The Unique Predisposition to Criminal Violations in Frontotemporal Dementia

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Brain disorders can lead to criminal violations. Patients with frontotemporal dementia (FTD) are particularly prone to sociopathic behavior while retaining knowledge of their acts and of moral and conventional rules. This report describes four FTD patients who committed criminal violations in the presence of clear consciousness and sufficiently intact cognition. They understood the nature of their acts and the potential consequences, but did not feel sufficiently concerned to be deterred. FTD involves a unique pathologic combination affecting the ventromedial prefrontal cortex, with altered moral feelings, right anterior temporal loss of emotional empathy, and orbitofrontal changes with disinhibited, compulsive behavior. These case histories and the literature indicate that those with right temporal FTD retain the capacity to tell right from wrong but have the slow and insidious loss of the capacity for moral rationality. Patients with early FTD present a challenge to the criminal justice system to consider alterations in moral cognition before ascribing criminal responsibility.

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Epidemiological data and clinical information indicate a relationship between criminal behavior and brain disorders. As many as 94 percent of homicide offenders, 61 percent of habitually aggressive persons, and 78 percent of sex offenders may have brain dysfunction. Acquired sociopathy, or antisocial acts with disturbances in the moral emotions linked to the interests or welfare of others, occurs in those with brain lesions affecting the inner or ventromedial prefrontal cortex (vmPFC). Investigations show that lesions in the vmPFC impair moral judgment, ²⁻⁴ and early-life lesions impair the development of moral decision-making. 5,6 Other factors that may contribute to impaired moral cognition or to the mental processes that underlie morality include loss of empathy or sympathy and disinhibited, compulsive behavior.

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Frontotemporal dementia (FTD) is a progressive neurodegenerative disorder previously known as Pick's disease. It affects the frontal and anterior temporal regions, especially the vmPFC, orbitofrontal cortex, and anterior temporal regions. 8,9 On average, FTD has an age of onset in the late 50s, with an equal incidence among men and women and potential autosomal dominant inheritance.^{7,8} Although the disorder is termed dementia, early in the course most patients have a personality change with relatively intact cognition (i.e., early FTD is less an impairment in memory, language, or perception than a disorder of abnormal behavior). Subgroups of FTD patients can develop primary progressive aphasia, semantic deficits, parkinsonism evolving to progressive supranuclear palsy (PSP), corticobasal degeneration, or motor wasting and motor neuron disease (MND).8 The core features of the usual behavioral variant FTD are transgression of social norms including sociopathic behavior, a loss of empathy or appreciation of the feelings of others, and disinhibited, compulsive acts. Patients with FTD can commit criminal violations while retaining the ability to know moral rules and conventions. 10

Among brain disorders, sociopathy is particularly associated with FTD, much more so than with Alzheimer's disease (AD), vascular dementia, or other neurodegenerative disorders, with the possible exception of Huntington's disease.⁷ These patients pose a potential dilemma for the law. Currently, the paraphrased M'Naughten standard for not guilty by reason of insanity requires that the perpetrator be incapable, by reason of mental illness, of understanding the nature of the criminal act or of knowing that the act was wrong. 11 In this report, we examine four FTD patients with sociopathy from our dementia research databases. The patients gave consent to be enrolled in these databases for the de-identified use of their clinical information. We examined their sociopathic behavior and their mental state at the time of the acts. Did they commit prohibitive acts in a culpable mental state? (Access to the deidentified data set was approved by the University of California Los Angeles Institutional Review Board.)

Case Reports

Patient I

A left-handed male in his sixties began stalking and attempting to molest children for the first time in his life. ¹² He followed children home from school and tried to touch them. On one occasion, he put his arm around a young boy and then struck him when he tried to pull away. On another occasion, he stood at the foot of a pool and stared at the children for a prolonged time. When he exposed himself to his neighbor's children, he was arrested. The patient did not deny his actions, could describe them in detail, and endorsed them as wrong and harmful. Despite this, he stated that he did not feel that he was causing harm at the time of his acts.

The patient's personality had deteriorated over the prior four years, with decreased concern for others, disinhibition, and compulsive hoarding. He had caused disturbances at work, such as intruding into others' conversations and walking into others' offices. He was taking supplies into his office, constantly pilfering and taking samples, and hiding money. He compulsively took photographs of the sunset every night. In restaurants, he filled his pockets with sugar, napkins, and other items. In addition, he ate indiscriminately, even going through waste containers and eating garbage. He stopped showering and wore the same clothes every day. The family

history was positive for an unspecified dementia in his mother.

