

Reviewing Delusional Misidentification Syndromes with Examples

Örneklerle Sanrısız Yanlış Tanımlama Sendromlarının Gözden Geçirilmesi

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ABSTRACT

Delusional misidentification syndrome is characterized by individuals perceiving familiar people, places, and objects as different entities, often associated with delusional disorders. These disorders are typically linked to abnormalities in cognitive processes, resulting in incongruent and unalterable beliefs. The loss of familiarity is believed to be the consequence leading to the emergence of these disorders. While commonly associated with psychiatric illnesses, they are also frequently observed in conjunction with neurodegenerative diseases. Diagnosis is primarily established through clinical evaluation. However, cases of these syndromes pose a significant burden on caregivers. Therefore, it is crucial not to overlook the possibility of dementia in these syndromes. This consideration is vital for providing appropriate support and treatment to patients and their families. This case report aimed to provide a detailed examination of this topic by presenting five cases.

Keywords: delusional misidentification syndrome; delusional disorders; Capgras syndrome; mirror self-misidentification syndrome; Alzheimer's dementia; vascular dementia; Lewy body dementia.

ÖZ

Sanrısız yanlış tanımlama sendromu, bireylerin tanıdık insanları, yerleri ve nesnelere farklı varlıklar olarak algılamasıyla karakterize edilir ve sıklıkla sanrısız bozukluklarla ilişkilendirilir. Bu bozukluklar genellikle bireyin düşünce süreçlerindeki anormalliklerle ilişkilendirilir ve gerçekle uyumsuz ve değiştirilemez inanışlarla sonuçlanır. Bu bozuklukların aşinalığın kaybolması sonucu ortaya çıktığı düşünülmektedir. Genellikle psikiyatrik hastalıklarla ilişkilendirilmeseler de aynı zamanda nörodejeneratif hastalıklarla birlikte de sıkça görülür. Tanı, öncelikle klinik değerlendirme yoluyla konmaktadır. Ancak vakalar, bakım verenler üzerinde ciddi bir yük oluşturur. Dolayısıyla, bu sendromlarda demans olasılığının göz ardı edilmemesi önemlidir. Bu durum, hastalara ve ailelerine sağlanacak uygun destek ve tedavi açısından kritiktir. Bu olgu sunumunda beş olgu sunularak bu konuya dair detaylı bir değerlendirme sağlanması amaçlandı. **Anahtar kelimeler:** Sanrısız yanlış tanımlama sendromu; sanrısız bozukluklar; Capgras sendromu; aynada kendini yanlış tanımlama sendromu; Alzheimer demans; vasküler demans; Lewy cisimcikli demans.

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INTRODUCTION

Delusional misidentification syndrome (DMS) is a delusional syndrome characterized by the mistaken identification or perception of familiar individuals, places, and objects (1,2). It can manifest in various forms: Capgras syndrome, which is characterized by the belief that a known person has been replaced by an impostor (3); Fregoli syndrome, where a stranger is mistakenly identified as a familiar person who has changed appearance (4); intermetamorphosis, where the belief is that two known

individuals have switched places (5); subjective doubles, where a person believes they have an identical twin who acts independently (6); and mirror self-misidentification syndrome, where one misidentifies their reflection as a separate person. Although these syndromes are primarily associated with psychiatric disorders, they have also been observed in neurodegenerative diseases and organic brain damage. DMS can accompany neurodegenerative diseases, particularly Lewy body dementia, vascular dementia, and Alzheimer's dementia. In order not to overlook dementia in these delusional syndromes that are more commonly associated with psychiatric illnesses, we aimed to present five cases to review this topic.

