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SOCIAL BEHAVIOUR VS. PSYCHIATRIC FEATURES OF FRONTOTEMPORAL DEMENTIA
Clinical report of two cases

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SUMMARY

Behavioural disturbances are prominent in frontotemporal dementia (FTD), a focal, non-Alzheimer type of dementia. Although most patients with FTD present with socially inappropriate behaviour, compulsive-like acts, poor insight and disinhibition, the presence of psychiatric features including delusions, hallucinations, and paranoia can lead to a misclassification of FTD as psychiatric disorder. In the absence of cognitive deficits non-experts fail to recognize these social changes as dementia symptoms.

We report two individuals who met current clinical criteria for behavioural or frontal variant FTD (bv-FTD), with the aim of distinguishing between psychotic symptoms and the often bizarre personality and behaviour change found in FTD. Also we review the literature on the noncognitive neuropsyhiatric manifestation of this disorder.

Clinical findings presented, and a literature review, indicate that psychotic symptoms are rare in FTD. Better awareness of behavioural symptoms in clinical practice is necessary in order to avoid misdiagnosis of FTD as psychiatric disorder.

Key words: *bv-frontotemporal dementia - behavioural disturbances - psychotic symptoms*

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INTRODUCTION

With baby boomers now reaching late middle age, degenerative diseases are becoming an increasingly important health issue. One such disorder, frontotemporal dementia (FTD) is particularly devastating to patients and their families, as symptoms include changes in behaviour and the erosion of personal relationships, often during the earliest stages. These disease features place great demands on caregivers and on society at large. FTD is a focal dementia of non-Alzheimer type, clinically characterized as either behavioural or aphasic variants with prominent frontal and temporal lobar atrophy (Neary et al. 1998, McKhann et al. 2001). FTD is the most common cause of early onset dementia after Alzheimer's disease (AD) (Ratnavalli et al. 2002). Once subsumed under the diagnosis of Pick's disease (Pick 1892), this heterogeneous group of focal dementias was unrecognized for many years. The behavioural variant of FTD (bv-FTD), the most common manifestation of FTD, is primarily characterized by disinhibition or apathy, socially inappropriate behaviour, compulsive-like acts, and poor insight which can all lead to a misdiagnosis of FTD as a late onset schizophrenia or another psychotic disorder (Passant et al. 2005, Mendez et al. 2008). The bv-FTD has also been called frontal variant FTD or fv-FTD. Due to the symptoms, FTD can be mistaken for AD, or another psychiatric disorder (Mendez et al. 2007). Therefore, the evidence of possible psychotic symptoms (delusions, hallucinations, and paranoia) must be distinguished from the often bizarre personality and behaviour changes of FTD. The overlap between FTD and psychotic symptoms exists and therefore may lead to misleading diagnoses.

Here, we present two cases of bv-FTD and discuss the difference in clinical presentation between the often bizarre personality and behaviour changes in FTD and psychotic symptoms (delusions, hallucinations, and paranoia). In the absence of pathology, investigators depend on the clinical criteria in the diagnosis of FTD.

CASE REPORTS

Case 1. A 54-year-old right-handed man was admitted to the University Psychiatric hospital Ljubljana, because he was not up to all the demands of his work. He had been a bus driver for several years, until he caused a traffic accident. Following the event his employer moved him to a less demanding job, which he could not fulfil and remained unsuccessful in performing even the least demanding operations. His co-workers had complained about his smell and lack of his personal hygiene. He never changed his clothes. Although his co-workers bought him new clothes, he ignored them. They noticed that he became apathetic and unconcerned. His father died four years ago, while his mother was still alive. None of them had been reported to have dementia. He had never married; he has siblings, who described him as an introvert throughout his life. During the last couple of years, however, his siblings noticed his increasing lack of social interests.

He was neither interested in family visits, nor did he answer any phone calls. On examination, his ability to follow conversation was severely reduced, he answered only simple questions. However, he spontaneously commented on events and persons, often inappropriately. On Mini Mental State Examination (Folstein et al. 1975) he scored 23/30. No failure was observed on orientation items, receiving 10/10. On the Brief neuropsychological cognitive examination he scored 14/30 (Tonkonogy 1997), indicating moderate cognitive impairment with mental flexibility and attention deficits mostly compromised. His comprehension, logical reasoning, and abstract thinking were reduced. His short-term, long-term memory and recognition tests were also impaired. Several intrusions were also noticed. He was unable to perform Trailmaking A test (Armitage 1946) and tests of motoric control. On verbal fluency test (Thurstone & Thurstone 1949) he was able to generate only three correct words in three minutes. No symptoms of psychosis or affective disorders were observed. The rest of the neurologic, general medical examination, and laboratory tests were normal. An MRI scan revealed mild bilateral frontotemporal atrophy. EEG showed no significant changes.