On examination, his thought processes were linear, and he did not endorse hallucinations, delusions, or paranoia. His Mini-Mental State Examination (MMSE) score was 29/30.13 He was oriented to place and time, and his basic attention span was normal. Language examination revealed verbal stereotypes and decreased naming, but normal comprehension and repetition. Memory was slightly impaired, but visuospatial skills were normal. Abstractions were concrete, and he had perseverations and impaired set-shifting. Neuropsychological tests confirmed the presence of mild decreased memory and declines in naming and executive functions. The neurological examination disclosed normal cranial nerve, coordination, motor, sensory, and reflex testing. Magnetic resonance imaging (MRI) of the brain revealed no abnormalities. Positron emission tomography (PET) imaging showed decreased metabolism in the right anterior temporal lobe.

The patient met consensus clinical criteria for FTD. Despite cognitive deficits, he had sufficient cognition to recall and understand his behavior and its wrongfulness. His lack of empathy and disinhibited compulsive tendency appeared to drive his pedophilic behavior. He was started on paroxetine 20 mg, divalproex 500/750 mg, and conjugated estrogens 0.625 mg. With this medical regimen and increased supervision, the patient had significant behavioral improvement. His case was not prosecuted.

Patient 2

A right-handed woman in her fifties presented with an 18-month progressive personality change accompanied by petty theft at retail establishments. She stole merchandise and would go behind counters and take items without concern for payment, even when she personally knew the merchants. Only the intervention of her family prevented the merchants from pursuing legal action and prosecution. Her family also described her as becoming disinhibited, with a tendency to talk to strangers. When her specific behaviors were pointed out to her, she could describe them in detail, and she endorsed knowing that they were wrong. When asked why she engaged in such behavior, she would shrug and say, "That's me."

There were other behavioral changes. The patient had become disinhibited, with excessively personal or familiar comments about others. She frequently made puns and burst into laugher. Yet, her concern for others was generally decreased. For example, when asked about the recent death of a close relative, she verbally expressed sadness and then quickly lapsed into laughter and light-hearted responsiveness. There was a compulsive tendency, particularly with regard to money, which she would hoard and hide in different places. She developed an addiction to ice cream and gained a considerable amount of weight. She had decreased personal hygiene with increased sloppiness in dress, often wearing the same clothes repeatedly. In her family history, her grandmother had dementia in her sixties and her brother had motor neuron disease (MND) and died in his fifties.

On examination, her spontaneous verbal output had stereotypical phrases and many intrusion of laughter. Her MMSE score was 23/30, primarily because of naming or language difficulty. She had decreased confrontational naming, but her auditory comprehension was preserved at the sentence level. On an auditory verbal learning task of memory, she had a memory retrieval deficit. Her visuospatial constructions were normal, but her interpretation of proverbs was concrete. The findings in the remainder of the examination were normal, including cranial nerves, coordination, motor testing, reflexes, and sensory tests. Her MRI was unremarkable, but singlephoton emission tomography (SPECT) imaging showed hypoperfusion in both anterior temporal lobes, more right than left.

The patient was diagnosed with FTD on a familial, autosomal dominant basis and was treated with sertraline for compulsive-type behavior. The patient was observed for two years and showed worsening language, executive functions, and semantic deficits.

Patient 3

A man in his fifth decade of life was detained after grabbing a woman's buttocks. On other occasions, he had repeatedly made lewd comments, such as describing a woman's appearance when naked. He was also found to have condoms and sildenafil stashed at work. On being confronted, he recalled his egregious behaviors in detail, described them as inappropriate, and understood why he got into significant trouble because of them. Additional legal action ensued when he invaded his neighbor's house looking for undelivered mail. These behaviors were totally un-

characteristic of the patient and constituted a personality change. When asked why he had engaged in the actions, he described an inability to restrain himself at the time.

He had an insidiously progressive personality change for about a year, with decreased empathy or concern for his victims. He made excessive and inappropriate jokes at work and had multiple driving errors. He had a tendency toward impulsive acts and repetitive behavior, including multiple trips for coffee or to the market. He developed a tendency to eat sweets every day, which was uncharacteristic of him, and he became sloppy in his dress, soiling the front of his clothes while eating. Coincident with these changes, he had decreased agility, decreased gait, and a decline in his fine hand coordination and handwriting.

