An unusual delusion of duplication in a patient affected by Dementia with Lewy bodies

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Abstract

Background: Dementia with Lewy bodies (DLB) is the second most frequent diagnosis of progressive degenerative dementia in older people. Delusions are common features in DLB and, among them, Capgras syndrome represents the most frequent disturbance, characterized by the recurrent and transient belief that a familiar person, often a close family member or caregiver, has been replaced by an identical-looking imposter. However, other delusional conditions near to misidentification syndromes can occur in DLB patients and may represent a major psychiatric disorder, although rarely studied systematically.

Case presentation: We reported on a female patient affected by DLB who presented with an unusual delusion of duplication. Referring to the female professional caregiver engaged by her relatives for her care, the patient constantly described the presence of two different female persons, with a disorder framed in the context of a delusion of duplication.

A brain 99Tc-hexamethylpropyleneamineoxime SPECT was performed showing moderate hypoperfusion in both occipital lobes, and associated with marked decreased perfusion in parieto-fronto-temporal lobes bilaterally.

Conclusions: An occipital hypoperfusion was identified, although in association with a marked global decrease of perfusion in the remaining lobes. The role of posterior lobes is certainly important in all misidentification syndromes where a natural dissociation between recognition and identification is present. Moreover, the concomitant presence of severe attentional and executive deficits evocative for a frontal syndrome and the marked global decrease of perfusion in the remaining lobes at the SPECT scan also suggest a possible dysfunction in an abnormal connectivity between anterior and posterior areas.

Keywords: Dementia with Lewy bodies, Delusion of duplication, Misidentification syndromes, Capgras syndrome

Background

Dementia with Lewy bodies (DLB) is a progressive degenerative dementia, with core features characterized by fluctuating cognitive symptoms with pronounced variations in attention and alertness, recurrent visual hallucinations and associated features of parkinsonism [1], which is recognized as the second most common form of severe cognitive impairment in older people [2].

Delusions are common features in DLB, and patients affected by this condition are firmly hold on their false beliefs also in the presence of strong contradictory evidence. Among delusional disturbances in DLB, Capgras syndrome represents the most frequent disorder affecting approximately 17% of DLB patients and is characterized by the recurrent and transient belief that a familiar person, often a close family member or caregiver, has been replaced by an identical-looking imposter [3]. Capgras syndrome belongs to delusional misidentification syndromes (see Additional file 1: Table S1), among which two major groups were proposed, based on content-specific misidentifications [4, 5]: the Capgras type (also including...
the Fregoli delusion, a delusional belief that one or more familiar persons, usually persecutors following the patient, are masquerading as several other people) and the Clonal Pluralization type. In the last, patients firmly believe that multiple exact copies of places, objects, self or others exist, such as in the misidentification of reflection, in the reduplicative paramnesia, and in the clonal pluralization of the self [6–8]. However, other delusional conditions near to misidentification syndromes can occur in several patients with DLB and may represent a major psychiatric disorders, although rarely studied systematically. Here we reported on a female patient affected by DLB who presented with an unusual delusion of duplication.

Case presentation

A 77-year-old woman had a history of DLB since 2013 when she was 75. She has a primary school education and was a retired saleswoman. Previous clinical history was unremarkable, she did not assume any medication, and there was no family history of psychiatric or movement disorders. Her initial motor symptoms, started approximately six months before the first examination, were bilateral bradykinesia, followed by rigidity and mild postural instability with associated marked hypomimia and mild hypophonia. Her relatives signaled also a history of likely rapid eye movement sleep behavior (RBD) disorder. Mood and affect were normal, with the exception of mild symptoms of apathy described mainly as a lack of motivation. She was initially treated with levodopa/carbidopa 400/100 mg daily with good initial improvement of parkinsonian symptoms. In agreement with the patient, no medications were introduced to treat RBD symptoms, which were considered not significant by the same patient.

However, three months later, she noticed a moderate increase of her motor symptoms and selegiline 10 mg daily was added. After another month, we were contacted by her relatives because she has started to present a significant cognitive impairment with fluctuating episodes of confusion and visual hallucinations, and, although less marked, with delusional jealousy and persecutory ideas, sometimes with behavioral problems (aggressiveness). Brain MRI was normal and without significant atrophy. Her dementia work-up did not reveal any reversible causes, while results of laboratory tests were normal. Although clinical feature were not suggestive for prion disease or epilepsy, an EEG was done and was normal.

