A Case of Capgras Syndrome With Frontotemporal Dementia

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Abstract: Capgras syndrome (CS), also called imposter syndrome, is a rare psychiatric condition that is characterized by the delusion that a family relative or close friend has been replaced by an identical imposter. Here, we describe a 69-year-old man with CS who presented to the Kemal Arikan Psychiatry Clinic with an ongoing belief that his wife had been replaced by an identical imposter. MRI showed selective anterior left temporal lobe atrophy. Quantitative EEG showed bilateral frontal and temporal slowing. Neuropsychological profiling identified a broad range of deficits in the areas of naming, executive function, and long-term memory. On the basis of these findings, we diagnosed frontotemporal dementia. This case demonstrates that CS can clinically accompany frontotemporal dementia.

Key Words: Capgras syndrome, frontotemporal dementia, misidentification syndrome, magnetic resonance imaging

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CS = Capgras syndrome. **FTD** = frontotemporal dementia.

Capgras syndrome (CS) clinically manifests as an "imposter" delusion whereby the patient believes that a person close to him or her (eg, a relative or friend) has been replaced by an identical imposter (Kimura, 1986). The delusion does not appear to be a facial recognition or memory issue because CS patients often recognize that the imposter looks identical to the actual person they are referring to (Barelle and Luaute, 2018; Berson, 1983). However, the delusion can become a significant disability because the patient can become aggressive toward the perceived imposter.

Although CS is most frequently observed in patients with schizophrenia, it has also been identified in individuals with affective psychosis and organic psychotic syndromes (Kimura 1986), as well as in patients with dementia (Harwood et al, 1999). For instance, Harwood et al (1999) reported CS presentation in 10% of patients who had been diagnosed with Alzheimer disease. Harciarek and Kertesz (2008) also claimed that misidentification syndromes such as CS, reduplicative paramnesia of place, or phantom boarder phenomenon can occur in other types of dementia such as Lewy body dementia and semantic dementia.

CS has also been observed in patients with acquired brain lesions. A comprehensive literature review of clinical cases by Darby and Prasad (2016) showed that lesion-related delusional misidentification syndromes most commonly occur in cases where the right cerebral hemisphere is affected, in particular, the frontal lobe. CS can also manifest throughout the course of other neurodegenerative disorders, such as Huntington disease, and can even be brought about by drug-induced psychosis (Gama Marques, 2015; Gama Marques and Carnot, 2016).

CASE REPORT

A 69-year-old, right-handed man presented to the Kemal Arikan Psychiatry Clinic with a persistent conviction that his wife of 43 years had been replaced by an identical imposter. The man was a well-educated person and held a master's degree in business administration. He had retired 10 years ago, after working as a chief accountant for various large-scale international companies. He had also lectured on business accounting at a respected institution in Turkey for a couple of years. With regard to employment, the patient's wife described him as a highly conscientious, ambitious, and hard-working person. In his private life, she described him as a cultured person who loved to read and had a full social life. The patient also enjoyed sports and had played in an amateur-league football team until a few years ago. He was also a dutiful and caring father of two now-adult children and had seen to their education and personal development.

Three years ago, the patient started experiencing neuropsychological issues. Family members reported noticing that the patient began to find it difficult to form words (anomia). However, as this did not significantly affect the patient's life, the family did not seek medical help at that time. A few months before his referral to our clinic, the patient developed CS, which is what prompted

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him to seek medical help. There was no family history of psychiatric or behavioral problems and, before this, the patient had never been given a psychiatric or neurologic diagnosis, nor had he ever used any psychotropic medications or illegal substances. His blood chemistry, a vitamin and hormone panel, and serological tests (such as for venereal disease and the human immunodeficiency virus) were all normal.

When questioned in detail about his imposter delusion, the patient acknowledged that, although the imposter resembled his wife, he perceived his wife to be an imposter. This delusion persisted daily and caused moderate to severe distress to the family, with the patient often exhibiting negative emotional reactions (namely aggression) toward his wife. Sometimes, he even prevented her from doing household chores because only "his wife" was allowed to do them. The patient also misidentified his home, frequently not believing it to be his real house with his own furniture.

