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The behavioral variant of frontotemporal dementia: An analysis of the literature and a case report

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ABSTRACT

The aim of this case report is to underline the importance of possible legal consequences of the behavioral variant of frontotemporal dementia (bvFTD). This disease is associated with antisocial behavior, impulse control disorder and cognitive and personality impairment, which are often the earliest manifestations of the bvFTD. One of the antisocial behaviors possibly associated with this neurodegenerative disease is pathological stealing. This case report is about a 50-year-old Italian man who had a regular life until 2010. In 2010 and 2011, some critical events occurred: he lost his job, his father-in-law, to whom he was particularly close, died, and his wife had a serious illness. He began to show symptoms of depression, a significant weight loss, apathy, poor self-care, and lack of interest in the activities of his family. He became disengaged from his prior activities, emotionally detached from his family and developed compulsive hoarding. Moreover, he had uninhibited behaviors, a memory retrieval deficit, executive dysfunctions and impulsive behaviors. In January 2012, the subject began stealing objects, particularly components of computer, without premeditation or concern for resulting legal actions. He was then diagnosed affected by bvFTD. He was charged with theft and attempted theft and the Court asked for a psychiatric evaluation, in order to analyze the effect of the neurodegenerative disease on his behavior. To answer to the Court, the Authors analyzed his history of life and made a mental examination. The subject was considered mentally insane at the time of his crimes. This is an example of the practical application in judicial cases of the latest knowledge and evidence in the literature about the frontotemporal dementia, a disease associated with antisocial behaviors that could create tensions with the criminal law. The focus of the paper is to explain how the behavioral symptoms of bvFTD can have legal implications and how to deal with legal aspects of the behaviors induced by a neuro-psychiatric condition, such as bvFTD.

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Contents

1. Introduction

Forensic psychiatrists were asked to evaluate a 50-year-old man afflicted by a neurodegenerative progressive disease with behaviors' anomalies, defined as frontotemporal dementia (FTD).

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The goal of this evaluation was to analyze the subject's mental condition at the time of his crimes and to express an opinion about his mental capacity, in order to decide if he was or not guilty by reason of insanity (Italian Penal Code, art. 85: "No one can be punished for an act envisioned by law as a crime if, at the time when it was committed, he/she did not have mental capacity. A person has mental capacity if he/she has the ability to understand right from wrong and to determine his/her volition and his/her behavior"), and his social dangerousness (Italian Penal Code, art. 203: "Socially dangerous is a person, even if not imputable or not punishable /…/ when it is likely to commit further offenses punishable by law as a criminal offense").

Before proceeding to the presentation of the case, it seems useful to retrace the most significant evidences related to the frontotemporal dementia (FTD).

2. Frontotemporal dementia (FTD)

Frontotemporal dementia (FTD) refers to a complex of syndromes resulting from degeneration of the frontal and temporal lobes (Kurz, Kurz, Ellis, & Lautenschlager, 2014).

The recent International consensus paper recognizes four clinical variants of FTD (Chare et al., 2014). The behavioral variant (bvFTD) (Rascovsky et al., 2011) and three primary progressive aphasia syndromes (PPA): semantic variant or sv-PPA; non fluent/agrammatic variant or nfv-PPA; logopenic variant or lv-PPA (Gorno-Tempini et al., 2011; Sieben et al., 2012). These syndromes have different initial presentations, which reflect the different localizations of the brain damage, but in all of them, frontal and temporal lobes are diffusely involved (Seltman & Matthews, 2012). Frequently, they are linked with movement and neuromuscular disorders (Rohan & Matej, 2014).

FTD is the second most frequent form of pre-senile dementia, following Alzheimer's disease (AD) among people below the age of 65 years (Ratnavalli, Brayne, Dawson, & Hodges, 2002; Rosso et al., 2003). The estimated prevalence is 15–22 per 100,000 in the age group between 45 and 65 years (Onyike & Diehl-Schmid, 2013; Ratnavalli et al., 2002). A Netherlands study found a prevalence of 3.6 per 100,000 among the 50- to 59-year-old and 9.4 per 100,000 among the 60- to 69-year-old population (Rosso et al., 2003). Knopman, Petersen, Edland, Cha, and Rocca (2004), studying the cases of frontotemporal lobar degeneration (FTLD) in Rochester, MN, from 1990 through 1994, identified an incidence rates (new cases per 100,000 person-years) of 2.2 for ages 40 to 49, 3.3 for ages 50 to 59, and 8.9 for ages 60 to 69, finding a prevalence higher than in previous studies in young people.

Male and female are equally afflicted (Johnson et al., 2005; Rosso et al., 2003); symptoms onset is usually before the age of 65 years with a mean age of approximately 58 years (Johnson et al., 2005), even if younger patients in their thirties have been described (Mackenzie, Foti, Woulfe, & Hurwitz, 2008).

Prognosis is poor: survival from symptoms onset varies between 6 and 11 years (Seltman & Matthews, 2012). Main causes of death are pneumonia, circulatory system failure and cachexia (Hodges, Davies, Xuereb, Krill, & Halliday, 2003; Nunnemann et al., 2011). Motor neuron involvement is associated with significantly poorer prognosis (Hodges et al., 2003).