A neuropsychological evaluation was done (Table 1) which showed a moderate-severe cognitive impairment mainly characterized by severe attention and executive deficits and frontal syndrome, with associated severe deficit of visual-spatial functions. Rey Copy and the Clock drawing tests are reported in Fig. 1. Moderate/severe impairment in long-term verbal and visual-spatial memory recall was registered with deficit of fluency for semantic categories (mainly due to attentional fluctuations). A brain 99Tc-hexamethylpropyleneamineoxime single-photon emission computed tomography (SPECT) was performed showing moderate hypoperfusion in both occipital lobes, and associated with marked decreased perfusion in parieto-fronto-temporal lobes bilaterally (Fig. 2). At this time, she was diagnosed as DLB.

Thus, delusional jealousy and persecutory ideas were treated with the simultaneous suspension of selegiline and the introduction of quetiapine 12.5 mg/daily. In the following two weeks, both disappearance of these delusions and the normalization of behavioral problems were observed, although a moderate impairment of bradkynesia and rigidity was noted.

However, after four months, her family members reported the appearance of an unusual delusion, characterized by the fact that the patient, referring to the female professional caregiver engaged by her relatives for her care, constantly described the presence of two different female persons, sometimes with different physical characteristics, sometimes similar, with a disorder framed in the context of a delusion of duplication. It was reported, in fact, that the patient always talked about the “two ladies”, despite the complaints of family members who indicated the obvious presence of a single person, without substantial criticism of the delusional disorder by the patient. Also in the absence of the professional caregiver, the aberrant conviction of the existence of two different ladies was constantly present, while the caregiver was never recognized as a unique person by the patient. In this respect, during the conversation the patient said: “Today I have arrived to the hospital with my son, without the two ladies who usually take care of me.”, “They are nice people, even if they do not speak much. I do not remember their names, but both speak with a foreign accent”. When we asked if they were both with blond hair, her response was: “I do not remember exactly the color of their hair, but it seems to be of a different color”.

Thus, levodopa was decreased to 200 mg/daily, with no disappearance of the delusion of duplication, but with a clear worsening of parkinsonism. At a follow-up visit, two months later, with the same professional caregiver present during the interview, the patient was referring to her as if it actually were two people. The delusional disorder of duplication was not present in relation to other people or other patient’s relatives. No visual or auditory hallucinations were referred.

Discussion

Here, we have described a particular delusion of duplication quite different both to the classical Capgras syndrome described in DLB and to the typical clonal pluralization syndromes. In fact, the delusion was not characterized by
the patient’s belief that her professional caregiver was
been replaced by an identical imposter, but, on the con-
trary, by the duplication of the same person. Further-
more, in Capgras syndrome, the imposter commonly
has features that are very similar to those of the original
person, although subtle physical differences are used to
differentiate the original person from the imposter. In
our case, according the description made by the patient
herself, the core of the delusional idea was not charac-
terized by the perception of an imposter, but was
simply described as a phenomenon of reduplication.

This delusion was not simply an unusual psychotic
symptom driven uniquely by medication, because an
important reduction of levodopa/carbidopa dosage did
not change its clinical presentation, while a total
suspension of dopaminergic treatment was impossible
for the worsening of parkinsonian symptoms.

Another striking difference with Capgras syndrome is
represented by the person involved in the appearance of
this delusion of duplication. In fact, while in Capgras
syndrome, patients report that one or more well-known
persons (usually family members) have been replaced by
substitutes [4], it is interesting to note that this delusion
of duplication was referred only to the professional

