ECT as a Useful Treatment Strategy for Difficult-to-treat FTD: A Case Report and Review of Literature

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INTRODUCTION

Frontotemporal dementia (FTD) defines a heterogeneous group of clinical syndromes marked by the progressive, focal neurodegeneration of the frontal and anterior temporal lobes. The variety of presentations causes special diagnostic difficulties that can significantly negatively influence patient care and advice for carers regarding the clinical status and prognosis. Those who develop FTD in midlife and have a predominance of behavioral symptoms may be mistaken for patients who have other psychiatric disorders like late-life depression or psychosis. When it first manifests in younger people, FTD might be mistaken for bipolar illness or schizophrenia. Due to the prevalence of repetitive, compulsive behaviors in Behavioural variants (bv-FTD), some individuals may initially be diagnosed with obsessive-compulsive disorder.

Presentation of FTD

The symptoms of FTD vary depending on the subtype of the disease. The behavioral variant of FTD is the most common subtype and is characterized by changes in personality and behavior, such as:

1. Loss of empathy and social skills
2. Lack of judgment and impulse control
3. Apathy and decreased motivation
4. Changes in eating habits
5. Repetitive behavior
6. Inability to follow social norms and rules
7. Inappropriate sexual behavior

The language variant of FTD is characterized by language problems, including difficulty speaking, understanding language, and writing.

Challenging Symptoms of FTD

The symptoms of frontotemporal dementia (FTD) can be challenging for patients and their families because they often involve changes in behavior and personality that are difficult to understand and manage. Some of the challenging symptoms of FTD include:
Loss of empathy and social skills: FTD can cause patients to become more self-centered and less interested in the needs and feelings of others. They may also struggle to understand social cues and norms, leading to inappropriate behavior in social situations.

Lack of judgment and impulse control: Patients with FTD may act impulsively and make poor decisions, such as spending money recklessly or engaging in risky behaviors.

Apathy and decreased motivation: FTD can cause patients to lose interest in activities they used to enjoy and become less motivated to engage with the world around them.

Changes in eating habits: Patients with FTD may develop unusual food preferences or eat inappropriately, such as eating too much or too little.

Repetitive behaviour: Patients with FTD may engage in repetitive behaviours, such as pacing, rocking, or repeating the same phrases or questions over and over.

Inability to follow social norms and rules: FTD can cause patients to disregard social norms and rules, leading to inappropriate behaviour in public or at home.

Inappropriate sexual behavior: Some patients with FTD may engage in inappropriate sexual behaviour, such as making sexual comments or advances towards others.

Difficulty in Diagnosis: FTD is often misdiagnosed as a psychiatric disorder such as depression, bipolar disorder, or personality disorder because of the similarities in symptoms.

Early onset: FTD can occur at an earlier age than other forms of dementia, with symptoms often appearing in individuals in their 50s or 60s.

Rapid progression: FTD is known for its rapid progression, with patients experiencing a decline in cognitive abilities and functional skills in a relatively short period.

Differential Diagnosis

Alzheimer’s disease: Alzheimer’s disease is the most common cause of dementia, and it can cause memory loss, language problems, and changes in behavior and personality that are similar to those seen in FTD. However, in Alzheimer’s disease, the symptoms typically start with memory loss, while behavioral and language problems characterize FTD.

Parkinson’s disease: Parkinson’s disease is a neurodegenerative disorder that can cause movement problems, such as tremors, rigidity, and slowness, as well as cognitive impairment. However, FTD does not typically present with movement problems.

Lewy body dementia: Lewy body dementia is a type of dementia that can cause visual hallucinations, movement problems, and changes in behavior and cognition. However, it also involves the presence of Lewy bodies and abnormal protein deposits in the brain that are not typically seen in FTD.

Huntington’s disease: Huntington’s disease is a genetic disorder that can cause movement problems, cognitive impairment, and changes in behavior and personality that are similar to those seen in FTD. However, Huntington’s disease also involves the presence of abnormal protein deposits in the brain that are not typically seen in FTD.

Primary progressive aphasia (PPA): PPA is a type of dementia that primarily affects language abilities, and it can cause difficulty speaking, understanding language, and writing. However, unlike FTD, PPA does not typically cause significant changes in behavior or personality.

Schizophrenia: Schizophrenia is a psychiatric disorder that can also cause changes in behavior and personality, but a progressive decline in cognitive abilities and functional skills characterizes FTD.

