Right Orbitofrontal Tumor With Pedophilia Symptom and Constructional Apraxia Sign

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Background: Orbitofrontal abnormalities are associated with poor impulse control, altered sexual behavior, and sociopathy.

Objective: To describe a patient with acquired pedophilia and a right orbitofrontal tumor who was unable to inhibit sexual urges despite preserved moral knowledge.

Design: Case report.

Results: The patient displayed impulsive sexual behavior with pedophilia, marked constructional apraxia, and agraphia. The behavioral symptoms and constructional deficits, including agraphia, resolved following tumor resection.

Conclusions: For patients with acquired sociopathy and paraphilia, an orbitofrontal localization requires consideration. This case further illustrates that constructional apraxia can arise from right prefrontal lobe dysfunction. Agraphia may represent a manifestation of constructional apraxia in the absence of aphasia and ideomotor apraxia.

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The orbitofrontal cortex contributes to moral-knowledge acquisition and social integration.1,2 Adult-acquired orbitofrontal damage may diminish impulse control and can be associated with sociopathic behavior.3-5 We describe a 40-year-old man who was treated with medroxyprogesterone acetate and a 12-step program for new-onset pedophilia. He was subsequently diagnosed as having a right orbitofrontal tumor. At the time of tumor discovery, his neurologic examination results were notable for a paucity of sensorimotor signs, marked constructional apraxia, and agraphia.

REPORT OF A CASE

A 40-year-old, right-handed man in an otherwise normal state of health developed an increasing interest in pornography, including child pornography. He had a preexisting strong interest in pornography dating back to adolescence, although he denied a previous attraction to children and had never experienced related social or marital problems as a consequence. Throughout the year 2000, he acquired an expanding collection of pornographic magazines and increasingly frequented Internet pornography sites. Much of this prurient material emphasized children and adolescents and was specifically targeted to purveyors of child pornography. He also solicited prostitution at “massage parlors,” which he had not previously done.

The patient went to great lengths to conceal his activities because he felt that they were unacceptable. However, he continued to act on his sexual impulses, stating that “the pleasure principle overrode” his urge restraint. He began making subtle sexual advances toward his prepubescent stepdaughter, which he was able to conceal from his wife for several weeks. Only after the stepdaughter informed the wife of the patient’s behavior did she discover with further investigation his emerging preoccupation with pornography, and child pornography in particular. The patient was legally removed from the home, diagnosed as having pedophilia, and prescribed medroxyprogesterone. He was found guilty of child molestation and was ordered by a judge to either undergo inpatient rehabilitation in a 12-step program for sexual addiction or go to jail. Despite his strong desire to avoid prison, he could not restrain himself from soliciting sexual favors from staff and other clients at the rehabilitation center and was expelled. The evening before his prison sentencing, he came to the University of Virginia Hospital (Charlottesville) emergency department complaining of a headache. A nonphysiologic cause was suspected, and the psychiatry service admitted him with...
Figure 1. Magnetic resonance imaging scans at the time of initial neurologic evaluation: T1 sagittal (A), contrast-enhanced coronal (B), and contrast-enhanced axial (C) views. In A and B, the tumor mass extends superiorly from the olfactory groove, displacing the right orbitofrontal cortex and distorting the dorsolateral prefrontal cortex. The tumor is capped by a large cystic portion.

A diagnosis of pedophilia, not otherwise specified, after he expressed suicidal ideation and a fear that he would rape his landlady. The day after his admission he complained of balance problems, and a neurologic consultation was obtained.

The patient's medical history was notable for a closed head injury 16 years earlier that was associated with a 2-minute loss of consciousness and no apparent neurological sequela, a 2-year history of migraines, and hypertension. He was without a previous psychiatric or developmental history and had exhibited no prior deviant sexual behavior. Medications included fluoxetine hydrochloride, amlodipine besylate, metoclopramide hydrochloride (for nausea), and medroxyprogesterone acetate at a dose of 10 mg/d. There was no family history of psychiatric disease. He had worked as a corrections officer prior to completing a master's degree in education in 1998, at which time he became a schoolteacher. He was currently in his second marriage, which prior to his developing sexual preoccupations had been stable for 2 years.

