Clinical note

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Neuropsychological, clinical and social issues in two patients with capgras syndrome

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Two patients suffering Capgras Syndrome (CS) were evaluated with neuroimaging and neuropsychological tests, the results of which are compared with the existing etiopathogenic theories. To date, the etiopathogeny of the CS continues to lack satisfactory explanation. However, several holistic models have been proposed to better understand the many different theoretical proposals circulating for CS. These theories are discussed and the psychosocial consequences of aggressiveness, an aspect not frequently commented in the literature, are analyzed. Risk for aggressiveness and its negative consequences should be taken into consideration upon diagnosis of CS in order to design effective preventive measures.

Keywords:
Capgras, Delusional misidentification, Aggressiveness, Neuroimaging, Neuropsychology

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Aspectos neuropsicológicos, clínicos y sociales de dos casos de síndrome de Capgras

En este trabajo se presentan dos casos de Síndrome de Capgras (SC), a los que se les han realizado pruebas de neuroimagen y neuropsicológicas, cuyos resultados se contrastan con las teorías etiopatogénicas existentes. En la actualidad, sigue sin explicarse satisfactoriamente la etiopatogenia del SC aunque se han propuesto diversos e interesantes modelos holísticos que tratan de aprehender algunos aspectos de las diferentes perspectivas teóricas, que se discutirán. Así mismo, se analizan las consecuencias psicosociales que la agresividad ha generado en estos dos enfermos y que han sido poco reconocidas en la literatura. El SC nos debería alertar acerca del riesgo de agresividad así como de sus consecuencias desfavorables, con el fin de tomar las medidas oportunas para prevenirlas.

Palabras clave:
Capgras, Delirios de falsa identificación, Agresividad, Neuroimagen, Neuropsicología

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INTRODUCTION

The misidentification syndromes (MIS) include four basic subtypes: Capgras Syndrome (CS), Fregoli Syndrome, Intermetamorphosis Syndrome and the Subjective Doubles Syndrome.[1]

MISs may occur within the context of different nosological settings, such as schizophrenia (above all the paranoid subtype)[2, 3], mood state disorders and organic mental disorders. Cases have also been described in association to different somatic diseases, which must always be ruled out.[4]

CS, first described in 1923,[5] is the most frequent of all of them, this occurring in more than 4% of the patients with psychosis.[6, 7] The individual suffering it believes that a person, generally a relative, is replaced by an imposter who is identical to said person.

There are numerous etiopathogenic hypotheses generating this syndrome, which gives us a glimpse of the complexity underlying such a daily phenomenon as that of recognizing a familiar person.

From the cognitive theories, Ellis and Young[8] uphold that the MIS are caused by errors during information processing at different times during the facial recognition process. In CS, the feeling of familiarity that would help recognition is an additional emotion element that is absent.

CS, together with another disorder, prosopagnosia, are those which have made it possible for us to go deeper into the neuroanatomic model of facial recognition.[9, 10] Prosopagnosia is a condition characterized by alteration in facial recognition, but in which the subject can identify the persons when listening to their voices or by their clothes, and the subject does not pose the possibility of the other being an imposter. This defect is associated to abnormalities of the right hemisphere.[11] In this sense, the CS may be the "mirror image" of the prosopagnosia.[9, 12]

There are several stages in facial recognition: first, recognizing it as an object, then feeling it as something familiar. Another stage may be access to biographic and semantic information evoked by this face, the so-called "personal identity node," which can also be accessed by listening to the voice of the person.[10] However, the processing of all this information seems to be in parallel and not successive.

There would be an occipital-temporal ventral pathway to recognize the face as such, and another occipital-limbic and more dorsal one, from the cortex of visual association to the posterior-inferior parietal cortex and to the cingulate gyrus for the sensation of familiarity.[10] Thus, in prosopagnosia, the ventral pathways of explicit facial recognition would be altered. However, the dorsal pathway, responsible for emotional processing, associated to the facial recognition, would be conserved.[13] The result of this would be that in prosopagnosia, the faces would not be identified, however the emotional response associated to the familiar faces would be maintained. In CS, the contrary defect would occur: the ventral pathway would remain intact, without altering perception, but the involvement of the dorsal pathway would cause inappropriate emotional responses.[12, 14] Thus, in CS, the face is recognized, but the emotional experience of familiarity is lacking, with the conclusion that the other person is an imposter.

More recent approaches[15] only involved the ventral pathway which, when more or less harmed, would give rise to a symptom of prosopagnosia or CS. The amygdala would then be the key piece, since the emotionally significant memory would depend on it. There would be a first facial recognition to which the amygdala would add the affective component.