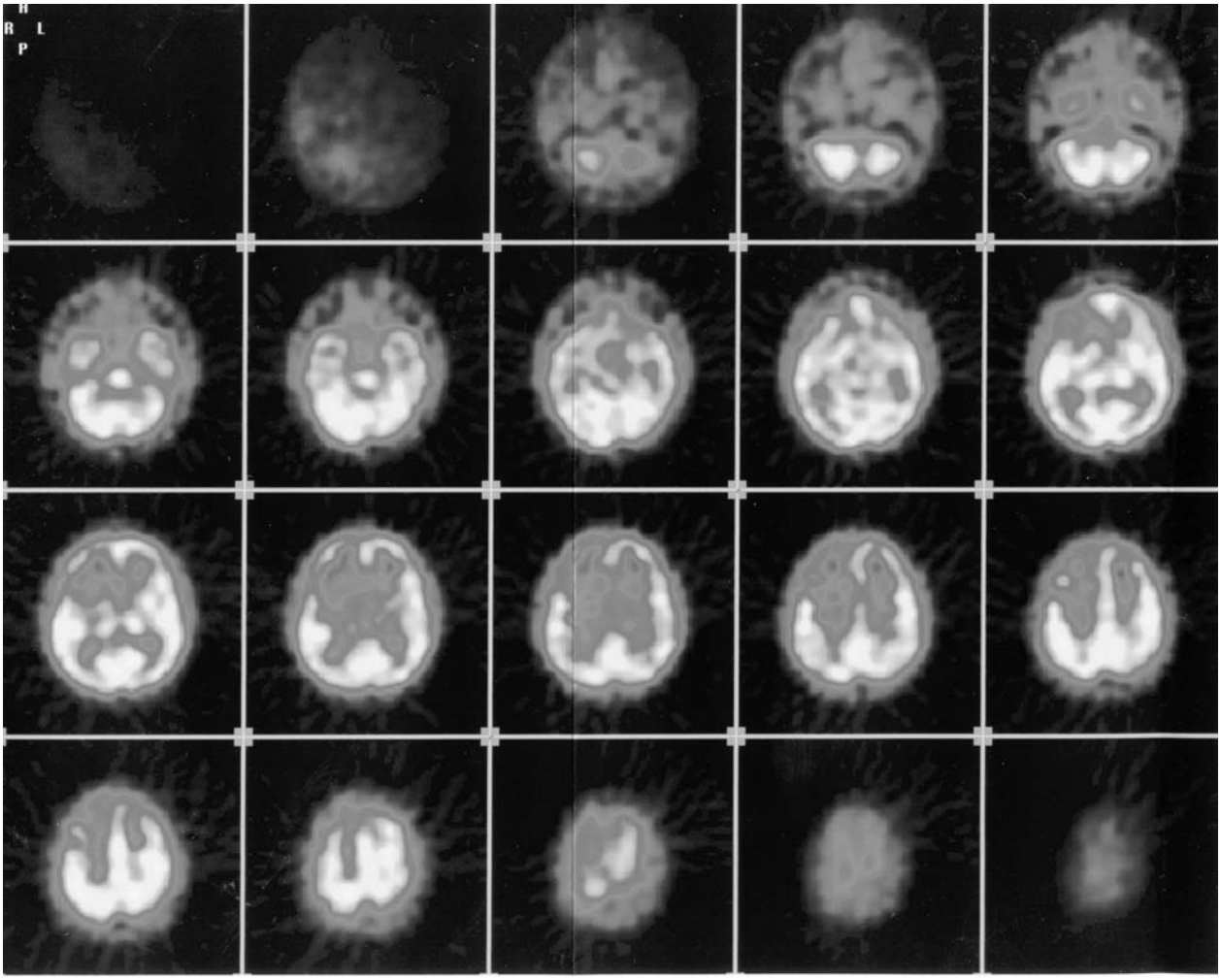

Figure 1. A brain SPECT study- in axial sections, the frontal part of the brain revealed severe perfusion deficits, which appears as a darker, redder colour more prominent on the right side, case 2.

Case 2. A 77-year-old right-handed man presented with a history of gradually increasing shopping difficulties e.g. pathological collection and storage of objects in his apartment and car. For the last few years, he became untidy and hoarded rubbish. In his apartment there was not enough room even for sleeping. Due to severe hygienic neglect and bad smell spreading out from his apartment, neighbours demanded that sanitary inspectors intervene. He could not, however, understand what the neighbours were arguing about. He thought that the smell was due to a malfunction of the sewage system. He completed technical college, was married twice, and has three children. For the last two years he was living alone, not even allowing his children to visit him at home. His father had dementia of late onset.