CASE 1

A 78-year-old right-handed male patient, who is literate, presented to the clinic with episodic memory complaints that have been occurring for three years, along with a behavioral impairment of greeting himself in the mirror as someone else. The problems started with complaints of losing objects three years ago, followed by difficulties in recognizing family members and rummaging through rooms. All his needs are taken care of by his relatives. His medical history includes known diabetes mellitus, epilepsy, stroke, coronary artery disease, and hypertension. There is no significant family history. Neurological examination is unremarkable. In the neuropsychological evaluation, the Beck depression inventory (BDI) score was 7 which indicated no clinical depression. Also, the mini-mental state examination (MMSE) score was 13 (moderate dementia). While spatial orientation is preserved, there are impairments in time orientation, registration memory, and perceptual organization. The clinical dementia rating (CDR) score is 16. A brain imaging was performed using a magnetic resonance imaging (MRI) was performed. Fazekas scale score, which is a scale that indicates the level of small vessel disease was stage 2, and global atrophy in MRI at the sequence of fluid-attenuated inversion recovery (FLAIR) was observed (Figure 1A). The coexistence of Alzheimer's dementia and vascular dementia was considered. Mirror self-misidentification syndrome was improved after the treatment by donepezil 10 mg/day and memantine 20 mg/day into the six weeks without any improvement in neurocognitive test scores.

CASE 2

A 79-year-old right-handed female patient, who is literate, presented to our clinic with complaints of hoarding items, walking naked at home and talking to someone else in the mirror, which has been occurring for two years but has worsened in the past eight months. During the inquiry, it was revealed that she had a pre-existing REM sleep behavior disorder, and within the past two years, she had developed misbehaviors such as rummaging through rooms, urinary incontinence, aimless wandering at home, and closing curtains due to the belief of being watched. Her self-care needs are taken care of by her relatives. She has a medical history of controlled hypertension. There is no significant family history. Neurological examination showed slowness in walking, mild bilateral rigidity, and echolalia. In the neuropsychological evaluation, the MMSE score was 9 (moderate-advanced dementia), and the BDI could not be performed due to inconsistency. The

CDR score is 16. The Koedam score, which indicates atrophy of the posterior cerebral areas, was found to be stage 2 (Figure 1B). Lewy body dementia was considered in the patient. After rivastigmine 9.5 mg/day transdermal patch and 12.5 mg/day quetiapine treatment, misbehaviors including mirror self-misidentification syndrome were resolved in one month.

CASE 3

A 48-year-old right-handed male patient, who has received eight years of education, presented to the clinic with an increase in difficulties remembering proper names and a facial recognition impairment last six months. During the inquiry, it was revealed that he couldn't even recognize his mother's face, found her expressions strange, experienced difficulties with orientation, developed a lack of interest in his surroundings, and exhibited behavioral problems such as swearing in public. The symptoms were found to be gradual and progressive over two years. His functional abilities were preserved. He has a history of cerebrovascular event two years ago. There is a family history of an unspecified psychiatric illness in his sibling. Apart from neurocognitive disorders, right hemihypoesthesia was detected in the neurological examination. BDI score was found 28. Personal and actual information, as well as spatial and temporal orientation, were preserved. However, there were difficulties in verbal memory processes, including encoding and retrieval, while lexical fluency was relatively preserved but semantic fluency was severely impaired. Dorsal and ventral pathway functions related to visuospatial abilities were impaired. Cranial imaging was performed with an MRI diffusion sequence including FLAIR due to claustrophobia, revealing chronic encephalomalacic areas in the left parietal and right temporal region (Figure 1C-D). Vascular dementia was considered in the patient. Treatment was initiated with rivastigmine 4.6 mg/day transdermal patch and 50 mg/day sertraline. The rivastigmine dose was increased to 13.3 mg. The sertraline dose was not changed. Although facial recognition did not improve, swearing and apathy improved in three months.