The patient's wife also reported that the patient had recently developed an excessive and impulsive desire for sexual activity. For example, he often suggested that he and his wife have "public" sexual intercourse (ie, while in the presence of their children and grandchildren), which caused a great deal of embarrassment. As well as misidentifying his wife, the patient also often accused her of having an affair with another man. In addition, he was consistently compulsive in vocalizing his political beliefs and frequently forced conversations on the topic of political issues, even with strangers (eg, on public transport). According to his wife, the patient also extensively confabulated stories about visits to countries and places that he had never been to. All of these symptoms followed the onset of CS, and it was their cumulative detrimental effect that led the family to seek medical help.

ASSESSMENTS

Neurologic Examination

On examination, the patient was cooperative and was oriented to time, place, and people. He exhibited good self-grooming and hygiene. His speech was spontaneous and was appropriate for the situation. His muscle strength was normal, and no pathological reflexes were identified. Ataxia and dysmetria were absent, based on the finger-to-nose test and performance of rapidly alternating movements. Spontaneous gait and tandem walking were also normal; however, mild ideomotor apraxia was observed when the patient was asked to mime brushing his teeth and combing his hair, although his knowledge on the use of these tools was intact.

Neuroimaging—MRI

An MRI including sagittal and axial T_2-weighted and coronal FLAIR sequences showed prominent atrophy of the patient's left temporal lobe, including the hippocampus. Interestingly, the atrophy was unilateral and was not detected in the right temporal lobe or either frontal lobe (Figure 1).

Neuroimaging—EEG

Quantitative EEG and absolute power mapping was performed to quantify the power of commonly analyzed EEG power bands: Delta (1–4 Hz), theta (4–8 Hz), alpha (8–12 Hz), beta (12–25 Hz), and high-beta (25–30 Hz) showed a significant increase in slow theta and delta waves in the frontal and temporal lobe electrodes as compared to the population mean (ie, *z*-scored results) (Figures 2 and 3). A significant increase in delta and delta bands suggests a slowing of brain activity. This result is consistent with the MRI findings in that slowing was more prominent on the left side of the brain.

Neuropsychological Testing

The patient completed an interview-based neuropsychological battery composed of various tests measuring memory, language, executive functions, abstract reasoning, and visuospatial functions (Table 1). The tests were administered by an experienced neuropsychologist (S.A.K.). All of the tests are international neuropsychological measures except the Verbal Memory Processes Scale (Öktem, 1992), which was developed in Turkey. However, all of the tests have

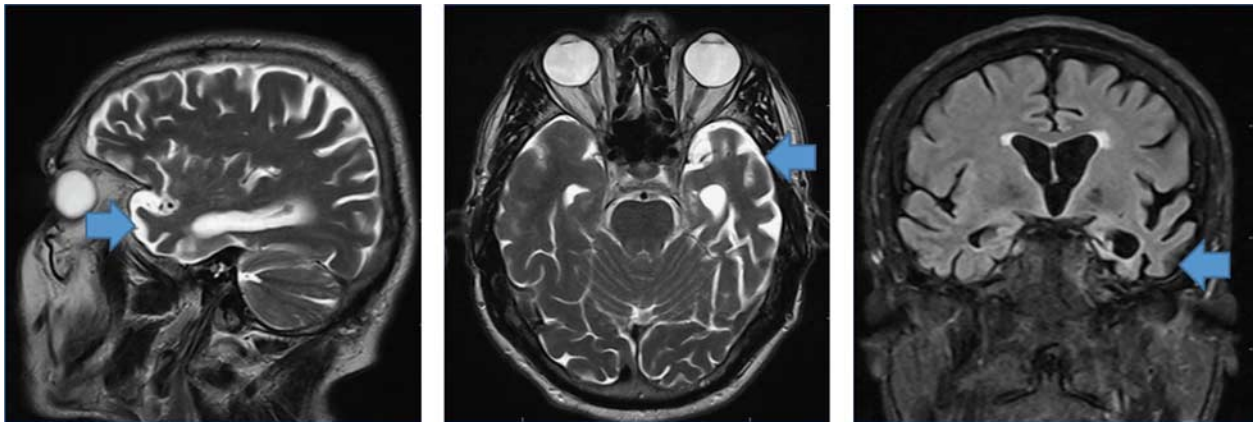

FIGURE 1. MRI showing atrophy in the left temporal lobe (indicated with arrows). (Figure 1 can be viewed in color online at www.cogbehavneurol.com.)