However, as indicated by Donoso et al.[10], in the CS, perception of the face of the relative does not seem to be necessary in order to believe that the relative has been impersonated. In fact, cases have been described, as that of a female patient with paranoid psychosis that reactivated a CS in relation to her daughter, while her daughter was living in a foreign country and only maintained telephone contact. Because there was no existing visual contact between them, the case illustrates that the CS cannot be explain by itself as an alteration of the facial processing.

Currently, therefore, the failure of exclusive facial processing, stressing the inability to integrate recognition of a familiar face with the emotional memories linked to it, in such a way that the expected familiarity sensation does not occur, is rejected.

Some authors, based on modern imaging techniques, suggest that there are abnormalities of the right hemisphere[16-18] or bilateral ones in the CS, stressing frontal, temporal and parietal involvement.[17, 19-21] Bathia[22], in this sense, recommends performing a magnetic resonance in all patients suffering CS. Neurophysiological research has provided some empirical support to theses findings.[23] Recently, Gainotti et al. have attempted to unify the neuroanatomical model with the cognitive proposals.[24]

Therefore, it seems that a delusion of false identification is a complex process that is not limited to a simple problem of facial processing, but rather that is due to a multiple dysfunction based on cognitive processes involved in the interpretation of abnormal perceptions and the formation of beliefs.[25] The complexity of this syndrome has been pointed out in recent reviews on different etiopathogenic models.[26, 27]

From a clinical point of view, a relation between the violent reactions against persons and the presence of an active psychotic disorder has been indicated in many studies, with a high incidence of serious violent acts in the delusional disorders compared with other mental disorders.[28, 29] Among mental patients, those having well-structured delusions are more frequently involved in violent acts against persons than those with chronic undifferentiated psychosis.[30] Precisely, one of the characteristics of the CS is its potential for dangerousness for third parties. This relation between the false identification syndrome (FIS) and aggressiveness has been poorly studied. Silva et al.[31] compared two groups of patients with psychosis and aggressive behaviors, one with FIS and another with another type of delusions. Those with FIS more commonly had grandiose ideas, thought disorders, generalized hostility, excitability and a previous history of violent behavior than the delusional and violent subjects but without FIS. However, those who suffered FIS less frequently attached with weapons.

It has been suggested that the degree of threat perceived by the patient from the erroneously identified person is the most important factor to determine the response of the patient to their delusion.[32] Another one of the possible predictive factors indicates is that of toxic substance usage, especially alcohol, superadded to psychosis.[33]

In the following, two clinical cases of Capgras syndrome with aggressive behaviors towards the relatives are presented.

Case 1

F is a 49-year old man who was admitted involuntarily after having been involved in a heteroaggressive episode against his mother.

The patient had no medical background of interest and had never received psychiatric treatment even though the

disease seems to have begun when he was 24 years old after the death of his father and he broke up with his significant other. At that time, he began to suffer behavior alterations: he quit work and studies, he shut himself up at home and began to show an interest for metaphysical and spiritual subjects, neglecting his personal appearance. In the months prior to his admission, on several occasions, he had violent behaviors towards his mother, so that his siblings decided to report him, and the judge issued a restraining order regarding his family home, which he broke in order to attack his mother, after which he was brought to the hospital.

During his admission in the Acute Unit, F manifested that for several months, he had the idea that his relatives have been impersonated by identical doubles, justifying the aggression on his mother because it was an imposter who had snuck into his home and he was only trying to get her out.

F is capable of detecting imposters because of several differences, basically physical. According to him, they have sharper cheeks due to cosmetic surgery and that a nodule has been implanted in their throat that distorts their voice. Furthermore, he says he has installed a security system in his room that detects, by means of electromagnetic emissions, when the persons are actors or not. The patient explains that there is a conspiracy by the mafia against him in which the actors impersonating his family participate. He denies alterations in sensory-perception. He has no disease awareness.

On admission to the ward, treatment was begun with risperidone 6 mg/day, this being increased to 9 mg/day, introducing risperidone 50 mg in depot formulation, every 14 days. His evolution has been favorable, reviewing the delusional ideation and adequately recognizing his relatives. However, significant residual symptoms are observed.

Because the restraining order regarding the home is still in force, the patient had to be discharged to a residential site, where he currently remains.

Case 2

G is a 53-year old patient who was admitted by his psychiatrist due to a psychotic episode with aggressive behaviors in his usual setting.

G does not have any medical background of interest except harmful alcohol consumption. He is diagnosed of paranoid schizophrenia, having been admitted to our center several times due to psychotic decompensation. He is living in the street and does not know any relative. Good adherence to treatment or disease awareness is lacking.

The onset of his symptoms seems to go back to when he was 24 years old. G then had self-referential and harm ideation and MIS that makes him think his father, a military man, had been replaced by another person, who he knew was not a military man. After committing multiple violent acts against his family, he left the family home, changing his place of residence to another province. Since then, he has been living in poverty. He has not seen his family for 10 years and does not allow us to contact them, since, according to him, we would only find impostors.

