Behavioral Disturbance From Suspected Frontotemporal Dementia And Comorbid Paraphilia: A Case Report
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Citation

Abstract
Frontotemporal dementia is a degenerative disorder of the frontal and temporal lobes of the brain which results in disturbances in behavior, personality, and language. Onset usually occurs between 45-60 years of age. Presentation occurs as one of three types: a behavioral variant, a semantic dementia, or a progressive non-fluent aphasia. Behavioral disinhibition can be a prominent disturbance of this condition. Paraphilias are a group of recurrent behaviors or stereotypic urges with sexually arousing objects or situations. These activities are aberrant and impulsive and often involve humiliation to oneself or others. Such actions can result in considerable social impairment. This is the only report known to the authors of a patient with suspected comorbid frontotemporal dementia and paraphilia and illustrates a case with a complex differential diagnosis.

INTRODUCTION
Frontotemporal Dementia
Frontotemporal dementia (FTD) comprises approximately 3% of all dementia cases and involves the frontal and temporal lobes of the brain.(1) Onset usually occurs in middle life, and patients survive approximately eight years after diagnosis.(2,3) Previously known as Pick’s Disease for its histopathological neuronal inclusions (“Pick bodies”), FTD is now classified as a distinct illness with three subtypes: a behavioral variant, a semantic dementia, and a progressive nonfluent aphasia.(3) Occurring initially and throughout the illness, personality change and inappropriate social conduct are hallmarks of the disease.(4)

The pathological finding characteristic of frontotemporal dementia is an accumulation of abnormal tau or ubiquitin proteins in the brain.(1) While these proteins normally promote structural support and provide nutrition, a mutation of chromosome 17 leads to abnormal tau or ubiquitin accumulation, gliosis, and formation of interneuronal bodies. This results in frontal and temporal lobe neuronal death which manifests as dementia.(4)

Diagnosis is made following a comprehensive history, physical examination, mental status assessment, neurodiagnostic imaging, and neuropsychological testing.(1) The history may note evidence of impaired insight, interpersonal difficulties, and personality or behavioral alterations, including disinhibition, impulsivity, poor personal hygiene, distractibility, and hyperorality.(4) Compulsive or perseverative behaviors may occur early in the disease and can be mistaken for a psychiatric disorder, and inappropriate sexual behaviors might result in social or legal problems.(4) Speech is often affected and may progress to aspontaneity or echolalia. Significant memory loss occurs only in later stages.(1) Physical examination may reveal incontinence, akinesia, and primitive frontal lobe reflexes.(1) An electroencephalogram (EEG), neuroimaging, and laboratory studies help rule out other causes of dementia.(1) The EEG may reveal non-specific focal background slowing, or it may remain normal despite clinically evident dementia. Neuroimaging might depict selective atrophy or hypoperfusion in frontal or temporal lobes, but this is not specific for FTD.(1,2) Atrophy in an asymptomatic individual cannot predict whether clinical FTD will appear.(2) Conversely, normal neuroimaging does not rule out diagnosis in early stages of disease.(1) In short, imaging can identify pathology, but the diagnosis is made clinically, based on impaired frontal lobe functioning confirmed by neuropsychological testing.(1)

No disease-modifying agents are known to reverse frontotemporal dementia. Behavioral strategies are the first-line interventions for management of behavioral difficulties, and symptom-targeted pharmacotherapy is prescribed if needed.(1) Antidepressant medicines have been administered with mixed results.(5-7) Antipsychotic drugs can decrease disinhibition, while carbamazepine is documented for decreasing hypersexuality.(8-11) Benefits in prescribing
Paraphilias are recurrent, intense, sexually arousing fantasies, urges, or behaviors that occur over a period of at least six months. They involve inanimate objects and suffering or humiliation of oneself, a partner, or non-consenting individual, causing distress or impairment in social or occupational functioning. It is difficult to estimate the prevalence of paraphilias, because few people with paraphilias seek treatment. In a sample of 112 voluntarily admitted male psychiatric inpatients, 13% reported symptoms consistent with at least one lifetime paraphilia, most commonly voyeurism, exhibitionism, or sexual masochism. These subjects had a history of frequent psychiatric hospitalizations, suicide attempts, and childhood sexual abuse. Paraphilias are more common among men with impulse control and post-traumatic stress disorders.