Major depressive disorder: Major depressive disorder can cause symptoms such as apathy, decreased motivation, and changes in appetite and sleep patterns, which are also seen in FTD. However, in FTD, these symptoms are typically accompanied by other behavioral and cognitive changes, and they are progressive and worsen over time.

Bipolar disorder: Bipolar disorder can cause changes in mood and behavior, such as impulsivity, decreased inhibitions, and changes in activity level, which are also seen in FTD. However, in bipolar disorder, these symptoms typically occur in the context of distinct episodes of mania or hypomania, and they do not typically progress over time.

Personality disorder: Borderline personality disorder can cause impulsivity, mood instability, and interpersonal difficulties, which can overlap...
with some of the behavioral symptoms seen in FTD. However, these symptoms are typically chronic and pervasive in borderline personality disorder, and they do not progress over time as in FTD.

**Obsessive-compulsive disorder (OCD):** OCD typically has an earlier onset chronic, fluctuating course, whereas FTD typically has a later onset and a more gradual, progressive course. In OCD, the symptoms are typically centered around specific fears or concerns and are associated with repetitive behaviors or mental acts to reduce anxiety or prevent harm. In FTD, the symptoms are typically related to changes in social and emotional functioning, such as loss of empathy, disinhibition, and apathy.

**How to differentiate FTD with other psychiatric disorders**

**Neurological symptoms:** FTD is characterized by the presence of neurological symptoms, such as changes in speech, motor function, and executive functioning, that are not typically seen in primary psychiatric disorders. These symptoms can be identified through neurological examinations and imaging tests.

**Onset and progression:** FTD typically have a later onset (in middle or late adulthood) and a more gradual, progressive course than primary psychiatric disorders, which often have an earlier onset and a chronic or episodic course.

**Nature of symptoms:** FTD symptoms are typically related to changes in social and emotional functioning, such as loss of empathy, disinhibition, and apathy, while symptoms of primary psychiatric disorders are more varied and may include mood disturbances, anxiety, or psychotic symptoms.

**Response to treatment:** FTD has no known cure or effective treatment, while primary psychiatric disorders can often be successfully managed with psychotherapy, medication, or a combination of both.

**Family history:** FTD is often associated with a family history of the disorder, while primary psychiatric disorders may or may not have a genetic component.

To differentiate FTD from other psychiatric disorders, a combination of clinical assessment, neurological examinations, and imaging tests such as MRI or PET scans are used. It is important to rule out other potential causes of cognitive impairment, such as vitamin deficiencies, infections, or tumors. A neuropsychological evaluation can also help identify patterns of cognitive deficits typical of FTD.

Here we present a case of a 58-year-old male in whom diagnoses were revised multiple times before finally reaching the diagnosis of frontotemporal dementia.

A middle-aged adult male of average build presented to the psychiatry OPD with chief complaints of anxiety for 9 months, decreased interaction for 5 months, fearfulness, agitation and disinhibited behavior for 4 months. Over the past 3 months, he also started to hold food and saliva in his mouth for a long and would maintain odd postures for prolonged durations. He developed repeated behaviors in the last 1 month.

As per the informants with the onset of illness, he started to express worries regarding his office work and inability to handle increased work stress as his promotion was announced. He would interact very less and had a blank face. Later, he also exhibited disinhibited behavior where he would undress and roam naked in the house. His repetitive behaviors included going and standing on the door multiple times, crawling on the floor and again sitting. Throughout the course, his interaction persistently decreased with spontaneous speech reduced to a few brief sentences. A dietary preference for eating only bananas was present. His sleep was 1 to 2 hours, and family members maintained his self-care. As per the family members, there was extreme agitation for the past 1-month where the patient would run here and there and if stopped, would push the family members such that they even might get hurt. Activities of daily living were severely affected. There was also the history of becoming fearful, lasting for a few hours throughout the illness. There was no history of any observable muttering behavior and no history of persistent, pervasive low mood. Past history was suggestive of a year-long episode of decreased interaction, anxiety, suspiciousness and sleep disturbance at 51 years of age following a similar promotion at the job. This was effectively managed using multiple antipsychotics. An episode of 3 months with hypomanic features occurred 3 years later. A positive family history of psychosis in the brother was present. Personal history and pre-morbid personality were non-contributory.
Before presenting to us, the patient had received olanzapine 10 mg for 3 months, amisulpride 200 mg, risperidone 6 mg, trifluoperazine 10 mg, haloperidol 10 mg, blonanserin 8 mg for 1-month each and escitalopram 10 mg for 15 days. However, the response was only transient. After catatonic symptoms began, family members consulted our department and a lorazepam trial with 4 mg showed improvement in catatonic symptoms and oral lorazepam 6 mg/day was added. However, even after 20 days, catatonic symptoms persisted and he was hospitalized.