On examination, the patient was talkative with fluent speech which was sometimes difficult to interrupt. Comprehension was normal. No symptoms of psychosis or affective disorders were observed. On MMSE, he scored 29/30 (Folstein et al. 1975). A more detailed neuropsychological exam showed moderate cognitive decline. On the Brief neuropsychological cognitive

examination he scored 18/30 (Tonkonogy 1997). He had great difficulty in naming pictures of common objects, despite the fact that he was able to describe their function. Similarly he was unable to recall names of actual political figures. He scored in the normal range on tests of visuospatial skills, but his abilities were significantly reduced on tasks requiring processing of new, incomplete or unconventional information. On verbal fluency test (Thurstone & Thurstone 1949) he was able to generate seven correct words in one minute. Mental flexibility was reduced, he was unable to shift mental sets, and several perseverations were noticed. An MRI scan revealed bilateral frontotemporal atrophy and a broader Sylvian fissure. A brain SPECT (single-photon emission computerised tomography) showed severe frontal perfusion deficit, more prominent on the right side (Figure 1). Except for the presence of palmomental and snout reflexes, the neurologic exam was normal. The rest of the medical examination, and the laboratory tests were normal.

DISCUSSION

Clinicians often attribute psychotic symptoms to FTD. This report of two cases who met clinical criteria for FTD (Neary at al. 1998, McKhann et al. 2001), however, does not document a specific tendency for delusions, hallucinations or paranoid ideation at presentation and follow up over a two year period. Although FTD may resemble schizophrenia (Stone et al. 2003), many reports of FTD patients with delusions or hallucinations were probably referring to alternative diagnoses. As Köhler et al. (2007) stated, psychosis in old age seems much more common than previously thought and so does psychosis with late onset. Of all cases, they found that 21% were over 50 years of age at illness onset. A typical case of schizophrenia should not be mistaken for other disorders because of its lifelong course.

There have been reports presenting a clinical course consistent with FTD with psychotic symptoms (Miller et al. 1993, Waddington et al. 1995, Neary et al. 1998, Köhler et al. 2007). In contrast, in more extensive series of fv-FTD, there have been reports of patients with delusions and hallucinations (Edwards-Lee et al. 1997, Bozeat et al. 2000). Recently, Mendez and colleagues (Mendez et al. 2008) reported that in a large cohort on 86 FTD patients, only two patients (2.3%) suffered delusions, one of whom had paranoid ideation and no hallucinations, while 17.4% of AD patients had delusions and paranoia. In contrast, another study failed to find any psychosis among well-characterized patients with FTD (Le Ber et al. 2006). Finally, three clinicopathological series with definitive frontotemporal lobar degeneration on autopsy found no psychosis among 19 patients (Passant et al. 2005), no hallucinations among a cohort of 48 patients (Liscic et al. 2007), and only one clearly defined paranoid-hallucinatory state among 20 patients (Gustafson 1993).

The infrequency of delusions, hallucinations or paranoia in FTD, particularly as compared to AD, is important for what it reveals about brain mechanisms of psychosis. Lesions in the mesiotemporal area may result in delusions that involve fear or a disturbed sense of threat through disruption of limbic functions that link perception to emotional states (Richardson & Malloy 2001). Recently, a 65-year-old woman with depression, personality changes and bizarre delusions with acoustic and gustatory hallucinations was reported (Kerssens et al. 2006), but her symptoms and course were more suggestive of psychotic depression than of FTD. Depression and dementia, in particular of frontal-lobe type dementia, should be looked for in elder individuals who live in extreme squalor, extreme self-neglect, unhygienic conditions, accompanied by a self-imposed isolation called a Diogenes syndrome (Clark et al. 1975). These elderly patients tend to be aloof, suspicious, emotionally labile, aggressive, and reality-distorting individuals. Apathy is a common symptom in depression as well as in FTD. However, in depression it is accompanied by depressive mood or depressive thoughts, which are usually not present in FTD. Anatomically, the bv-FTD syndrome is associated with predominantly orbitofrontal cortex (OFC) and dorsolateral frontal atrophy (Viskontas et al. 2007).

The language impairments are not commonly seen in bv-FTD (Wittenberg et al. 2008), therefore, the impairment in word fluency, which we found in our cases may present „executive" dysfunction such as initiation and activation of retrieval strategies that are thought to be dependent on frontal lobe functions, particularly in the left hemisphere (Waarkentin & Passant 1998). Also, occasional memory loss in FTD patients may in part reflect word finding difficulties stemming from language dysfunction (Liscic et al. 2007). Therefore, the two presented cases together with a literature review, suggest that revisions are needed in order to improve the already established diagnostic and research criteria for bv-FTD (Raskovsky et al. 2007). Improvements in differential diagnosis will most likely come from incorporating behavioural and social measures, since these symptoms define the disorder and are the first to emerge.

CONCLUSION

The relative paucity of psychotic symptoms in FTD has several implications. First, these are important implications for differential diagnosis, while in the past 21% of autopsy-confirmed FTD cases have been misdiagnosed with psychosis in FTD or schizophrenia (Passant et al. 2005). Second, the lack of psychosis in FTD may indicate that the temporal-limbic system is necessary to develop paranoid false beliefs. Further work is required in order to corroborate these clinical findings, particularly among FTD patients followed by autopsy.

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