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COTARD SYNDROME IN SEMANTIC DEMENTIA

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Abstract

Background—Semantic dementia is a neurodegenerative disorder characterized by the loss of meaning of words or concepts. semantic dementia can offer potential insights into the mechanisms of content-specific delusions.

Objective—The authors present a rare case of semantic dementia with Cotard syndrome, a delusion characterized by nihilism or self-negation.

Method—The semantic deficits and other features of semantic dementia were evaluated in relation to the patient's Cotard syndrome.

Results—Mrs. A developed the delusional belief that she was wasting and dying. This occurred after she lost knowledge for her somatic discomforts and sensations and for the organs that were the source of these sensations. Her nihilistic beliefs appeared to emerge from her misunderstanding of her somatic sensations.

Conclusion—This unique patient suggests that a mechanism for Cotard syndrome is difficulty interpreting the nature and source of internal pains and sensations. We propose that loss of semantic knowledge about one's own body may lead to the delusion of nihilism or death.

INTRODUCTION

Semantic dementia is a neurodegenerative disorder characterized by the loss of semantics, or the meaning of nouns and objects.^{1,2} It is one of three major frontotemporal degenerations which have their usual onset in the presenium. Although these syndromes are pathologically heterogeneous, most patients with semantic dementia have anterior temporal pathology with abnormal intraneuronal ubiquitin-positive inclusions containing deposits of misfolded transactive response DNA-binding protein 43 (TDP-43).¹ Clinically, the earliest and most common deficit in semantic dementia is semantic anomia,² or language difficulty characterized by the loss of meaning of words. The deficits in semantic dementia, however, extend beyond language to involve a multimodal or “amodal” impairment in the knowledge of objects, faces, smells, other perceptions, or concepts.^{3,4}

This report describes a patient with semantic dementia who developed Cotard syndrome involving the conviction that she was dead or dying. Cotard syndrome is the delusion of nihilism or denial of general or particular features of the self.⁵ The patient's loss of semantic

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knowledge had extended to the nature and source of her internal discomforts. This unique case offers an opportunity to clarify the pathophysiology of Cotard syndrome. We consider the nature of her semantic and underlying deficits and how they might facilitate the emergence of nihilistic beliefs.

CASE REPORT

Mrs. A was a 56-year-old woman who presented with feelings of being dead and a complaint of progressive word finding difficulty. Over three years, she was progressively impaired in understanding nouns and their meaning. Her difficulty was affecting her ability to make herself understood in conversations and to understand instructions. Her husband gave an example of her inability to understand the word “laundry” but normal comprehension for “washing clothes.” Another example was that she used to be a vegetarian but lost the meaning of that word “vegetarian” and began eating animal products.

Like many patients with semantic dementia, Mrs. A became rigid and developed compulsive preoccupations.⁴ These included rigidly maintaining routines such as going to bed at a certain time and eating certain foods such as eating only burritos for every meal. The patient also became preoccupied with somatic sensations, which she variously referred to as body aches, pains, discomforts, and inner “traveling” feelings. Mrs. A stated that all her prior external pains were transferred to the inside of her body, from her mouth and surrounding oral area all the way down to the insides of her feet. She could not otherwise describe her sensations, but incessantly complained about them at every conversation. The patient could not describe where her internal sensations were located. She could not point to the part of the body that was bothering her, but would indicate the sensations or discomforts as occurring in her torso.

Mrs. A reported that her sensations indicated that she was wasting away inside. She stated: “my body is not working from the inside and doesn't feel right. I am dying from the inside.” No evidence to the contrary could alter her conviction of dying. At one point, felt that she had completely died and then come back to life. She could not elaborate on her feeling of having died except that it felt as if her “insides were gone.” There was no religious aspect to her belief, nor did she report other unusual beliefs, hallucinations, or misidentifications of persons or place. Although she had a history of a non-psychotic, depressive episode, she denied that she was currently depressed and did not otherwise endorse symptoms of depression such as decreased mood, anhedonia, or decreased energy, sleep, or appetite. Her past medical history was remarkable for Hashimoto's thyroiditis and migraine headache.

Mrs. A's examination showed her to be alert and attentive, with digit span of six forward and four in reverse, eager to converse, and emotional detached from her matter-of-fact reports of nihilism and dying. Her neurological examination, including cranial nerves, gait and coordination, motor, and sensory examinations, was entirely intact, and her reflexes were normal, symmetrical, and without pathological reflexes.

Her language examination showed significant word comprehension difficulty. Her speech was fluent and without paraphasic errors but characterized by word-finding pauses and hesitations. Mrs. A was unable to generate a list of animal names or an additional list of foods. Confrontational naming was severely impaired, and she was unable to name common items such as a pen, glasses, and shoes. She scored 1/15 on the mini-Boston Naming Test,⁶ and, when later given the item names and asked to identify them, she did not know what they were. The semantic aspects of these items were frequently absent, i.e., when subsequently given the word, she could not point to the object or describe its function or use. Mrs. A had surprising difficulty identifying body parts and did not know the meaning of

chin, knee, nose, or hair. She did not know the organs from where her discomforts originated and could not recognize or describe major organs, such as heart, stomach, liver, or intestines from their names or pictures. Other language tests included normal repetition and comprehension to commands when she understood the nouns in the command. Consistent with surface dyslexia, she could read regular and nonsense words but misread irregular words in a phonetic fashion.

