



# Genital self-mutilation in a patient with frontotemporal dementia

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**ABSTRACT** - A case of a patient with frontotemporal dementia (FTD) who mutilated his penis is presented. Brain magnetic resonance imaging, single-photon emission computed tomography abnormalities and cognitive assessment are described. Genital self-mutilation (GSM) is a disturbing though yet not described symptom in FTD. A 60-year-old patient presented with a two-year history of social withdrawal and loss of attention to household responsibilities. He was hospitalized and eventually diagnosed with FTD. One day, the nurses observed that his penis was swollen and lacerated. It was discovered that the patient had mutilated his penis with a spoon. He did not have any insight and he completely denied his act. FTD is the second most common cause of dementia in people younger than 65 years. It is characterized by personality changes and impaired social conduct and represents the behavioral variant of the three clinical presentations of frontotemporal lobar degeneration. Sociopathy is a well described but underappreciated phenomenon in FTD patients. It is characterized by decreased insight and decreased awareness of patient's actions and their impact. In patients presenting with behavioral changes and GSM, FTD is one of the diagnoses to be taken in consideration.

**Key words:** frontotemporal dementia, genital self-mutilation, behavioral changes

## INTRODUCTION

Frontotemporal dementia (FTD) is the second most common cause of dementia in patients younger than 65 years (1). It is characterized by personality changes and impaired social conduct and represents the behavioral variant of the three clinical presentations of frontotemporal lobar degeneration (FTLD) (2). These changes are caused by progressive frontal and temporal lobe degenera-

tion (3). Behavioral disturbances vary among patients and there are two distinct behavioral syndromes: the apathetic subtype characterized by generalized loss of interest in activities and volition, loss of social emotions and decreased pain

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response; and the disinhibited subtype characterized by hyperorality, increased preference for sweet foods, exaggerated sensory responses and repetitive motor behavior (4). Patients with FTD commonly violate social norms, for example, by making sexually inappropriate comments, stealing, or public urinating (5). Cognitive impairment is not as pronounced as behavioral changes early in the course of the disease when many patients perform well on traditional neuropsychological tests. As the disease progresses, the number and severity of behavioral changes increase and cognitive impairment emerges, especially concerning executive functioning (6).

Sociopathic behavior has been described in FTD patients and can manifest itself as sexual aggressiveness and inappropriate sexual advances (7). Genital self-mutilation (GSM) has not yet been documented in FTD patients. We report on a patient with FTD developing inappropriate sexual behavior manifesting as mutilation of his penis.

## CASE REPORT

A 60-year-old patient presented with a two-year history of social withdrawal and loss of attention to household responsibilities. The patient himself denied having any problems. He had no memory difficulties and was able to manage the household chores. He had 12 years of formal education and had worked as a mechanic. Family history was negative for cognitive or psychiatric disorders.

Physical and neurological examination revealed only positive Babinski sign bilaterally. The patient scored 22/30 on the Mini Mental State Examination (MMSE). Cognitive assessment revealed multiple and severe dysfunctions, mainly in executive and attention tasks. He scored below normal range on digit span forward and backward (test of immediate attention) and Stroop Interference test; Trail A and B tests (tests of executive functions). He also scored in the impaired range on tests of naming and verbal fluency. His memory was still preserved. Results of neuropsychological assessment are presented in Table 1.

Brain magnetic resonance imaging showed bifrontal atrophy. Single-photon emission computed tomography revealed hypoperfusion in bilateral medial frontal and orbitofrontal regions of the cerebral cortex and in cingulate gyri. Cerebrospinal fluid examination revealed elevated tau protein and normal amyloid beta 42 protein levels (Table 2).

Table 1. *Results of neuropsychological assessment*

Test	Score
Mini Mental State Examination Test	22/30
<i>Long-term memory</i>	
Rey auditory- verbal learning test –immediate recall	30/75
Rey auditory -verbal learning test – delayed recall	5/15
Rey–sterrieth complex figure–recall	14/36
<i>Language</i>	
The Boston Naming test	38
Token Test	15/50
<i>Praxis</i>	
Rey–sterrieth complex figure–copy	31/36
<i>Attention and executive functions</i>	
Trail Making test A	65
Trail Making test B	321
Digit span	4
Controlled association letters test	6
The Stroop color-word interference test (mean reading time in seconds for the 24/ item card)	63

Table 2. *Results of cerebrospinal fluid examination*

Test	Result
hTAU Ag	301 ng/L
Phospho TAU (181P)	62 ng/L
Beta-AMINOID (1-42)	565 ng/L

The patient was diagnosed with the behavioral variant of FTLD (2). His behavioral problems progressed during a two-year follow up period and the patient became socially inappropriate with childish attitude. He maintained a stereotyped way of life with little social activity. According to his wife, he showed no sexual interest. Cognitive abilities deteriorated and he developed language difficulty with non-fluent speech. Despite all this, the patient was still functioning on his own and had preserved visuospatial functions. Cognitive assessment was repeated and perseveration and phonemic and semantic paraphasia were noted. He now scored 14/30 on the MMSE. Verbal fluency was greatly reduced as he was able to produce only three words beginning with “S” and 8 animals in one-minute period. On assessment of executive functions, he performed poorly having difficulties with alteration required in Part B of the Trails and incongruent condition of the Stroop task.