Exhibitionism is a paraphilia in which an individual has experienced sexually arousing fantasies, urges, or behaviors involving exposing genitals to an unsuspecting stranger, causing distress and interpersonal difficulty. The exact etiology is unknown. Psychodynamic explanations propose that exhibitionism develops as a defense mechanism against the anxiety of dependency, with tension reduced by projection. It may also be an attempt to exert sadistic control and avenge perceived past mistreatments.

Coprophilia includes sexual fantasies, urges, or behaviors involving exposing genitals to an unsuspecting stranger, causing distress and interpersonal difficulty. Coprophilia has been linked to early developmental orderliness, parsimony, and obstinacy in those with anal eroticism. There is an association between obsessional neuroses, sadism, and anal eroticism.

Excessive love of cleanliness might be a form of reaction formation against coprophilic tendencies. Before diagnosing paraphilia, clinicians must obtain a detailed history, perform a physical examination, and assess mental status because impairment in judgment, social skills, or impulse control may be found in other conditions, such as mental retardation, dementia, brain injury, delirium, intoxication, mania, or psychosis. Data on successful interventions are limited, since affected individuals rarely engage in treatment. There is no specific therapy for people with paraphilias; current options aim to decrease sexual arousal and hypersexual behavior. Serotonergic agents (e.g., buspirone, antidepressant drugs) may be helpful in reducing inappropriate behaviors, particularly in patients with compulsive activities. Hormone-based therapies (e.g., estrogens, antiandrogens, gonadotropin-releasing hormone agonists) also evidence clinical benefits. Mood stabilizers might help in treating people with paraphilias. Cognitive behavior therapy is recommended as an adjunct to pharmacotherapy, and psychodynamic therapy can reduce coprophilia and associated coprophagia.

### CASE REPORT

Reporting of this case study followed the ethical guidelines of the University of Louisville School of Medicine for reporting case studies. A 59-year-old Caucasian male was brought by police to an emergency room for psychiatric evaluation after urinating and defecating in public areas at a homeless shelter, resulting in aggression towards him by other residents. He acknowledged these behaviors without remorse, stating that “it’s easier to go there instead of the bathroom.” He was admitted to the psychiatric service for evaluation of these behavioral disturbances. His medical history was significant for poorly controlled type 2 diabetes mellitus, diabetic retinopathy and neuropathy, hyperlipidemia, hypertension, peripheral vascular disease, coronary artery disease, two myocardial infarcts, congestive heart failure, and cataracts. There was no history of concussion, meningitis, encephalitis, cerebrovascular accident, seizure, or syncope. His outpatient medications included aspirin, hydrochlorothiazide, triamterene, hydralazine, insulin, metformin, isosorbide, metoprolol, and rosuvastatin. He smoked cigarettes, reported past heavy alcohol consumption for years with sobriety for the past decade, and denied use of illicit substances. There was a family history of diabetes in his father and late-age onset dementia in his mother.

