The Characteristics of Capgras Syndrome in Patients Diagnosed as Probable Alzheimer Disease

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Objective: We assessed the characteristics of Capgras syndrome in patients diagnosed as probable Alzheimer disease, and compared the clinical and imaging findings with other previous reports.

Methods: We obtained medical records of patients with Capgras syndrome diagnosed as probable Alzheimer disease in three hospitals from March 2009 to March 2010. The basic characteristics, neuropsychologic tests, brain imaging data were investigated.

Results: The mean age of the patients was 74.7 years old and all of the patients experienced this syndrome in relation to a spouse. Brain magnetic resonance imaging showed mild-to-moderate global atrophy, variable hippocampal atrophy in this study. The Mini-Mental State Examination mean scores were 18.78 and clinical dementia rating scores were from 1 to 2. All of the patients took medicine of acetylcholine esterase inhibitors and two patients recovered with antipsychotic medication. Five patients had showed continuous Capgras syndrome, at the last follow-up day, for a year.

Conclusion: Capgras syndrome patients in this study who were diagnosed as probable Alzheimer disease showed similar results to the individuals from other reports. More study is needed to estimate the numbers and characteristics of patients with this syndrome.

Keywords: Capgras syndrome; Dementia; Alzheimer disease

INTRODUCTION

In 1923, Capgras first reported a case of a woman who had the delusion that close family members were replaced by look-alike imposters. He named these symptoms ‘delusion of doubles’ [1]. At this time, Capgras syndrome is well known for the transient, recurrent or sustained delusional belief that a person is replaced by an imposter or a double with a close resemblance to the original [2]. It is the selective duplication of people with whom the patient has strong emotional bonds and associative memories. These patients can consciously recognize familiar faces but cannot emotionally connect with them. Sometimes it is called the converse of prosopagnosia [3]. Initially, Capgras syndrome is assumed to be a rare phenomenon reported to be associated with psychiatric diseases, such as schizophrenia [4]. However, it has also been described in neurological diseases such as stroke, epilepsy, brain tumor and neurodegenerative diseases, such as Alzheimer disease [5] or Lewy body disease [6]. Three patients with Alzheimer disease presenting Capgras syndrome were reported in Korea [7]. We reported six cases and reviewed three previously reported cases. The aim of this report was to assess the characteristics of Capgras syndrome in patients diagnosed as probable Alzheimer disease, and to compare the clinical and imaging findings with previous other reports.

MATERIALS AND METHODS

We obtained all medical records in which the term ‘Capgras syndrome’ had been used by the clinical neurologists in three hospitals (Seoul National University Bundang Hospital, Bobath Memorial Hospital, and Hyoja Geriatric Hospital) from March 2009 to March 2010. We reviewed all the medical records of these patients to confirm true Capgras syndrome. Specifically, to be includ-
ed in this report, the description of Capgras syndrome clearly had to be a delusional belief that someone had been replaced by an imposter or a double. Patients were excluded if the description was more suggestive of a lack of facial recognition (prosopagnosia). Among patients with Capgras syndrome, we just investigated the patients diagnosed with probable Alzheimer disease, according to the National Institute of Neurological and Communicative Disorders and Stroke and the Alzheimer disease and Related Disorders Association [8]. The diagnosis was made clinically, and also supported by neuro-imaging. The disease onset in history was defined by the patient or reliable caregiver. We obtained the basic characteristics including age, sex, the duration of Alzheimer disease, person whom the patients had delusions, symptom duration of Capgras syndrome. Additionally, we investigated follow-up duration, state of symptoms (resolving or continuing), types of medications. Mini-Mental State Examination (MMSE) was administered as a brief screening test of global cognitive functioning and the functional impairment of the patient was assessed with the clinical dementia rating (CDR) scores. All of the patients had brain MRI studies at least once. The Patients with stroke, seizures, delirium, or psychosis were excluded before the onset of dementia.

RESULTS

Nine patients were reviewed, we summarized our six cases and previously three reported cases (Table 1). Five patients were women in nine patients. There were no differences between men and women in our cases. Mean age of the patients was 74.7 years old (59 to 86 years old). The time of the onset of Capgras syndrome was from 1 to 7 years after the onset of dementia. All patients had experienced this syndrome in relation to a spouse. In our six cases, one patient had the reduplicative paramnesia and two patients had multiple imposters. The brain MRI studies showed mild-to-moderate global atrophy, variable hippocampal atrophy, and mild degree T2-weighted increased signal changes in the subcortical and periventricular white matter that was believed to be nonspecific. MMSE mean scores were 18.78 and CDR scores were 1 at seven patients and 2 at two patients. Among nine patients, five patients showed continuous Capgras syndrome during follow-up period about one year. All patients took medicine of acetylcholine esterase inhibitors. Two patients recovered with antipsychotic medication, the others recovered without antipsychotics.