In addition to his partially incoherent language, focused on the delusional theme of harm, self-referential and false identification, he had auditory pseudohallucinations and phenomena of control and thought diffusion. Significant negative symptoms were added to these.

During admission, treatment was initiated with risperidone in depot formulation (75 mg/14 days), with good tolerability. The patient is becoming progressively calmer and coherent, with disappearance of the hallucinations and control phenomena and thought diffusion. The delusional ideation persists, although with greater affective distancing from it. G has accepted the need for psychiatric treatment and has also permitted us to arrange his admission to a shelter.

Table 1 shows the sociodemographic and clinical characteristics of the patients while table 2 shows the results of the complementary tests performed.

The neuropsychological evaluation was performed during his hospital admission. The performance of each patient, whenever possible, was compared with the age and/or educational level, obtaining the corresponding Z score and performance level. The tests administered and their results are shown in table 3.

CONCLUSIONS

We have presented two typical cases in persons suffering paranoid schizophrenia. In both, extensive physical and complementary examinations were performed, not finding significant organic alterations that could explain the delusional symptoms. Regarding the neuroimaging tests, even though some alterations have been found, they are non-specific.

When the results of the neuropsychological tests were analyzed, we found that the screening test (MMSE) showed an acceptable and similar result. However the specific evaluation shows the presence of a dysfunction of the attentional-frontal profile in both cases (although the cognitive profile is not identical), in agreement with the

Table 1 Sociodemographic and clinical characteristics

Características		Case 1	Case 2
Sociodemographics	Age	49	53
	Gender	♂	♂
	Studies	First Year in Law School	Degree in Sociology
	Work	No	No
	Partner	No	No
	Residence	Family home	Living in the street
Clinical factors	Age at onset of disease	25 years	24 years
	Years of evolution	24 years	29 years
	Toxics	No toxic habits	Tobacco Hashish/ Alcohol
	Delusions	Self-referential, of harm, mystics and false identification	Self-referential, of harm and false identification
	Control and Thought Diffusion Phenomenon	No	Sí
	Sensory-Perception Disorders	No	Sí
	Heteroaggressiveness		
	Physical/Verbal	Sí	Sí

Table 2 Complementary tests

	Case 1	Case 2
BRAIN MRI	Without significant alterations	Without significant alterations
BRAIN SPECT	Examination consistent with frontal hypoperfusion	Significant irregularity on the cortex that seems clearer in the right anterior frontal and left posterior parietal. Bilateral temporal hypoperfusion. Noticeably irregular nuclei of the base.
EEG	Little consistent harmful focal activity in left temporal zone	Registry of normal bioelectric activity in relation to the patient's age
Thyroid hormones	Normal	Normal

literature reviewed. From a qualitative analyses, standing out are the errors that both patients made in the verbal memory test, with a high number of intrusions during the learning and recall trials, especially in case number 2. The presence of these intrusions indicates that the verbal memory may be secondarily affected by frontal dysfunction, because of the difficulty to discriminate the "target" stimuli, belonging to the test and presented externally from the stimuli that were self-generated and recovered from the internal global "storage." The *familiarity* that some words or others should arouse during the test is therefore found to be altered and gives rise to "false memories" (in this regards, consult, for example, the review of Parkin[14]).

From the Ellis and Young[8] model presented in the introduction, we can conclude that these patients have an acceptable ability to discriminate faces and the ventral pathway, responsible for processing explicit recognition, would be globally preserved. One methodological limitation of our study was the impossibility to present photographs of their relatives to the patients. We could not evaluate the recognition and sensation of familiarity that these

Table 3 Neurological tests administered and results[1]