Although no relatives were available for questioning, review of his medical records revealed that he had been verbally abused by his father and sexually assaulted by a classmate at age 12. After graduating from college and serving in the military, he had worked intermittently as a minister and had spent five years in prison for an arson conviction. For the previous 11 years, he was unemployed, divorced, and living in shelters. In the past, he had engaged in masochistic sexual behaviors with women, paying them to hurt him. There was no history of suicide attempts, self-mutilation.
psychiatric medication use, or psychiatric hospitalizations. He first received psychiatric care in 2007 for anxiety and depression and had told his female interviewer that he liked women to wash his hair and had requested that she do the same. At that time, his memory was intact for immediate, recent, and remote events. He was diagnosed with alcohol dependence (in remission) and sexual masochism. He declined all treatment options and did not obtain further mental health care. A month prior to this hospitalization, he had been evaluated in an emergency room for public nudity and inappropriate defecation. He did not meet criteria for inpatient admission, so he was discharged and referred for neuropsychological assessment. This was requested to evaluate his decision-making capacity and to identify possible cognitive or organic explanations for his behavior. During testing, the patient’s social worker reported that the patient defecated and urinated in public areas and in his clothing, and he would decline hygiene items offered by shelter staff. The patient responded that he “didn’t care” and “was used to it.” He expressed awareness of others’ negative reactions to his behavior, but he did not appear to be concerned by it or motivated to change it. He was reported to periodically compulsively touch signs, phones, and wires with his right hand and elbow. In the interview, his speech was fluent with normal prosody, tone and volume, without dysarthria or word-finding difficulty. Specific scores from the neuropsychological tests are unavailable, but the summary stated that his intellectual functioning was below average but not in the range of mental retardation. He had difficulty understanding and following directions, and he displayed impaired executive functioning through poorly organized work, lack of integration, and perseverative tendencies. He possessed impaired perceptual and spatial reasoning, verbal and visual memory, and slowed information processing. He demonstrated well-preserved verbal comprehension abilities on tasks that were overlearned or when prompted with verbal cues. However, thought processes became tangential and disorganized on free recall tasks despite good effort, and he was unable to initiate behaviors. His behavior was apathetic and avolitional, consistent with and attributed to frontal lobe impairment. The evaluator felt that disorganization and social confusion (and conflict) occurred when he trusted flawed reasoning instead of utilizing external feedback from his environment. Testing concluded with diagnoses of frontotemporal dementia and schizotypal personality traits. Physical examination was unremarkable except for plantar neuropathic pain. A complete blood count, serum chemistry, thyroid tests, and cyanocobalamin and folate levels were within normal limits, except for a serum glucose > 600 mg/dl. Urinalysis evidenced glucosuria. A rapid plasma reagin test was nonreactive, which excluded neurosyphilis. Except for glucosuria, a urinalysis, urine culture, and overflow incontinence evaluation were unremarkable when he was evaluated two months earlier for hyperglycemia and “fecal/urinary incontinence in public;” bowel function studies were not initiated. A review of computerized tomography brain scans completed in 2009 and 2010 were notable for mild anterior temporal atrophy and minor, old, small-vessel ischemic changes in the right frontal lobe.

On mental status examination, the patient appeared his stated age and was cooperative and jovial with good eye contact. He was disheveled, wearing clothing soiled with feces and urine. Speech was pressured but cheerful in tone and with normal rate and volume. Mood was euthymic and affect was bright. He denied suicidal and homicidal ideation. His knowledge base appeared average. Thought processes were logical but circumstantial at times. Thought content was devoid of hallucinations, delusions, or paranoia. He was alert, awake, and fully oriented. Memory was intact to immediate and remote events, but he displayed mild impairment in recalling recent events. Insight, judgment, and reliability of information were impaired. Following admission to the psychiatric service, his interactions with peers and staff were initially appropriate. Later, he began to smear feces on walls, walk about naked, wear soiled clothes, and ask female personnel to bathe and change him. He denied being sexually aroused and minimized these incidents without evident embarrassment, remorse, or concern for his inappropriate behavior towards staff. He scored 21/30 on the Folstein Mini Mental State Exam and 25/30 on the St. Louis University Mental Status examination, scores consistent with mild cognitive impairment. These inappropriate behaviors could be attributed to disinhibition characteristic of frontotemporal dementia, or to impaired judgment and cognition observed in some cases of hyperglycemia.(28) Given his medical history, it was also possible that he had another dementing illness secondary to vascular or ischemic changes. In addition, his history of sexual trauma and documented inappropriate interpersonal behaviors in the absence of cognitive change suggested that personality pathology and/or an impulse control disorder (i.e., paraphilia) could explain this presentation. A diagnostic question of frontotemporal dementia (FTD) and paraphilia (exhibitionism and
coprophilia) was raised, so medicine and neurology consultations were requested to assist with diagnosis and management. The medicine consultant advised a diabetic diet, sliding scale insulin, and oral hypoglycemic agents, which resulted in improved glycemic control. Given the results of the neuropsychological testing one month earlier, the absence of different behaviors since that time, and the lack of new, focal neurological deficits on physical exam, the consulting neurologist felt that repeated brain imaging (last scan having been done within the previous year) was not sufficiently warranted. A diagnosis of vascular dementia could not be given based on old imaging and the patient’s intact memory at the time of imaging. The patient admitted to the neurologist that after being hospitalized for a few days, he realized that some of his behaviors at the shelter had been socially unacceptable. In addition to displaying inaccuracies on the clock drawing test and minor apraxia, the patient’s score on the Folstein Mini-Mental Status Exam was consistent with cognitive impairment. The neurological assessment rendered a diagnosis of frontotemporal dementia compounded by hyperglycemia. Other than medication for improved glycemic control, the neurology team recommended no other pharmacotherapy for his diagnoses. It did recommend behavioral management by nursing staff, with limit setting and scheduled toileting every two hours.