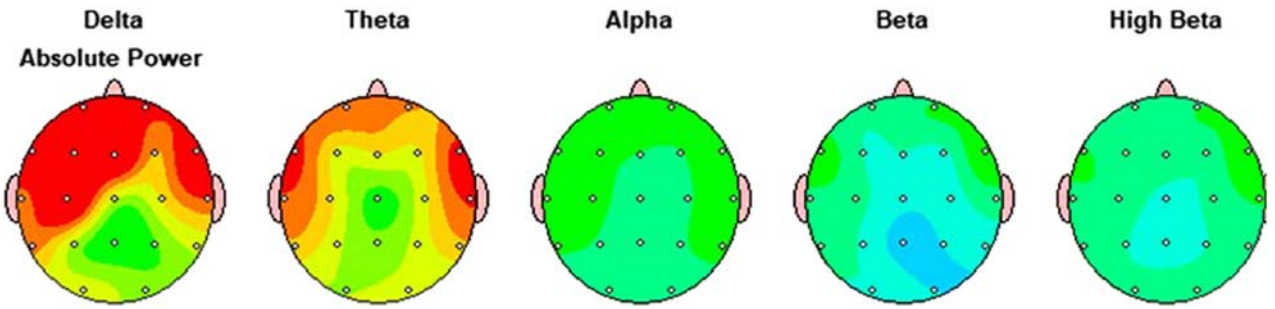

FIGURE 2. *Z*-scored fast fourier transform summary. (Figure 2 can be viewed in color online at www.cogbehavneurol.com.)

been validated for the Turkish population and this specific patient's age group; education-adjusted norms are also available.

Memory

The Verbal Memory Processes Scale measures both verbal short-term and long-term memory, including delayed retrieval and recognition after delay. During the test, a 10-item word list is repeated eight times. Regarding short-term memory, the patient remembered only four of the items, indicating a severe deficit. For delayed recall, the patient was unable to recall any of the items, indicating a severe impairment. And, in terms of recognition after delay, the patient recognized only half of the items that he had been shown and falsely recognized five words. These results indicate mixed-type memory impairment, with both hippocampal and frontal deficits.

Language

On the Boston Naming Test (Lansing et al, 1999), the patient was able to name only four items (out of 31) spontaneously, indicating severe anomia. For the remainder of the test, he required a phonemic or semantic cue. The patient also had semantic, phonemic, and neologistic paraphasias. During the Cookie Theft task from the Boston Diagnostic Aphasia Examination (Giles et al, 1996), the patient used neologisms and exhibited phonemic paraphasias. These results indicate severe naming and language impairment.

Executive Functions (see Faria et al, 2015, for Details of the Tests)

During the Digit Span Test, the patient was able to repeat only five digits forward and only two digits backward. This performance was interpreted as showing problems with attention span and working memory. During the Clock-Drawing Test, the patient wrote numbers greater than 12 on the clock face and placed the clock hands incorrectly. This pattern indicates a planning and conceptual deficit in addition to a perseverative pattern. On the Stroop Test, the patient could not name any of the colors under the incongruent condition. This performance shows problems with interference control and inhibition. During the Verbal Fluency Test, the patient is first required to produce names from a certain category (eg, animals), then to name objects starting with the letters K,