On examination, he was alert and attentive, with an MMSE score of 27/30. Language was fluent, and auditory comprehension and naming were intact. On an auditory verbal learning task, memory was normal except for a mild retrieval deficit. He did not have ideomotor apraxia. His visuospatial skills were intact, and he was sufficiently abstract on proverb interpretations. On the rest of his neurologic examination, he walked *en bloc*, with decreased associative movements. Extraocular movements showed decreased voluntary saccades in the upward and downward directions. The lower cranial nerves suggested some masking of his face, but there was no pseudobulbar palsy. He had increased tone, particularly in the axial plane, but also to a degree in his arms. There were no reflex or sensory changes. A computed tomographic scan showed no lesions, but a SPECT scan showed frontal and temporal hypometabolism.

This patient had an FTD spectrum disorder with early PSP, a not infrequent combination. ¹⁴ He was started on physical and occupational therapy and was prescribed coenzyme Q and memantine, with future consideration for antiparkinson therapy. The initial behavioral management focused on education and behavioral intervention rather than psychoactive medications. Legal action was concluded without incarceration.

Patient 4

A right-handed man in his early fifties had a hitand-run accident and left the scene without concern. He had struck a van with passengers but kept driving. The police stopped him a short distance away from the scene, and he did not deny his action. Leaving the scene of an accident was not characteristic of his premorbid personality, yet he had had several recent traffic violations. There was no evidence that the patient had had a seizure or any alteration of awareness during the accident. He could recall and describe the accident, knew that it was wrong to leave the scene, but did not feel the need to stop at the time.

Over the prior two years, the patient's pervasive behavior had significantly changed. He had become disengaged and emotionally detached; for example, he did not react to the death of his mother and he did not visit his wife during her hospitalization for blood clots. He had periods of laughing inappropriately and childlike excitement. His wife described him as having no restraint in what he said, with a tendency to blurt out distressing comments. He was no longer embarrassed over passing gas or belching in public or appearing partially clothed in front of others. The patient had a tendency toward hyperorality, especially for peanuts, and had a decline in personal hygiene. Other aspects of the history included dysarthria and a recent tendency to choke on liquids. His mother had died of AD at 84; otherwise, there were no known familial neurologic conditions.

On examination, he had evidence of MND. He had dysarthric speech and upper-extremity fasciculations. On mental status assessment, he was fully attentive, with an MMSE score of 27/30. His language was normal, and his memory was intact on an auditory verbal learning task. His visuospatial constructions were normal, and his proverb interpretation was abstract. The rest of his neurologic examination showed intact cranial nerves except for the dysarthria. The results of gait, coordination, reflex, and motor testing were normal except for the fasciculations. He had normal MRI and PET scans early in his course, but there was denervation on an upper-extremity electromyogram.

The patient's diagnosis was FTD-MND, which occurs in 1 to 15 percent of FTD patients. Although the early PET was normal, follow-up revealed deteriorations of personality and cognition characteristic of FTD. Most of his course involved progression of MND and management of his worsening dysarthria, dysphagia, and motor weakness. He received riluzole 50 mg every 12 hours, among other medications, but continued to deteriorate and eventually died about one year after his accident, precluding further legal consequences.

Discussion

These FTD patients illustrated the problem of sociopathic behavior from frontal brain disorders. They developed pedophilia and committed theft, sexual harassment, and automobile violations. All four had an awareness of their behavior at the time of the acts and understood that it was wrong. They had preserved knowledge of moral behavior and of potential consequences, but they went ahead anyway, in an unempathic, impulsive, and often compulsive, manner. In addition to manifesting the behavioral features of FTD, these patients had the spectrum of FTD-related conditions, including semantic deficits (Patient 2), an autosomal dominant inheritance (Patient 2), PSP (Patient 3), and MND (Patient 4).

The early diagnosis of FTD can be difficult, particularly in court. The clinical diagnosis of this disorder is based on the core behavioral criteria of an insidious and progressive personality change with impairments in social interpersonal conduct, impairments in regulation of personal conduct, early emotional blunting, and early loss of insight.8 There is no definitive test for FTD, and neuroimaging, which may show abnormalities in frontotemporal regions, is only supportive and not diagnostic. Hence, it is absolutely essential to document clinical changes in individuals by obtaining similar confirmatory information from third parties in the individual's environment. Ultimately, only long-term clinical follow-up and documentation of clinical progression to cognitive impairments and dementia can establish the diagnosis of FTD.

