects tested for anosognosia with the Wada technique had neurological disorders, but they were not typical of the older patients who most often present with stroke and loss of awareness, and are likely to have associated vascular and degenerative brain disease (Koyama, Mochizuki, Mitsui, & Marui, 1998; Mineura et al., 1995). The finding that patients undergoing the Wada procedure showed no consistent association between anosognosia and confabulation on a test of simple sensory discrimination (Lu et al., 1997) might, therefore, be set against the finding that lack of awareness and confabulation tend to associate in a more representative population after stroke (Feinberg, Roane, Kwan, Schindler, & Haber, 1994), although double dissociations can also be inferred from case descriptions reported in the neuropsychological literature. More generally, the suspension of hemispheric functions may not be strictly comparable to the regional dysfunctions seen in clinical practice.

When it has been possible to study the rarer chronic forms of anosognosia for hemiplegia, it is significant that such patients often have extensive right frontoparietal and subcortical lesions (e.g. Berti et al., 1998), and were reported to show verbal inventions about capacity for activity as well as more bizarre beliefs on the topic of their paralysed limbs.

In this respect, lesions and degenerations involving the frontal and parietal lobes of the right hemisphere have been associated with distinctive disorders of memory retrieval, visuospatial processing, veridical assessment and reality monitoring (e.g. Rapcsak, Polster, Comer, & Rubens, 1994; Venneri et al., 2000). Right sided frontoparietal brain damage has also been more commonly aligned with a range of neuropsychiatric symptoms including confabulation, delusions based on autobiographical memory falsifications, delusional misidentification syndromes and persecutory states (e.g. Binetti et al., 1995; Conway & Tacchi, 1996; Ellis, 1994; Shanks & Venneri, 2002; Venneri et al., 2000).

The evidence presented supports a conclusion that EN had a significant deficit involving the associative right frontal cortex in addition to the precentral gyrus. The content of EN's many confabulations about activities logically implies, at the highest cognitive level, the lack of explicit awareness of disability demonstrated by the neurological and neuropsychological examinations. Loss of awareness following stroke would seem to entail a, usually transient, disturbance in the structure of normal consciousness and the experiential content seems often to include a process of "filling in" manifest as confabulation. This alteration in awareness may be supported by contingent right frontal and/or parietal dysfunction causing a suspension or change, albeit often partial, in the ability to monitor and check the 'real' and especially to assess the truth of mental contents. In acute and milder cases these disturbances would likely fall short of the more florid and established reality distortions seen in EN. Failures in reality monitoring/checking from this frontal impairment are known to facilitate the appearance of abnormal beliefs whose content is then determined by emotional and motivational factors as well as by specific regional dysfunctions (Shanks & Venneri, 2002; Venneri et al., 2000).

In this case at least, therefore, there may be a barrier to the natural awareness of hemiparesis as part of a wider syndrome of reality monitoring failures which also have allowed the development of other abnormal beliefs. The content of these false beliefs often seems psychogenically driven and either gainful for the patients or explicative in terms of their changed appreciation of their environment. The neuropsychiatric symptoms may be viewed, therefore, as integral with whatever mechanisms are promoting persistent anosognosia, and this approach might have heuristic value for the interpretation of other similar cases and for future research.

This hypothesis could be tested by studies of reality assessment and susceptibility to provoked confabulation in acute or subacute stroke patients with and without lack of awareness. Cases of hemiparesis with anosognosia might show a more extensive or subregionally relevant disorder of frontal lobe function, and this also could be assessed by functional brain imaging. Detailed evaluation of the idea that frontal dysfunction may be an embedded factor in the genesis and maintenance of lost awareness after stroke will, therefore, require data from the study of both transient and persistent cases with or without demonstrable frontal involvement. The present case, however, offers evidence that a functional lesion extending into the associative cortex of the right frontal lobe, as shown by the rCBF pattern, can lead to distinctive neuropsychological and phenomenological changes in chronic anosognosia. These features suggest the probable contribution of such regional dysfunction (either transient or persistent) in the genesis and endurance of anosognosia.

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