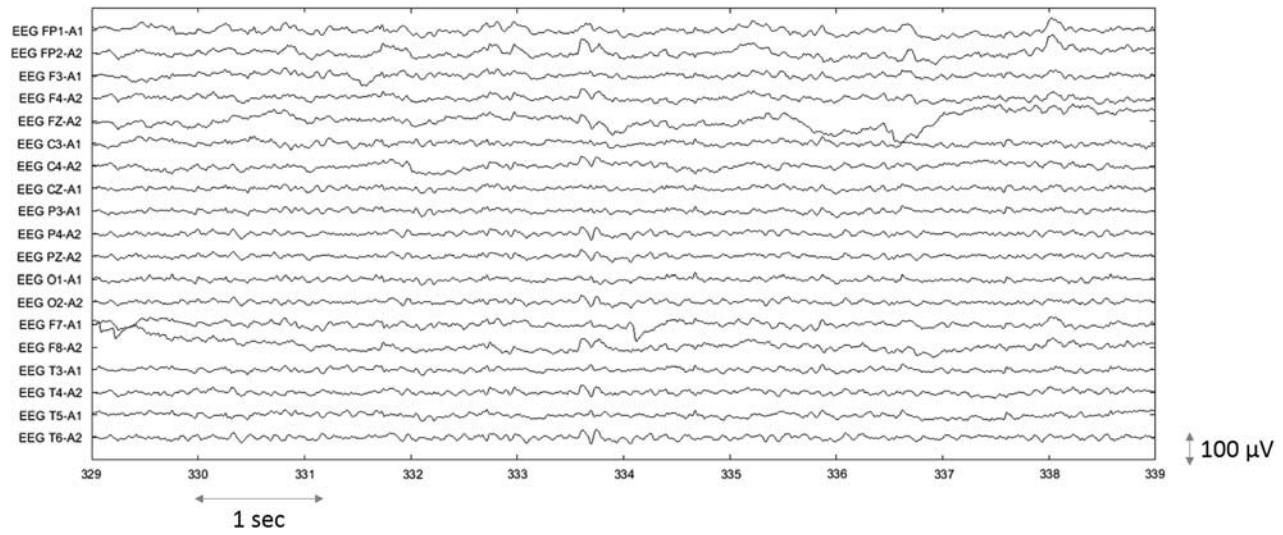

FIGURE 3. EEG trace with prominent frontal slowing.

TABLE 1. The Patient's Neuropsychological Evaluation of Memory (Short-term, Long-term, Delayed Recall), Language, Executive Functions, Abstract Reasoning, and Visuospatial Planning

Ability	Test	Interpretation
Memory	Verbal Memory Processes Scale	Severely impaired short-term memory and mixed-type memory impairment (frontal and limbic)
Language	Boston Naming Test	Severe anomia
	Cookie Theft task	Severe language impairment
Executive Functions	Digit Span Test	Problems with attention span and working memory
	Clock-Drawing Test	Planning and conceptual deficit
	Stroop Test	Severely impaired interference control and inhibition
	Verbal Fluency Test	Severely impaired semantic information processing
Abstract reasoning	WAIS–IV Similarities subtest	Severely impaired abstract reasoning
Visuospatial planning	Benton Line Orientation Test	Mild impairment
	Benton Facial Recognition Test	Normal

WAIS–IV = Wechsler Adult Intelligence Scale, Fourth Edition.

A, and S. The patient managed to produce only two animal names in the allotted 1 minute and only seven words starting with the letters K, A, and S. This performance is consistent with severely impaired semantic information processing.

Abstract Reasoning

We used the Similarities subtest of the Wechsler Adult Intelligence Scale, Fourth Edition (Wechsler, 2008), to measure the patient's abstract reasoning. The patient could detect the similarity correctly for only two out of 20 items, indicating severe impairment.

Visuospatial Planning

We used the Benton Judgment of Line Orientation Test (Qualls et al, 2000) and the Benton Facial Recognition Test (Tranel et al, 2009) to measure the patient's visuospatial function. The Line Orientation Test involves finding the match of lines with different orientations to the lines in a response card. The patient's performance was only slightly impaired as he scored 17 out of 30; a score of 20 is considered to be normal. The Benton Facial Recognition Test involves recognition of a target face among a set of different faces. The patient's score on this test was normal (41 out of 54). These findings indicate that the patient's visuospatial function was largely preserved.