	Case 1	Case 2
MMSE (Mini-Mental State Examination).	32/35	32/35
Direct Digit Span WAIS-II	6 Z= 0.007 Average	5 Z= -0.692 Average
Inverse Digit Span WAIS-III	5 Z= 0.351 Average	5 Z= 0.351 Average
Barcelona Direct Mental Control Battery [2]	6 C= 95 Superior	3 C= 95 Superior
Barcelona Inverse Mental Control Battery [2]	2 C <10 Dysfunction	6 C= 95 Superior
Trail Making A (time and errors)	0.54" Z=-2.647 Significant dysfunction 0 errors	0.49" Z=-1.311 From average to mild dysfunction 0 errors
Trail Making B (time and errors)	2.29" Z= -4.623 Significant dysfunction 0 errors	2.31" Z= -3.079 Significant dysfunction 1 error
Alternate tracing of figures	Correct	Perserverative and motor programming errors
WCST: Wisconsin Card Sorting Test - No. of categories completed	0/6 C ≤ 1 Significant dysfunction Qualitatively: performed inferred with by internal and external distractors	0/6 C ≤ 1 Significant dysfunction
Phonetic Fluency (F-A-S) (correct and errors)	21 Z=-2.116 Significant dysfunction 7 errors	41 Z= -0.330 Average 1 error
Animal semantic fluency (correct and errors)	12 Z=-1.833 Mild dysfunction 1 error	27 Z= 0.44 Average-high 3 errors
Rey Auditory Verbal Learning Test		
- Immediate learning (test I)	3 Z=-2.0 Mild to important Dysf	4 Z= -1.533 Mild Dysf
- Learning (test V)	10 Z=-0.684 Average	4 Z= -3.227 Import. Dysf.
- Total words (tests I to V)	38 Z=-1.068 Average-low	24 Z= -2.947 Import. Dysf.
- Long term memory	7 Z=-0.727 Average to middle-low	4 Z= - 1.566 Mild Dysf
- Long term recognition	10 Z=-0.846 Average-low	6 Z=-2.0 Mild to Import Dysf.
- False positive recognition A+B	8	5
- Error in tests I to VII	21 (12 intrusions, 9 repetitions)	24 (22 intrusions, 2 repetitions)
Clock-Order	8.5/10 Adequate Qualitatively: peculiar performance	10/10 Adequate
Clock-Copy	9/10 Adequate	8/10 Borderline
Digit coding WAIS-III (correct and errors)	57 Pe=9/19 Average 0 errors	29 Pe= 5/19 Mild Dsyf. 1 error
Barcelona Face matching battery		
- Correct	5 C< 10* Dysfunction	5 C 20 Average-low
- Errors	1/6 errors	1/6 errors
- With time	10 C < 10** Import. Dysfunction.	14 C 50 Average
Identification of faces/objects/emotions. Own elaboration battery		
- Inverted faces	2/7 errors	1/7 errors
- Non-inverted faced	1/7 errors	2/7 errors
- Masked pictures	1/28 errors	2/28 errors
- Unmasked pictures	1/28 errors	2/28 errors
- Emotional expression – screening: surprises, happiness, neutral	1/6 errors Qualitatively: does not confabulate	4/6 errors Qualitatively: confabulates

[1] The direct score, Z score and corresponding level of performance are shown (in some cases, the C "percentile" or the "Pe" Score scale are shown.
[2] Time measured with stopwatch is shown (performance is similar without using a stopwatch for the time)

photographs could provoke, therefore limiting the analysis of the dorsal route.

Finally, although it must be confirmed in later studies improving the methodology, we have found signs that the analysis of the emotional expression of unfamiliar faces could be altered in the CS and could be correlated with a confabulatory tendency. Alteration of the recognition of the emotional expression in this syndrome is consistent with the proposal of Breen et al.[15] on the participation of the amygdala in facial recognition.

These results make it possible to continue adding to the phenomenological discussion mentioned in the introduction regarding the Capgras *delusion*. As we have seen, Dietl et al.[25] have described a case in which the picture was precipitated in the absence of a relative, only reactivating the (false) memory of this person. The visual stimulation (having recently "seen" the relative) does not seem to be essential. However, on the other hand, it is essential that the memory regarding this person is altered (since the sensation of familiarity fails), and a *delusional belief* is reactivated at the current moment in absence of perceptual alterations.

The fact that, under certain circumstances, the alterations in recall may precipitate a CS (or be precipitating it, as occurs in Case 2), without being accompanied by important perceptual alterations, opens up the line for future research. Although the experimental tests that we have used have some limitations, the data presented herein invite us to propose a neuropsychological hypothesis. An example is, regarding the links between frontal dysfunction, facial expression recognition deficit and the associated confabulatory tendency (see table 3), in presence of a conserved perceptual ability. In addition, in relationship to the traditional memory tests, it could be questioned if the frontal dsyfunction, altered recall of familiar persons and the appearance of errors (intrusions) in the formal evaluation of memory are significantly related. It would be desirable for future investigations on the CS to include neuropsychological protocols that make it possible to clarify these questions, contributing to the integration of cognitive and psychopathological features making up this clinical condition.

On the other hand, from the clinical point of view, although the association of the MIS with violence behaviors and even homicides has been described, not much is known about their psychosocial consequences: injuries to third parties, distancing from the family home, institutionalization, marginality, poverty, etc. in the two cases presented by us, we have been able to observe a combination of the situations described, it being possible to see that all of them affect the course and prognoses of both pictures. We know that, to a large degree, the course and prognoses of patients affected by CS are related closely with the resolution or effective treatment of the underlying disorder in which it develops. Therefore, antipsychotic treatment was administered in both cases.

However, this is not sufficient. With early detection of MIS by the mental health care professionals, and being aware of the greater risk these patients have of committing violent acts, we would be capable of preventing many aggressions and of avoiding the negative psychosocial consequences for a favorable evolution of the disease.

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