Mental status examination found blunt affect, decreased speech productivity, and impairment in concentration and recall. Deficits in memory, fluency and visuospatial functions were prominent (Addenbrooke’s cognitive Examination III score - 53/100). The patient scored 13 on BFCRS mainly in ambleness, automatic obedience, grimacing, staring, echolalia and stereotypy was present (Bush-Francis Catatonia Rating Scale score - 13). CT head revealed diffuse cerebral atrophy with prominent frontal and temporal lobe involvement. Investigations including vitamin D3, S. folate, RBS, lipid profile, viral markers, LFT, KFT, CBC, and serum electrolytes were done to rule out the reversible causes of dementia as well as any organic cause of catatonia. The final diagnosis of frontotemporal dementia with catatonic symptoms was made.

The patient was started on quetiapine due to its sedative property and relatively better tolerability when higher dosages of antipsychotics are needed for managing BPSD slowly increased to 400 mg/day and donepezil 5 mg and memantine 5 mg/day. To manage catatonic symptoms and agitation which were severe enough to put him and his family in danger and was not getting controlled on oral antipsychotics ECT was started after a discussion with the family. Possible benefits and adverse consequences were explained and informed consent for the ECT was obtained from family members. BFCRS score before starting ECT was 13 and reduced to 0 after 6 modified ECTs. The patient also showed improvement in repetitive behavior, agitation, and sleep disturbance. The patient became calm and manageable, although he still required assistance performing ADL. No intramuscular antipsychotics were required to restrain the patient after ECT and he got maintained on oral quetiapine 400 mg/day. The patient was discharged and followed up in OPD after 15 days and after 1 month. Improvement was sustained and family members reported no noticeable further decline in cognitive abilities.

Pharmacological Management for Cognitive Symptoms of Dementia

The approved pharmaceutical treatments for cognitive impairment in Alzheimer’s dementia include cholinesterase inhibitors (donepezil, rivastigmine, and galantamine) and NMDA antagonists (memantine). These drugs slow the disease progression. But there is little proof that they are helpful for other types of dementia.²

Apathy or disinhibition may be present to variable degrees in people with frontotemporal dementia. They might become less interested in self-care, socializing, and other obligations, or they might act in ways that are unsuitable for social situations.²

Pharmacological treatment for dementia’s behavioral and psychological symptoms.

The most prevalent BPSD symptoms in dementia include apathy, depression, agitation, hostility, wandering behavior, disturbed sleep, and psychotic symptoms.

Escitalopram may be able to reduce agitation when it is not severe enough to start antipsychotics, according to recent research.¹ Other selective serotonin reuptake inhibitors (SSRI), including escitalopram, are probably useful in reducing agitation in dementia patients. Further research is necessary to back up this therapy choice, though.

The only medication recognized for the treatment of agitation and violence in dementia patients is risperidone. For severe psychotic symptoms and agitation, low-dose atypical antipsychotics (Aripiprazole, Risperidone, Quetiapine, etc.) may be an option. In dementia caused by Lewy body disease and Parkinson’s disease, quetiapine and clozapine may be considered if there are severe psychotic symptoms.²

Further Somatic Therapies

During the course of illness, many patients with dementia exhibit agitation or violence. Interventions in behavior, environment, and medicine may not
always be effective in treating very serious cases of symptoms that could be fatal. These patients may be candidates for ECT. Challenges may be encountered in diagnosing and treating FTD in presence of other comorbid psychiatric disorders like depression, BPAD.\textsuperscript{3,4} Besides ECT also shows drastic improvement in cases with catatonic symptoms.\textsuperscript{2} There are case reports where the catatonic symptoms could not be distinguished from symptoms of FTD until they were resolved with ECT.\textsuperscript{5} There are systematic reviews of the published literature advocating that ECT is a highly efficacious modality in controlling agitation even in patients with dementia.\textsuperscript{6} Safety issues, ranging from delirium, seizures, stuttering and slurred speech to serious cardiovascular events have been noted in studies. The studies are less and also it remains unclear whether certain events such as spontaneous seizures and confusion could be attributed to treatment or the prevailing condition of the patients.\textsuperscript{7}

\textbf{References}