Patients with usual behavioral variant FTD manifest inappropriate social behavior early in the disease, when the neurodegeneration is still localized or asymmetrical, and their general cognitive function is relatively intact.8 Most commonly, there is a loss of social tact and propriety, improper verbal or nonverbal communication, and unacceptable physical contact. Socially inappropriate behavior expands to encompass a failure to conform to lawful behavior in greater than one-half of patients with FTD.^{3,15} Among these patients, investigators have reported stealing (shoplifting, stealing food), unethical job conduct, indecent exposure, inappropriate sexual comments or behavior, illegal driving acts, and physical assaults or violence. 12,15-17 In one study, 16 (57%) of the FTD patients had had sociopathic behavior compared with only 2 (27%) of the AD patients.³ The FTD patients with sociopathic acts were aware of their behavior and knew that it was wrong but did not prevent themselves from acting.³ They lacked premeditation and claimed subsequent remorse, but did not act on it or express concern for the consequences.

The behavior of FTD patients is reminiscent of the famous case of Phineas Gage who sustained bilateral vmPFC injury from an explosion that propelled an iron rod through his brain, except that, in FTD, the behavioral changes are gradual and insidious in onset.^{17,18} Acquired sociopathy occurs from focal vmPFC lesions, 4,19,20 and, although poorly visualized on neuroimaging, the neuropathology of early FTD includes the vmPFC. 9,21 Patients with vmPFC lesions have diminished emotional experience with reduced sociomoral emotions, such as compassion, shame, guilt, and regret. 4,5,22-25 The vmPFC, with its rich interconnections with limbic structures, mediates these strong, automatic, negative "gut reactions" to moral violations that prevent individuals from implementing morally impermissible actions.²⁶⁻³⁰ In a unique study, FTD patients were more impaired in their ability to respond immediately to emotionally based moral (personal) vignettes than were AD patients and normal control subjects. 26,27,31 Yet, those with vmPFC lesions are aware of their actions, have preserved logical reasoning and knowledge of social and moral norms, and can anticipate outcomes. 6,10,32

In addition to vmPFC involvement, early FTD involves other brain areas that affect the occurrence of sociopathic behavior. In FTD, there is decreased emotional empathy, particularly associated with right anterior temporal disease, as in at least three of our patients. 33,34 This variant of FTD is particularly prone to interpersonal coldness and a lack of responsiveness to others' distress. 34,35 In other studies of FTD patients, decreased emotional empathy and reduced responsiveness to victims correlates with damage to the right ventromedial-anterior temporal network. 35-38 Finally, the lack of strong moral emotions in FTD and the loss of empathy cannot override drives, possibly released by orbitofrontal dysfunction, for disinhibition, compulsions, or behavioral tendencies, such as pedophilia. 7,39,40 In sum, the unique neuropathological involvement in FTD, particularly with right anterior involvement,

makes these patients susceptible to committing sociopathic acts.

Do FTD patients have culpable mental states (mens rea) at the time of their acts? Are they responsible agents? FTD patients with sociopathy would not pass most legal criteria for judgments of not guilty by reason of insanity. On the basis of a restrictive M'Naughten rule, the U.S. Congress passed the Comprehensive Crime Control Act in 1984, which requires an insanity defense to establish, by "clear and convincing evidence," that "at the time of the commission of the acts constituting the offense, the defendant, as a result of a severe mental disease or defect, was unable to appreciate the nature and quality or the wrongfulness of his acts" (18 U.S.C. § 17). 41 Under these guidelines, FTD patients would not qualify for not guilty by reason of insanity, because their disease did not cause a "defect of reason." They did not have a general decreased capacity for rationality nor would they be exonerated because of an internal coercion or irresistible impulse. Nevertheless, they have a specific, brain-based impairment in moral reasoning. Anglo-American jurisprudence distinguishes between reason-based law and a natural law based on what a reasonable person would do in a like circumstance. 11 Arguably, under the law, a reasonable person is someone whose impulses are restrained by intact moral cognition (i.e., moral rationality). 42 Without the normal internal restraint of intuitive moral emotions and empathy, FTD patients may not possess the faculties of a reasonable person sufficient to bring reason to bear on their drives and to abstain from criminal violations. These considerations deserve a reappraisal of how we view criminal violations among brain-injured patients and how we can incorporate neurological factors involved in moral capacity or moral cognition.⁴²

In conclusion, in FTD, sociopathic behavior is consistent with decreased emotional moral judgments plus a lack of empathy and disinhibited, compulsive drives consequent to the unique neuropathology of this disorder. FTD patients have impaired moral rationality from impaired moral cognition. These findings have implications for understanding brain-damaged patients and the law.

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