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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