At that time, he was admitted to the hospital. During hospital stay, he was seemingly calm but developed paranoia involving his roommate. In the second week of his stay, nurses observed that the glans of his penis was swollen and a hematoma of the orifice of urethra was observed (Fig. 1). A bloody spoon was found at his bedside. Urethroscopy was performed and lacerations of the urethra were found, consistent with injuries from penile insertion of a blunt object. He did not have any insight and completely disregarded his act. Lesions of his penis recovered without sequelae and the patient was discharged home.



Fig. 1. Hematoma of the orifice of urethra

## DISCUSSION

The clinical syndrome of FTD is characterized by progressive changes in behavior, personality and/or language, with relative preservation of memory. Profound alterations in behavior can be an early sign and are known to affect personality and social conduct, as well as social inhibition (8). Disinhibition affects domains of interpersonal conduct (i.e. social intrusiveness, rudeness, inappropriate singing and making animal noises), regulation (i.e. inappropriate laughter) and sexual propriety (i.e. inappropriate touching, kissing and grabbing, getting into bed with other patients, public masturbation) (9). Cognitive deficits mainly occur in the domains of attention, planning and problem solving, whereas primary tools of language, perception and spatial functions are well preserved. As the disease progresses, most FTD patients become increasingly disinhibited with a decline of social and interpersonal skills (10).

GSM is an uncommon disorder and is usually described in psychiatric patients, mainly psychotic or

intoxicated ones (11). Reviewing the literature, we did not find any description of GSM in patients with FTD. On the other hand, sociopathy is a well described but underappreciated phenomenon in FTD patients. It is characterized by decreased insight and decreased awareness of patient actions and their impact (7). Its pathological paradigm is involvement of the right anterior temporal lobe and right orbitofrontal cortex, which are responsible for acquisition and development of social and moral reasoning and suppression of socially aberrant behavior (12,13). Such emotional and moral deficits are typical for psychotic patients in whom penile self-mutilations have been described. This, we believe, is the psychological platform for the manifested self-mutilation in our patient.

GSM is a disturbing yet so far not described symptom in FTD. In patients presenting with behavioral changes and GSM, FTD is one of the diagnoses to be taken in consideration.

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## Genitalno samoosakaćenje u bolesnika s frontotemporalnom demencijom

SAŽETAK - Opisan je slučaj pacijenta s frontotemporalnom demencijom (FTD), koji je osakatio svoj penis. Opisane su abnormalnosti mozga otkrivene slikovnim prikazom mozga magnetskom rezonancijom, *single photon* emisijskom kompjutoriziranom tomografijom i kognitivna procjena. Genitalno samoosakaćenje (GSO) je do sada neopisan, ali uznemirujući simptom FTD-a. Bolesnik u dobi od 60 godina bio je unazad dvije godine socijalno povučen i nije obraćao pozornost ni na kakav posao u domaćinstvu. Bio je hospitaliziran i utvrđena je dijagnoza FTD-a. Jednog su dana medicinske sestre opazile da mu je penis natečen i ozlijeđen razderotinom. Otkriveno je da je pacijent žlicom osakatio svoj penis. On nije za to mario i u potpunosti zaniijekao taj čin. FTD je drugi najčešći uzrok demencije u ljudi mlađih od 65 godina. Karakteriziraju ga promjene osobnosti i oštećeno socijalno ponašanje i predstavlja bihevioralnu varijaciju triju kliničkih ispoljavanja degeneracije frontotemporalnog režnja. Sociopatija je dobro opisan, ali podcijenjen fenomen u bolesnika s FTD-om. Karakteriziraju je smanjeni uvid i smanjena svjesnost pacijenta o svojim aktivnostima i njihov međusobni utjecaj. Kod pacijenata s promjenama ponašanja i GSO treba pomisliti i na dijagnozu frontotemporalne demencije.

**Ključne riječi:** frontotemporalna demencija, samoosakaćenje genitalija, promjene ponašanja