Behavior and language changes, such as apathy, inertia, disihibition, mood changes, lack of empathy, tactless, semantic, agrammatic or logopenic aphasia, are usually the first symptoms. These symptoms are not specific and so it is difficult to make an early diagnosis of FTD and consequently the treatments are often delayed. According to a study performed by Woolley and colleagues, the 50.7% of patients with FTD received a prior psychiatric diagnosis, such as Depression, Schizophrenia or Bipolar Disorder (Wolley, Khan, Murthy, Miller, & Rankin, 2011). This percentage of prior psychiatric diagnosis is higher than in other neurodegenerative diseases.

The underlying pathology invariably leads to focal atrophy of frontal and temporal lobes and is referred to as Frontotemporal lobar degeneration (FTLD) (Neary et al., 1998; Perry & Miller, 2013; Riedl, Mackenzie, Förstl, Kurz, & Diehl-Schmid, 2014). Arnold Pick (1892), a neurologist in Prague, identified the first patient with FTDL in 1892 and so this lobar degeneration was named Pick's disease for more than a century. Now, the term Pick's disease identifies only a small subgroup of FTLD, while the term FTLD is used to identify a clinic-pathological complex, which includes two clinical syndromes (behavioral-variant of frontotemporal dementia and primary progressive aphasia) and three major underlying neuropathological subtypes (Riedl et al., 2014).[1]

A family history of FTD is found in approximately 40% of cases (Goldman et al., 2005; Rohrer et al., 2009), but only 10%–30% have an autosomal dominant inheritance pattern.[2]

2.1. The behavioral variant of FTD (bvFTD)

In this paper, we focus on the behavioral variant of frontotemporal dementia (bvFTD), since the subject we analyze in this work was diagnosed afflicted by this variant of FTD, underlining the behavioral aspects of this condition as they involve legal-psychiatric concerns.

The bvFTD is the most common subtype of FTLD (Onyike & Diehl-Schmid, 2013; Rabinovici & Miller, 2010) and it is a clinical syndrome characterized by early changes in personality and behavior with a progressive deterioration of personality, social comportments and cognition (Rascovsky et al., 2011).

The diagnosis of bvFTD is based on clinical diagnostic criteria. The International Consensus Paper, elaborated by the International Behavioral Variant FTD Criteria Consortium (FTDC) in 2011 and available on line established the criteria for a proper diagnosis of bvFTD, based on recent literature, particularly on the criteria established by Neary and colleagues in 1998 (Neary et al., 1998), and collective experience (Rascovsky et al., 2011).

The diagnostic criteria for bvFTD require the presence of a progressive deterioration of behavior and/or cognition based on observation or history (provided by a knowledgeable informant). If this criterion is satisfied, there are three further levels of certainty. The diagnosis of bvFTD is "possible" if there are almost three of six clinically discriminating features: early behavioral disinhibition, early apathy or inertia, early loss of sympathy or empathy, early perseverative, stereotyped or compulsive/ritualistic behavior, hyper-orality and dietary changes, neuropsychological profile with executive/generation impairment with relative sparing of memory and visuospatial functions. The diagnosis of bvFTD is "probable" if criteria for possible bvFTD are present and the subject exhibits significant functional decline and there are imaging results consistent with bvFTD. "Definite" bvFTD includes histopathologic evidence of FTLD on biopsy or at post-mortem (see Footnote 1), or the presence of a known pathogenic mutation (see Footnote 2) (Rascovsky et al., 2011).

The 5th edition of the Diagnostic and Statistical Manual of Mental Disorders, published in 2013 by the American Psychiatric Association,

[1] The histopathology of FTLD in fact shows neuronal loss and astrocytic gliosis, but differs with regard to the abnormal processing and deposition of specific protein. These three neuropathological subtypes are so characterized by abnormal accumulation of proteins, microtubule-associated protein tau- MAPT, transactive response DNA-binding protein with molecular weight 43 kDa—TDP-43, and tumor-associated protein fused in sarcoma protein-FUS (Mackenzie et al., 2008; Sieben et al., 2012). In particular, three forms of molecular pathology are described connected with bvFTD: Tau, approximately 40%, more often with Parkinson, TDP-43, approximately 40%, more often associated with neuromuscular disorders (MND) and FUS, approximately 40%, also more often with MND (Rohrer et al., 2009; Snowden, Neary, & Mann, 2007).

[2] Mutations in five unrelated genes have been identified and cause almost all the familial FTD cases: microtubule-associated protein Tau (MAPT), chromosome 9 open reading frame 72 (C9orf72), progranulin (GRN), charged multivesicular body protein 2B (CHMP2B), valosin containg protein (VCP) (Ferrari et al., 2014; Josephs et al., 2011; Kurz et al., 2014; Loy, Schofield, Turner, & Kwok, 2013; Sieben et al., 2012). Mutations of C9orf72, MAPT and GRN explain over 80% of cases in FTLD families with a strong autosomal dominant family history (Riedl et al., 2014).

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