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Measuring and modifying abnormal social cognition in frontal variant frontotemporal dementia

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Abstract

We describe a 57-year-old man (MW) with frontal variant frontotemporal dementia (fv-FTD) who presented with a long history of drinking problem and marital disharmony followed by gradual changes in personality with disinhibition, stereotypic checking, overeating and a decline in self-care. Structural MRI imaging confirmed marked frontal atrophy involving particularly the ventromedial region. Performance on standard tests of frontal executive function was largely unremarkable and MW obtained a perfect score on the Mini-Mental State Examination (MMSE). In contrast, an experimental battery of tasks designed to evaluate theory of mind (ToM) revealed marked deficits. MW's challenging and disruptive behaviours, notably obsessive checking of car suspension by rocking, and wandering, responded to behavioural modification regimes adapted from the neurorehabilitation literature. In conclusion, deficits in ToM may underline the gross abnormalities in social conduct, which characterise fv-FTD; ToM appears to dissociate from frontal executive function; and behavioural modification approaches can be of benefit in this disorder.

Keywords: Theory of mind; Social cognition; Frontotemporal dementia; Frontal lobes

Introduction

Frontotemporal dementia (FTD) is the term now preferred to describe non-Alzheimer type dementia affecting the frontal and/or temporal lobes [1–4]. FTD represents a continuum: at one end is the temporal form, known more commonly as semantic dementia [5], and, at the other, the frontal variant of FTD [6]. The temporal form manifests as progressive breakdown of knowledge causing anomia and comprehension impairment, while the frontal variant (fv-FTD) is characterised by changes in personality and social behaviour [6,7]. Patients with fv-FTD often present the psychiatric services where they cause considerable diagnostic difficulty particularly since they often perform within the normal range on traditional tests of executive function.

Studies employing single case neuropsychological methods have contributed greatly to our understanding of a wide range of cognitive processes. For instance, the intensive investigation of semantic dementia has been particularly valuable in informing models of memory and language [8]. This has, in turn, aided the diagnosis of semantic dementia [9]. The single case approach in semantic dementia has been facilitated by the availability of tasks and procedures derived from the field of cognitive neuropsychology. The use of these techniques is, however, less easily applicable to patients with fv-FTD who present with neuropsychiatric symptoms.

Although numerous clinical reports have established the link between frontal pathology and antisocial behaviour [10], only very recently have testable models emerged, which can be applied with patients with breakdown in social cognition. For instance, Stone et al. [11] adopted a neurodevelopmental stance, based on the concept of theory of mind (ToM, described more fully below) and were able to successfully measure deficits in social cognition in their adult patients with acquired frontal pathology.

The patient described in this paper presented with marked behavioural disturbance in the context of fv-FTD but showed little evidence of classic neuropsychological deficit. We focus on four key issues. Firstly, following Stone

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et al. [11], we address whether tests of ToM can measure deficits in social cognition as displayed by this patient. ToM, the ability to make inferences about the mental state of others, is held to be one of the principal psychological constructs underpinning normal human social functioning [12]. It develops in stages throughout childhood with increasing complexity and can now be measured using a range of tasks. We were interested, therefore, in whether the social dysfunction seen in our patient might reflect loss of ToM. The second issue concerns the dissociation of measures of ToM from traditional executive dysfunction. Thirdly, we consider the question of the early precedents of full-blown dementia in our patient. Finally, we describe attempts to modify these challenging behaviours using a range of techniques adapted from the neurorehabilitation literature.

Case history

MW is a 57-year-old right-handed male with a diagnosis of fv-FTD who is currently a day and respite patient on an inpatient psychiatric ward (January 2002). He left school at the age of 16 with an exemplary record having been head boy. He then obtained a diploma in engineering and worked with his father and brother in a small family firm that manufactured machine tools. He was married with two children. He first presented to psychiatric services at the age of 38 with a 10-year history of excessive alcohol consumption. Within the previous 3 days, he had undergone a period of forced abstinence to allow hand surgery. He admitted to experiencing hallucinations such as seeing rats and insects around the house and he felt compelled to hunt for them. He also believed he had special powers such as the ability to buckle bicycle wheels by thought. His wife described him as believing that people were trying to break into the house. He tried to climb up and jump off the wardrobe and wandered around the village they lived in at night. MW was assessed as suffering from acute alcohol withdrawal and was subsequently admitted to hospital for successful detoxification.

MW was able to return to work and put his alcohol problem behind him. However, in an interview 6 months later, his wife stated that her husband had changed from being the sociable outgoing person she married. She said that she received no appreciation from him and that he generally thought that she was a "pain in the neck." She never knew what he was thinking and was never sure where she was with him. There was now very infrequent sexual contact and indeed they had started to sleep in separate bedrooms. Attempts were made to engage the couple in marital therapy. Correspondence at the time indicates this was unsuccessful as both partners felt the real issue was that MW needed individual therapy to "cure his problem of not being able to do things."

Four years later, at the age of 42, MW commenced drinking again but at this stage was still working. His wife reports that she was puzzled at the time as to why her husband had started drinking again as he had weathered personal losses, such as the death of his sister, and crises at work without resorting to alcohol. At the age of 47, he was admitted again for detoxification following the loss of his driving licence due to crashing his car while drunk. He was discharged on a prescription of disulfiram that his wife administered to him.

MW's next contact with psychiatric services was in 1997 at the age of 53. He was still at home living with his wife; however, 18 months previously the family firm had been declared bankrupt. It transpired that the business had been in financial trouble for some time and, since 1992, MW had made several disastrous decisions involving the couple's savings and mortgage. He was now unemployed and the couple were heavily in debt to the Inland Revenue with little income. His wife stated that 2 years prior to this contact with services, her husband's personality had undergone changes. He had become much more subdued. He would not sit down for family meals, preferring to eat alone. He failed to appear at his daughter's graduation.

Over the course of the next 2 years (1997–1999), MW's behaviour had progressively deteriorated. He became more disinhibited, for example, asking people for money or stealing. He became over-preoccupied with counting and checking. He made unnecessary purchases of food, developing a preference for sweet items. His mood at times was euphoric and he would giggle inappropriately and for no reason. He developed a paranoid ideation that his wife was intent to harm him and that people were going to steal from him. This led to obsessional checking of doors and windows in the home.

He was admitted for assessment, at the request of his wife, in early 1999 after 6 months of escalating and extreme behaviour. On admission, he was found to have grandiose delusions about selling machinery to the Falkland Island in a multimillion pound deal, and did not appear to have any insight into his unemployed status or that there might be anything psychiatrically wrong with him. Conversation with MW was difficult due to his preoccupations. His speech was monotonous but language output was syntactically, phonologically and semantically correct. On the ward, he displayed some limited checking behaviour of windows in the evenings and was at times inappropriately cheerful. In general, however, his behaviour was not as erratic and disruptive as his wife had described at home. He scored 30/30 on the MMSE, but it was noted that his memory for past events was inaccurate. The diagnosis on discharge was of early cognitive impairment most probably related to alcohol abuse.

Six months after this admission, he was seen in the Addenbrooke's Memory Clinic. His behavioural problems had become more marked and the clinical presentation had intensified. He would only ever eat sweet foods such as chocolate, which he had started to hoard. His daily routine had become stereotyped with a fixation on time. He had