CASE 4

An 86-year-old right-handed male patient, who has received 15 years of education, presented to the clinic with complaints of seeing and talking to things that are not present at home for six months. He believed that his relatives were replaced with other people for three months (Capgras syndrome). During the inquiry, it was revealed that he has been experiencing fluctuating orientation disorder, mixing up names, losing objects, difficulty in naming, confusion with directions, problems with financial calculations, and mistaking sheets for people for the past two years. He is able to take care of his self-care needs. He has a medical history of asthma and benign prostatic hyperplasia. There is no significant family history. Neurological examination showed limitations in upward gaze, festination, dizziness upon standing up from sitting, rigidity, and circumferential speech. In the neuropsychological evaluation, the BDI score was 5, and the MMSE score was 24. Personal and actual information and spatial orientation were preserved, but there were relative difficulties in time orientation, difficulties in retrieval in verbal memory processes, severe

impairments in semantic and lexical fluency, and impaired organization. The Koedam scale indicated stage 2-3 in brain imaging (Figure 1E). Although there was no change in the improvement in cognitive evaluation, Capgras syndrome significantly regressed three weeks after starting 4.6 mg/day transdermal rivastigmine treatment.

CASE 5

A 77-year-old right-handed male patient, who has received three years of education, presented to our clinic with the inability to recognize himself in the mirror and mistaking the reflection in the mirror for three months. During the inquiry, it was revealed that he has been experiencing repeated questioning and topic repetitions for at least five years. He has started to lose his sense of direction and is unable to leave the house alone, leading to a decline in his functionality. He requires assistance with self-care. There is no known medical condition. It was learned that his mother and sibling have complaints of unconfirmed forgetfulness. Neurological examination did not reveal any pathology. In the neuropsychological evaluation, the BDI score and MMSE score were 14. Personal and actual information, as well as spatial and temporal orientation, were moderately impaired. Simple attention and working memory processes were relatively preserved, but there were difficulties in encoding verbal memory processes. There was an impairment in the organization. The mesial temporal atrophy (MTA) score, which indicates hippocampal degeneration, was found to be stage 3-4 (Figure 1F). Alzheimer's dementia was considered in the patient. The donepezil/memantine combination was started with a dose of 5/10 and increased to a dose of 10/20 mg/day. There was no change in his misidentification.

DISCUSSION

Delusional misidentification syndrome is a delusional disorder in which individuals mistakenly identify familiar objects or people as something or someone else. It can be a neurological or psychiatric condition and is characterized by the delusional belief of misidentification. The common feature of all DMSs is the loss of familiarity. The most commonly seen form is Capgras syndrome, but other forms include Fregoli syndrome, intermetamorphosis, subjective doubles, and mirror self-misidentification syndrome. Among the cases we have encountered, case 4 was diagnosed with Capgras syndrome, and the other four exhibited the phenomenon of misidentifying oneself in the mirror.

In Capgras syndrome, individuals can not recognize a loved one or acquaintance and believe that they have been replaced by someone else. The phenomenon of misidentifying oneself in the mirror, on the other hand, is considered normal in animals and infants but is not expected in adults. It can be observed in conjunction with

disturbances in self-perception and visual-spatial perception associated with dementia disorders (7). These mental disorders typically manifest as symptoms of neurological or psychiatric conditions. They have been described in schizophrenia and schizoaffective disorders, and although they are more commonly associated with psychiatric illnesses, they can also occur in Alzheimer's dementia, Lewy body dementia, vascular dementia, epilepsy, cerebrovascular events, and advanced Parkinson's disease. The prevalence of these disorders has been reported to range between 5% and 82%. A review of 260 case reports related to DMS found that 174 (66.9%) of the cases had Capgras syndrome. Among the cases, 73% had a diagnosis of schizophrenia, 26.4% had dementia, and 16.7% had mood disorders (8). In two other studies, the rates of these phenomena occurring in different neurodegenerative diseases were reported to be between 16.6% and 27.8% in Lewy body dementia, and 15.8% in Alzheimer's disease (9,10). In another series of 47 patients with Capgras syndrome, it was found that 81% of the patients had a neurodegenerative disease, with Lewy body disease being the most common, followed by Alzheimer's disease. In individuals without neurodegenerative diseases, DMS typically emerges at a younger age and has been associated with psychiatric disorders, cerebrovascular events, and illicit drug use (11). Among our patients presented, the primary neurological diagnosis was as follows; one had a combination of Alzheimer's disease and vascular dementia (Case 1), two had Lewy body dementia (Case 2 and Case 4), one had vascular dementia (Case 3), and one had Alzheimer's disease (Case 5).

