Editorial

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Neuropsychiatry in Clinical Practice: The Challenge of Diagnosing Behavioral Variant Frontotemporal Dementia

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Editorial

The behavioral variant of Frontotemporal dementia (bvFTD) is an insidious neurodegenerative disease associated with progressive degeneration of the frontal lobes, anterior temporal lobes, or both [1]. Alterations in social cognition represent the core symptoms of bvFTD resulting in emotional disengagement and socially inappropriate responses or activities [2,3]. As is apparent in revised consortium criteria, additional neuropsychiatric symptoms including apathy and stereotypical and impulsive behavior are prominent in the clinical presentation [4]. Consequently, both neurodegenerative diseases and primary psychiatric disorders are crucial in the challenging differential diagnosis.

The differentiation between bvFTD and Alzheimer's disease (AD) has become easier by the use of biomarkers that are able to identify underlying AD pathology, such as the amyloid- β (A β) and tau [1,5]. However, to distinguish bvFTD from psychiatric disorders can still be difficult, particularly since biomarkers for bvFTD are less robust [6]. Previous studies indicated that as a result of symptomatic overlap between bvFTD and psychiatric disorders, bvFTD patients are clinically often mistaken for psychiatric patients and vice versa [7-10]. The current clinical criteria for bvFTD require that "if behavioral disturbance is better accounted for by a psychiatric diagnosis, a diagnosis of bvFTD has to be excluded" [4].

Despite clinical overlap, bvFTD patients do not often fulfill formal criteria for a psychiatric diagnosis, suggesting that it is valuable to apply formal criteria for psychiatric disorders [11]. Careful clinical phenotyping of overlapping symptoms can help to distinguish bvFTD from psychiatric disorders in clinical practice (**Figure 1**) [12,13].

	Overlap with bvFTD	Different from bvFTD
Major depression	• Apathy • Psychomotor agitation • Inhibition	• Disease awareness • Distress
Mania	• Disinhibition • Inappropriate behaviour • Inflated self esteem	• Reduced need of sleep • Fast disease course • Self-destructive behaviour
Schizophrenia	• Negative Symptoms • Reduced affect • Poverty of speech • Apathy	• More often delusions and hallucination
Obsessive Compulsive Disorder	• Stereotypical behaviour • Clinging to structure • Rituals	• Disease awareness • Distress • Young age of onset • Stereotypical behaviour is driven by fear
Autism Spectrum Disorders	• Clinging to structure • Rituals • Solitary minded	• Lifelong pattern • Problematic behaviour may be influenced by a well structured environment

Figure 1 Overlap and differentiation between bvFTD and psychiatric disorders in clinical practice.

The value of different symptom rating scales and clinical tools has been proven useful in clinical practice in case of suspected bvFTD when a psychiatric disorder is also probable (**Figure 2**) [11,14,15].

Presenting symptoms	Supporting probable or definite bvFTD	Supporting a psychiatric disorder
Mood and apathy	• Low score at the *Montgomery Asperg Depression Rating Scale (MADRS)*	• High score at the *Montgomery Asperg Depression Rating Scale (MADRS)* • High score at PANSS items *Tension, Anxiety and Guilt feeling*
Stereotypy	• High score at the *Stereotypical Rating Inventory*	• Low score at the *Stereotypical Rating Inventory* • Being male
Disinhibition	• Not fulfilling DSM criteria for Bipolar Disorder	• fulfilling criteria for Bipolar Disorder
Loss of empathy	• Low score at *Ekman 60 Faces test* (preferably score <20)	• High score at *Ekman 60 Faces test* (preferably score >46)
Psychotic Symptoms	• High score at PANSS items *Stereotypical thinking and Difficulty in abstract thinking*	• Low total score at the Negative subscale of PANSS
Indistinct behaviour	• Not Fulfilling formal *DSM criteria* for a psychiatric disorder	• Fulfilling *DSM criteria* for a psychiatric disorder
Symptom duration		
Long symptom duration without conversion from possible to probable bvFTD, lacking a genetic mutation	• Being female and absence of most of the following conditions: recent life events, mood problems, low intelligence	• Being male and one of the following conditions, or being female and at least 3 of the following conditions: recent life events, mood problems, cluster C personality traits, relationship problems, low intelligence

Figure 2 Clinical hallmarks and supportive measuring instruments in the differential diagnosis bvFTD and psychiatric disorders.

According to current criteria, the diagnostic certainty of bvFTD increases when Frontotemporal abnormalities are found on neuroimaging. In a large cohort of patients with late-onset behavioral changes, MRI had a sensitivity of 70% and a specificity of 93% for a bvFTD diagnosis [4]. The additional [18F]FDG-PET, when the MRI was inconclusive, had a sensitivity of 90% at the cost of a lower specificity (68%) [16]. [18F]FDG-PET is mainly useful when Frontotemporal hypo-metabolism is absent to exclude bvFTD diagnosis. The interpretation of neuroimaging results should especially be taken with caution in cases with a psychiatric differential diagnosis where [18F]FDG-PET is the only abnormal investigation and in cases with a genetic background where both MRI and [18F]FDG-PET can show a specific abnormalities [16-18]. Genetic screening especially for C9orf72 repeat expansion is emphasized [19,20]. particularly in cases with a remarkable (prolonged) disease course.

In clinical practice, bvFTD has a broad differential diagnosis including both neurodegenerative diseases and primary psychiatric disorders. The current criteria for bvFTD have clearly improved diagnostics but differentiating bvFTD from psychiatric disorders remains difficult. The challenge for the next decade is finding specific biomarkers for bvFTD on the one hand, and optimizing the neuropsychiatric diagnosis of bvFTD on the other hand. To this end, patient care for suspected bvFTD patients would be largely improved in a setting where neurologists and psychiatrists work hand in hand, ideally applying a consensus set of clinical rating scales next to their clinical expertise.

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