Her word comprehension difficulty affected scores on other neuropsychological measures. On her Mini-Mental State Examination,⁷ her score was only 10/30 because she could not perform the serial reversal items, recall, and three-step command due to comprehension difficulty. She could not complete the verbal learning memory test because of difficulty understanding the words; however, she demonstrated episodic, declarative memory for recent events, conversations, and instructions. Mrs. A was able to draw simple shapes and visuospatial constructions and perform simple calculations but could not comprehend similarities. On a supplemental series of 24 famous faces, she failed to recognize any of them, but she could match two sets of faces of the same persons, and she was able to recognize her own face in the mirror.

Magnetic resonance imaging showed bilateral anterior temporal lobe atrophy, most prominent on the left side than the right (See Figure). Single photon emission tomography showed hypoperfusion in the anterior temporal lobes, again more prominent on the left.

Mrs. A had a gradually progressive loss of semantic abilities, surface dyslexia with difficulty reading irregular words, and prosopagnosia involving the recognition of familiar faces. These findings, along with her neuroimaging, were consistent with semantic dementia, which, in this patient, was accompanied by Cotard syndrome. Her delusions of nihilism decreased to only an occasional report or preoccupation with risperidone 0.5 mg qd.

DISCUSSION

We report a patient with semantic dementia and Cotard syndrome. This unique patient affords insights into the nature of these false beliefs. The nihilistic delusions appeared to emerge from a preoccupation with somatic or internal sensations and inability to know their meaning or source. She had lost not only the ability to describe these sensations, but had also lost the identity of the body parts from where they emerged. Although delusions, including somatic delusions, are common in focal neurodegenerative diseases, actual reports of somatic delusions in semantic dementia are rare. They include a patient who also had somatic preoccupations and misinterpreted the sensation that worms were crawling under her skin and resulting in her veins and bunions.⁸ The emergence of Cotard syndrome when semantic dementia results in misinterpreted somatic sensations indicates that the delusion of nihilism in our patient with semantic dementia is more than just a rare coincidence of two infrequent conditions.

First described by Jules Cotard in 1880, the nihilistic beliefs in Cotard syndrome involve convictions of self-negation, which are firmly held despite proof or evidence to the contrary.⁹ Cotard syndrome usually extends to the delusion of being dead or dying; however, this denial of self-existence is absent in nearly a third of patients.⁹ Although most commonly associated with negative self-attribution from psychotic depression,^{9,10} Cotard syndrome can be associated with cognitive deficits from neurological disease. Reported causes include Parkinson's disease, migraine, brain tumors, traumatic brain injury, arteriovenous malformations, multiple sclerosis, and encephalitis.^{5,11} For example, the patient with encephalitis complained of missing both hands and felt that her internal organs had stopped working. Similar to our patient, who felt internal sensations but did not know

their meaning, this encephalitis patient reported that she did not feel her body “from the inside.” Both patients, therefore, concluded that they must not exist and were dead or dying.

Mrs. A has more than just a blunting of pain and sensations. She has decreased knowledge of internal body parts as indicated by inability to name, localize, and identify and describe major organs from their names or pictures. Her somatic sensations may be unable to trigger their corresponding meanings and associations. This can occur when sensations lead to altered feelings of familiarity,¹¹ possibly because of disconnection from limbic-emotional areas.¹⁰ Alternatively, the corresponding semantic information may simply be lost or decayed in patients with semantic dementia.³ Either way, there is a dissociation somatic sensations and their corresponding semantic attributes. In addition, because Cotard syndrome is a delusional belief, it also involves an inability to accurately evaluate reality.^{13,14} In this delusion, the problem assessing reality could result from impaired source monitoring, or the ability to apprehend the origin of her internal “aches and pains.”¹⁵

Her loss of knowledge of body parts resembles asomatognosia, or the neurological loss of awareness of parts of one's body, or somatoparaphrenia, or the denial of ownership of a limb or one half of the body.^{16,17} These neurological syndromes usually result from right-sided lesions temporoparietal and medial frontal lesions,¹⁹ areas that overlap with the pathology in semantic dementia. Mrs. A, however, did not have a focal lesion with feelings that contralateral parts of the body were missing or had disappeared.¹⁶ Moreover, although up to 8.3% of patients with semantic dementia may have Capgras syndrome or a related misidentification syndrome,¹⁷ Mrs. A did not have self-misidentification, as the patient was perfectly able to recognize her external self.

In conclusion, the emergence of Cotard syndrome in this patient with semantic dementia may have resulted from a dissociation of somatic sensations from semantic knowledge, followed by a failure to evaluate her “reality” of dying. This patient and the proposed mechanisms of nihilistic delusions offer intriguing insights and future directions for investigating the psychological reactions to somatic sensations. Research can further investigate the relationship between unrecognized somatic sensations and whether patients who misinterpret their discomforts or sensations develop delusions or other psychopathology.

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Figure 1. Magnetic resonance imaging shows bilateral anterior temporal atrophy, left worse than right, consistent with semantic dementia. From left to right, the images are T1-weighted, FLAIR, and T1-weighted with contrast injection.