Despite these interventions, inappropriate behaviors continued. The patient asked that his restroom be cleaned because he did not “make it to the toilet on time.” Without apparent remorse, he had smeared feces over the floor and walls. When encouraged to shower, he declined and insisted on wearing soiled clothes. He refused clean clothes and urinated on them instead. During a meeting to discuss the incident, the patient minimized these behaviors, smiling and stating, “It didn’t affect anyone else.” When told that he would be observed by staff and would be requested to clean himself after these “accidents,” the frequency of these behaviors subsequently diminished. However, he did once stand at the nursing station fully exposed, and when requested to pull his pants up, he told female staff that he did not know how to fasten his pants and needed help. He expressed no remorse or embarrassment regarding these behaviors, but instead appeared to derive pleasure or entertainment from exposing himself, handling feces, and requesting that female staff provide assistance with bathing, changing, and grooming. His capacity to void appropriately and to wear clean clothes when aware that he was being monitored indicated that his behavior had a volitional component and was not due solely to cognitive dysfunction.

**DISCUSSION**

This may be the first report of a person with suspected frontotemporal dementia and comorbid paraphilia. The patient had a history of socially inappropriate behaviors of uncertain etiology. It is unlikely that his behavior can be solely attributed to frontotemporal dementia or paraphilia, and it is difficult to know whether his social indifference and apathy were due to dementia or schizotypal personality traits. It is plausible that he derived some pleasure from these events, but restrained his expressions when questioned by staff members. Memory is often impaired in individuals with metabolic disturbances such as hyperglycemia, and individuals with FTD are noted to excessively prefer sweet foods.(28,29) Frequent and inappropriate urination may be attributed to a combination of brain pathology, diabetic hyperglycemic hyperosmolarity, and diuretic use. Many of the patients’ abnormal behaviors and social impairments can be observed in cases of FTD, paraphilia, and vascular or mixed dementias affecting the frontal lobes. However, he had received a diagnosis of paraphilia several years earlier in the absence of cognitive changes. Despite toileting schedules in the hospital, he continued to display inappropriate behaviors with apparent volitional control. On neuropsychological testing and memory screening examinations, he exhibited cognitive impairment. It is possible that the patient had pre-existing personality pathology with paraphilias (exhibitionism and coprophilia); perhaps these became more overt through the disinhibition of frontotemporal dementia and cognitive impairment secondary to hyperglycemia. In addition to a pre-existing paraphilia, it is possible that his personality changes and disinhibition were caused not by FTD, but rather by a recent-onset vascular dementia afflicting primarily frontal lobes and additional cognitive impairment from hyperglycemia. Additional neuroimaging could have provided evidence of increased brain atrophy, ischemic changes, or other pathology. A single and/or definitive diagnosis in this case is not determinable, but this case illustrates a complex differential diagnosis. Clinicians should consider the presence of paraphilia and/or FTD when evaluating patients with socially inappropriate or impulsively dyscontrolled behavior.

**References**


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