DISCUSSION

Taken together, the deficits displayed by our patient—dysexecutive neuropsychological pattern, disinhibition, and perseverations—and the MRI findings led to the diagnosis of probable frontotemporal dementia (FTD), in accordance with the Neary criteria (Mohandas and Rajmohan, 2009). The most likely subtype of FTD is semantic dementia, based on the patient's prominent naming deficits and MRI scans of characteristic anterior temporal lobe atrophy (Rascovsky and Grossman, 2013). On the other hand, the patient also exhibited severe executive dysfunction and impaired working memory, which is not typical of semantic dementia. This combination of the severe behavioral phenotype and the dysexecutive pattern suggests a behavioral variant of FTD (Pressman and Miller, 2014).

The patient was prescribed an antipsychotic (risperidone, titrated to 3 mg) to control his delusions and sexual disinhibition. He benefited significantly from this treatment; his wife reported a marked decrease in his disinhibited sexual behavior and CS symptoms after a few days. Despite a lack of evidence (Boxer et al, 2013; Li et al, 2015), off-label therapeutic trials of donepezil (10 mg) and memantine (20 mg) were initiated; however, according to his wife's report, the patient showed no improvement in naming and memory functions following these medications. In the last visit after 6 months of treatment initiation, the symptoms related to CS were still controlled with risperidone, and no improvement in the patient's other cognitive symptoms was described.

It is noteworthy that the patient exhibited marked asymmetrical temporal lobe atrophy on an MRI. Asymmetrical temporal lobe involvement has been reported in 35% of patients with a behavioral variant of FTD (Whitwell et al, 2013), with some studies demonstrating that the side of the brain that is affected in FTD confers the types of neuropsychological deficits that will be displayed by patients. For example, the right-sided variant is associated with behavioral dyscontrol, personality changes, aphasia, and prosopagnosia (Josephs et al, 2009), whereas the left-sided variant primarily manifests as language deficits (Boone et al, 1999; Razani et al, 2001). In the present case, MRI revealed exclusively left-sided involvement, although, interestingly, in addition to language impairment, the patient also displayed severe behavioral problems (reflected in CS).

A recent study reported that the brain lesions underlying CS are located in the right frontal cortex and the left retrosplenial cortex, which are associated with belief evaluation and familiarity, respectively (Darby et al, 2017). Therefore, in our patient's case, selective left temporal atrophy may not by itself have been responsible for the CS; it is possible that abnormalities in the neuronal networks within this region may have played a pathogenic role in the clinical phenotype. Interestingly, other authors have also proposed that CS occurs due to a lack of communication between the sensory and limbic systems (Ramachandran, 1998).

Harciarek and Kertesz (2008) studied a large group of patients with neurodegenerative disorders including Alzheimer disease, FTD, Parkinson disease, and Parkinson-plus

syndromes and reported that none of the patients with a behavioral variant of FTD presented with a misidentification syndrome. Josephs (2007) reported one case with FTD and misidentification syndrome; however, the subtype of FTD was not mentioned. On the other hand, Harciarek and Kertesz (2008) reported that 8.3% of patients with semantic dementia can have misidentification syndromes. Interestingly, in the case presented here, CS was the primary clinical symptom of the underlying neurologic pathology and appeared to cause the most distress and disability compared to other symptoms, such as disinhibition and anomia.

This case clearly demonstrates that CS can be a feature of FTD, along with language and behavioral impairments. The possibility that patients with neurodegenerative disorders can display misidentification symptoms should be highlighted to clinicians. In such patients, structural (MRI) and functional (EEG and PET) tests should be performed for a definitive diagnosis. One limitation of our report, however, is that we cannot provide histological confirmation of the diagnosis of FTD. FTD subtypes are notoriously associated with different neuropathological lesions, including motor neuron disease-type inclusions, which are characteristic of motor neuron disease with FTD, and tau pathology (also present in Alzheimer disease and other taupathies) (Kertesz et al, 2005). It would be of interest to determine whether the combination of FTD with CS and other misidentification syndromes is associated with a specific neuropathological pattern.

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