DISCUSSION

Capgras syndrome is classified as a delusional misidentification syndrome, a class of delusional beliefs that involves mainly people, places or objects. There are content-specific delusions predominate in neurologic patients, usually misidentification and reduplication syndromes, with beliefs that places (reduplicative paramnesia), people (Capgras or Fregoli syndrome), or events are transformed in identity or duplicated [9]. Capgras syndrome is representative delusional misidentification syndrome, and we reviewed our patients and compared with other previous reports. Capgras syndrome has been reported that women were prevalent in the psychiatric disorders [10]. But other recent reports described all misidentification syndrome (MIS) including Capgras syndrome in the patients with Alzheimer disease had similar ratios between

Table 1. The summary of nine patients with Capgras syndrome with probable Alzheimer disease

<table>
<thead>
<tr>
<th>Variable</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
<th>6</th>
<th>7</th>
<th>8</th>
<th>9</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex/Age</td>
<td>M/74</td>
<td>M/73</td>
<td>M/86</td>
<td>F/79</td>
<td>M/72</td>
<td>F/82</td>
<td>F/74</td>
<td>F/59</td>
<td>F/74</td>
</tr>
<tr>
<td>Disease duration (yr)</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>6</td>
<td>7</td>
<td>3</td>
<td>2</td>
<td>2</td>
<td>4</td>
</tr>
<tr>
<td>MMSE/CDR</td>
<td>25/1</td>
<td>16/1</td>
<td>21/1</td>
<td>15/2</td>
<td>14/2</td>
<td>15/1</td>
<td>17/1</td>
<td>18/1</td>
<td>28/1</td>
</tr>
<tr>
<td>Person/Other target</td>
<td>Wife</td>
<td>Wife</td>
<td>Wife/Daughter in law</td>
<td>Husband</td>
<td>Wife</td>
<td>Husband</td>
<td>Husband</td>
<td>Husband/ House</td>
<td>Husband/Mother</td>
</tr>
<tr>
<td>Symptom duration before diagnosis (yr or mo)</td>
<td>1 mo</td>
<td>1 mo</td>
<td>1 yr</td>
<td>1 yr</td>
<td>3 mo</td>
<td>2 yr</td>
<td>1 yr</td>
<td>3 yr</td>
<td>6 mo</td>
</tr>
<tr>
<td>Resolving or continuing (1 yr follow up)</td>
<td>Spontaneously resolved</td>
<td>Continued</td>
<td>Continued</td>
<td>Continued</td>
<td>Resolved</td>
<td>Spontaneously resolved</td>
<td>Continued</td>
<td>Continued</td>
<td>Continued</td>
</tr>
<tr>
<td>Additional medication for Capgras syndrome</td>
<td>No</td>
<td>No</td>
<td>Quetiapine 75 mg</td>
<td>No</td>
<td>Clozapine 12.5 mg</td>
<td>Quetiapine 25 mg</td>
<td>No</td>
<td>Olanzapine 2.5 mg</td>
<td>Olanzapine 2.5 mg</td>
</tr>
<tr>
<td>Brain MRI</td>
<td>Right</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>Brain PET</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
</tr>
</tbody>
</table>

Patient 1-6, six patients in this study; 7-9, previous reported Korean patients [6]. MMSE, mini-mental state examination; CDR, clinical dementia rating scores; MRI, magnetic resonance imaging; PET, positron emission tomography.
males and females [10-13]. Previous reports investigated that the mean age of Capgras syndrome in Alzheimer disease were 73.29 years old [11] or 72 years old [13]. The mean time of the onset of MIS in Alzheimer disease from the beginning of illness was 3.51 years (range, 0.4 to 11 years). Interestingly, about 27% among patients with Alzheimer disease developed at least one form of misidentification phenomenon within the first year of illness [11]. One reported it occurred at early stage of Alzheimer disease frequently [14], however, the other reported it occurred at late stage of Alzheimer disease frequently [12]. In this study, there is no sex differentiation and similar mean age ranged from early to late stage of Alzheimer disease like previous reports. Most of patients were likely to experience the syndrome in relation to a spouse. Additionally, Capgras syndrome may involve multiple imposters or have additional delusions, such as misidentification of familiar places (redundantive paramnesia) or phantom boarder phenomenon [11,12]. This study showed similar results in individuals as imposers and additional delusions. Mean MMSE scores were reported 19.76 ± 4.84 [11] or 10.4 ± 4.7 [12], our patients showed similar results, 18.78. Because there is no adjustment with age, years of education, sex, we could not compare them to other results. As regards the pharmacological management of Capgras syndrome, antipsychotic drugs are often prescribed. However, their efficacy has not been established. In this study, one patient showed spontaneous recovery without antipsychotics, and the other could not recover with antipsychotics. It has been known that Capgras syndrome is a disconnection between the temporal cortex and the limbic system. The faces are recognized in the temporal cortex and the emotion is related in the limbic system. The ventral temporal visual recognition system is intact, but disrupted connections to the limbic system (dorsal route) cause Capgras syndrome. In result, the patient knows who he or she is, but inappropriate affective responses to the faces generate this syndrome [15]. Patients with delusional misidentification syndromes usually have lesions in the right and/or bilateral hemispheres, suggesting the right-side dominance of the delusional misidentification syndrome [16]. In recent study, dementia patients with Lewy bodies had significant hypoperfusion in the bilateral fronto-temporal and parieto-occipital cortices compared to the controls, suggesting that the bilateral hemispheres might be dysfunctional in dementia with Lewy bodies with misidentifications [17]. This study has some limitations. The diagnosis of Alzheimer disease in this study is only based on clinical criteria and is not pathologically proven. Furthermore, it is difficult to analyze the associated lesions of Capgras syndrome due to a small number of patients.

In conclusion, Capgras syndrome patients were diagnosed as probable Alzheimer disease showed similar results to the individuals from other reports. There is not an accurate data of this syndrome with dementia in Korea except case reports. Further more study is needed to estimate the numbers and characteristics of patients with this syndrome.

REFERENCES