The pathophysiology of DMS appears to involve a disconnection syndrome where the connection between emotional information processing and face recognition is disrupted. It has been found that patients particularly have difficulty recalling past experiences with a hypoactive limbic system in the right hemisphere (12). Neuroimaging studies have also shown abnormal face recognition in these patients (13). Based on these findings, it has been suggested that there is a disconnection (caused by neurodegenerative or functional lesions) between the occipitotemporal cortex (involved in face recognition) and limbic circuits (neural circuits involved in the control of emotional expression) (14). In Lewy body dementia, hallucinations and misperceptions are common and are associated with Lewy body pathology in specific anatomical areas, which can involve disrupted cortical connections between the occipital and temporal lobes (15). A study examining 17 patients with Capgras syndrome based on antemortem imaging also found a neuroanatomical disconnection between impaired familiarity processing (left retrosplenial cortex) and belief evaluation (right frontal cortex) (16).

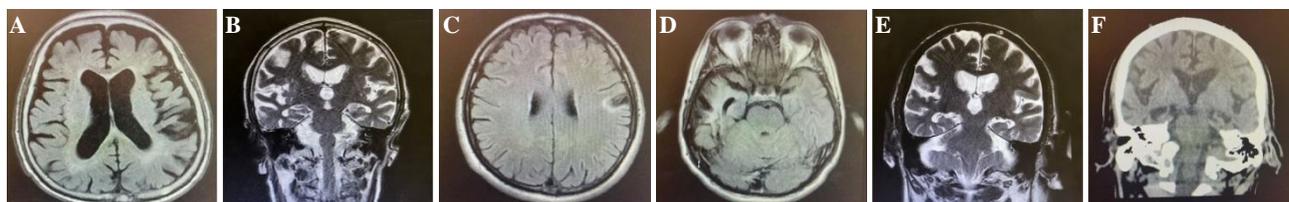


Figure 1. Magnetic resonance imaging of cases, A) case 1, B) case 2, C-D) case 3, E) case 4, F) case 5

DMS is a clinical diagnosis, and its diagnosis is based on the clinical evaluation of symptoms (8). DMS is one of the most challenging clinical findings for caregivers. In our five patients, the complaints related to these delusional syndromes were the reason for seeking medical attention, despite the presence of other symptoms before. This condition, which disrupts the trust relationship between the caregiver and the patient more than the loss of recognition itself, undermines the effectiveness and safety of care and intensifies the burden on the caregiver (17). The patient does not allow assistance with self-care from someone they perceive as an imposter or fake. Confusion, fear, and anger lead to the rejection of attention from the caregiver, and in some cases, it can even result in aggressive behaviors (17,18). Alternatively, individuals with mirror self-misidentification syndrome may feel anger towards their family members who try to convince them that they are using their own belongings at their own homes. Although there was no aggression in our patients, they exhibited persecutory feelings towards their caregivers. Pharmacological or non-pharmacological interventions have been shown to not completely eliminate DMS. However, antipsychotic medication combined with therapies that make the patient feel safe, such as music or reminiscence therapy, as well as transcranial magnetic stimulation methods, have been tried (19,20). Approaching the patient with empathy and showing interest in their anxieties related to their condition can enhance the effectiveness of treatments (21). Hospitalization is also an option in cases where the patient poses a risk to themselves or others. We have observed benefits from symptomatic treatments of dementia including acetylcholine esterase inhibitors and memantine in DMSs in dementia cases. Due to the overlap of their current symptoms with psychiatric disorders, our patients remained untreated in the neurology department for a long time, highlighting the importance of recognizing that these conditions can also be seen in neurodegenerative diseases.

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