During a neurologic examination, he solicited female team members for sexual favors. He was unconcerned that he had urinated on himself. He was slow to initiate leftward saccades and had mild left nasolabial fold flattening without facial weakness. Appendicular tone was increased bilaterally. There was no neglect. Abnormal glabellar, snout, and palmonental responses were present. The patient's gait was wide based, and as he walked, his step length diminished and side-to-side titubation occurred.

Magnetic resonance imaging revealed an enhancing anterior fossa skull base mass that displaced the right orbitofrontal lobe (Figure 1). Prior to resection (December 2000), bedside neurologic testing found the patient alert and completely oriented. He scored 25 of 30 on the Folstein Mini-Mental State Examination,6 missing points for delayed recall, impaired copy (Figure 2A), and an inability to write a legible sentence (Figure 2B). His memory, however, was intact according to a 16-item test of enhanced cued recall on which he freely retrieved 6 objects and the remaining 10 with cues. He named the previous 5 presidents. He was able to state digit spans of 7 going forward and 4 in reverse. On the clock-drawing test, he exhibited marked constructional apraxia, and this did not improve with the figure copy test (Figure 2A). Simultanagnosia was absent. Although spontaneous language output, repetition, comprehension, and reading skills were intact, his writing was illegible (Figure 2B). The patient was able to spell, and prosody was normal. During 1-minute intervals he named 5, 7, and 5 words beginning with C, F, and L, respectively (bottom of first percentile). He named 11 animals during 1 minute. He verbally shifted between letter and number sets, conceptualized, performed sequential hand movements, and inhibited motor responses on the Luria go–no go test.7 He was without ideomotor apraxia. Results of olfactory testing appeared normal because the patient correctly identified peanut butter and coffee by scent. He performed normally on a task of visuoperception (Luria figure-ground analysis8).

Histopathologic examination revealed a hemangio-pericytoma. Several days after tumor resection, the patient's walking and bladder control improved. He successfully participated in a Sexaholics Anonymous program. Seven months later, he was believed not to pose a threat to his stepdaughter and returned home. In October 2001, he developed a persistent headache and began secretly collecting pornography again. Magnetic resonance imaging showed tumor regrowth, and re-resection was accomplished in February 2002.

Two days after this surgery, his examination results were notable only for a slightly decreased left nasolabial fold. His Mini-Mental State Examination score was 30 of 30. Results of clock-drawing and figure copy tests were normal (Figure 2C), and his writing was legible (Figure 2D). During 1-minute intervals he named 18, 13, and 9 words beginning with C, F, and L, respectively (51st percentile). He named 26 animals during 1 minute and a digit span of 8 going forward and 5 in reverse.

The orbitofrontal cortex is involved in the regulation of social behavior. Lesions acquired very early in life impede social- and moral-knowledge acquisition, which may result in poor judgment, reduced impulse control, and sociopathy.2 A similar acquired sociopathy occurs with adult-onset damage, but previously established moral develop-
ment is preserved. Nevertheless, poor impulse regulation leads to bad judgment and sociopathic behavior. Our patient developed paraphilia late in his fourth decade and met the criteria for pedophilia according to the Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition. His symptoms resolved with the excision of a right orbitofrontal hemangiopericytoma, further establishing causality. The orbitofrontal disruption likely exacerbated a pre-existing interest in pornography, manifesting as sexual deviancy and pedophilia. To our knowledge, this is the first description of pedophilia as a specific manifestation of orbitofrontal syndrome.

Bedside orbitofrontal lobe assessments have low sensitivity. Anosmia is occasionally noted but was not present in our patient. Urinary incontinence, gait ataxia, frontal release signs, and word generation impairment (especially on controlled oral word association) are consistent with general prefrontal lesion localization. Severe constructional apraxia on both free-drawing and copy-drawing tests was an unexpected examination finding that is most often attributable to parietal dysfunction. Absent simultanagnosia and normal visuoperceptual performance on Luria figure-ground analysis suggest relatively intact parietal visuospatial function. Constructional apraxia likely resulted from an inability to execute the drawing task rather than a parietal-based visuospatial failure.

Constructional apraxia is classically associated with parieto-occipital damage and represents a functional consequence of visuospatial dysfunction. It has also been reported to occur with frontal lesions. Constructional apraxia in this patient likely arose from dysfunction of the dorsolateral prefrontal cortex or its connections, although precise localization is difficult given the tumor’s size and mass effect. Regardless, the patient’s intact memory retrieval, working memory, set shifting, and sequencing abilities indicate that dorsolateral prefrontal dysfunction was not pervasive. We do not know if constructional apraxia would have manifested from a similar dominant-sided lesion. Interestingly, frontal degeneration syndromes are associated with early decline of the orbitofrontal lobes and early preservation of drawing abilities. Our findings emphasize that extensive right orbitofrontal damage can produce constructional apraxia.
Our patient exhibited severe agraphia that resolved with resection of his anterior fossa tumor. Although agraphia is typically a disorder of language associated with dominant inferior parietal lobe abnormalities, it can be associated with visuospatial deficits, limb apraxia, and sensorimotor deficits. His agraphia is notable given the absence of limb apraxia, aphasias, and significant sensorimotor deficits. It likely represents a distinct manifestation of his overall constructional apraxia. Demonstrating a preservation of typing ability could have corroborated this hypothesis, but unfortunately this was not attempted prior to his tumor resection. Occasionally, agraphia has been reported with prefrontal lesions, although the mechanism for such deficits is unclear. Agraphia resulting from constructional apraxia is perhaps best considered pseudodysgraphia.

Orbitofrontal lesion research suggests that sociopathic behavior results from a loss of impulse control rather than a loss of moral knowledge. Functional magnetic resonance imaging studies indicate that orbitofrontal, dorsolateral prefrontal, and subcortical limbic structures are involved in behavioral self-regulation and response inhibition, including the conscious regulation of sexual urges. Our patient could not refrain from acting on his pedophilia despite the awareness that this behavior was inappropriate. The somatic marker hypothesis attempts to provide a physiologic explanation for this phenomenon. The orbitofrontal cortex receives afferents from the sensory cortex, amygdala, and hippocampus. It is in turn projects to brainstem autonomic nuclei. Therefore, the orbitofrontal lobes play a role in generating the autonomic responses that typify a variety of emotions. The cortex subsequently attaches a feeling, or somatic marker, to the emotional response; this higher-order interpretation guides behavioral response patterns to environmental stimuli. Disruption of this system can result in decision making that emphasizes immediate reward rather than long-term gain, impairing the subject’s ability to appropriately navigate social situations.

Because prompt surgical intervention was clinically indicated, the neuropsychological evaluation was limited to the bedside. Although a fairly comprehensive assessment of the patient’s cognitive strengths and weaknesses was accomplished, formal neuropsychological testing might have allowed for a finer localization of relevant signs and symptoms. It is also possible that formal neuropsychological testing would have facilitated an earlier diagnosis. Tests that emphasize frontal lobe functions, such as the Stroop Interference Test and Wisconsin Card Sorting Test, are sensitive indicators of frontal lobe dysfunction. It is unfortunate that data from such testing could not be obtained. In addition to these instruments, neuropsychological testing that is both sensitive and specific for orbitofrontal dysfunction has recently been developed. The Iowa Gambling Task requires the subject to select cards from 4 decks, and each card selected incurs either a financial gain or financial loss. Cards from 2 of the decks will occasionally result in a substantial payoff, but choosing from these decks ultimately results in a net loss. The other 2 decks are characterized by more conservative payoffs and penalties. Playing these decks results in a net financial gain. This paradigm can distinguish individuals with orbitofrontal dysfunction from control individuals because it is difficult for orbitofrontal-damaged subjects to restrain their exploration of the riskier, disadvantageous decks.

In summary, signs of orbitofrontal lobe dysfunction are often subtle. Physicians can overlook even large orbitofrontal lesions in patients with acquired sociopathy if not appropriately vigilant. Acquired sociopathy with concomitant constructional apraxia and pseudodysgraphia but not simultanagnosia could indicate the presence of right orbitofrontal dysfunction.

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