| Table 1 Neuropsychological findings of the subject at evaluation |
|-----------------------------|-----------------------------|-----------------------------|
| Test                        | Raw score | Adjusted score and/or (Cut-off) | Equivalent score and/or classification |
| MMSE                        | 18/30     | 17.7 (>24) | Impaired |
| ADL                         | 3/6       | Impaired   |
| IADL                        | 3/6       | Impaired   |
| Frontal assessment battery  | 10/18     | 11.5 (>13.4) | 0 |
| Stroop Test                 |           |           |
| Time interference           | 93.5      | 80.25 (<36.91) | 0 |
| Error interference          | 15        | 13 (<4.23)  | 0 |
| Attentional matrices        | 25        | 30.75 (>31) | 0 |
| Trail making test           | NP        | Impaired   |
| Digit span forward          | 4         | 4.65 (4.42) | 2 - Normal |
| Digit span backward         | 3         | 3.77 (2.66) | 2 - Normal |
| Rey Auditory Verbal Learning Test | | |
| Learning                    | 27/75     | 37 (>28.53) | 3 - Normal |
| Recall                      | 0/15      | Impaired   |
| Recognition (False recognition) | 15/15 (6) | Impaired   |
| Test of Corsi               |           |           |
| Direct                      | 2         | 2.68 (>3.46) | 0 - Impaired |
| Forward                     | 3         | 3.42 (>3.08) | 2 - Normal |
| Phonological verbal fluency | 31        | 40 (>16)   | 4 - Normal |
| Semantic verbal fluency     | 10        | 20 (>25)   | 0 - Impaired |
| Modified Card Sorting Test (MCST) | NP | Impaired   |
| Rey figure (Copy)           | 0         | (>23.74) | 0 - Impaired |
| Immediate recall            | 1         | 8.6 (>6.44) | 1 -At lower limit |
| Delay recall                | 0         | (>6.33)   | 0 - Impaired |
| Simple Figure Copy          | 3         | 4.3 (>7.18) | 0 - Impaired |
| Clock Test                  | 0         | (>3)      | Impaired |

Legend: Raw score, score test; Adjusted score: obtained by subtracting or adding the contribution of patient’s age and education; Equivalent score: adjusted scores converted to a five-point interval scale, ranging from 0 to 4 equivalent scores. Score 0 was equal or lower than the outer tolerance limit (5%), NP, not performable.
caregiver and not to family members. In this context, it should be considered as the caregiver is at the same time both the nearest and the more extraneous person. On the other hand, the clonal pluralization syndromes are characterized by the false belief of the existence of an identical copy (clone) of the patient’s self or of other individuals; in our case, the patient believed that there was another caregiver in addition to that real, but she did not refer to her as an exact copy, rather than as she was a different individual. This particular form of reduplicative delusion could explain why the patient did not show persecutory ideas neither toward the real caregiver nor the “other” caregiver: simply, she believed to have two caregivers.

This phenomenon of reduplication appears very interesting because its similarity to a type of hallucinatory disorder raises the question on its clear operating framework between these psychiatric conditions. However, the previous presence of other delusional disturbances and the fact that this belief, with a duration longer than two months, was fixed and not amenable to change in light of conflicting evidence, in addition to the absence of any other hallucinatory problem, depose with greater evidence to a classification within the spectrum of delusions, according to the DSM 5 [9]. In any case, it should be remembered as a previous study found a strong relationship between Capgras syndrome and visual hallucinations [3].

Another issue concerns the possible neurobiological mechanisms that may contribute to the appearance of this delusion of duplication. In fact, several studies have addressed their attention to Capgras syndrome, but other misidentification syndromes have been scarcely studied. With regard to Capgras syndrome, proposed alterations include the presence of impairment in facial processing [10, 11], dysfunction of working memory [12], altered connectivity among associative areas and limbic/paralimbic structures [13], bilateral dysfunction of fronto-temporal connectivity [14, 15], and right hemispheric hypo function [16, 17].

In our patient with this peculiar delusion of duplication, an occipital hypoperfusion was identified, although in association with a marked global decrease of perfusion in the remaining lobes. In this context, it should be kept in mind as a deficiency in occipital hypoperfusion is more frequently seen in DLB with respect to other dementia such as Alzheimer disease [18]. The role of posterior lobes is certainly important in all misidentification syndromes where a natural dissociation between recognition and identification is present and is confirmed by the findings of severe deficit of visual-spatial functions observed at the neuropsychological evaluation and typical of a posterior cerebral areas dysfunction. However, the concomitant presence of severe attentional and executive deficits evocative for a frontal syndrome and the marked global decrease of perfusion in the remaining lobes at the SPECT scan also suggest a possible dysfunction in an abnormal connectivity between anterior and posterior areas.

Moreover, the presence of a significant impairment in visual-spatial functions and in visual-spatial memory recall is in agreement with previous neuropsychological findings on delusional misidentifications reporting a low efficiency in the complex visuospatial organization tasks and in non-verbal memory [19]. In this context, there is a clear evidence for impaired visuoperceptual functions in the appearance of misidentificative psychotic symptoms in patients affected by Alzheimer’s disease [20].

**Conclusions**

In conclusion, we have described an unusual delusion of duplication in a patient affected by LBD. Neural correlates of this peculiar delusion of duplication in LBD still remain largely unclear, and future researches are required to better identify possible neurobiological mechanisms that may contribute to the appearance of this peculiar type of delusion.

**Additional file**

Additional file 1: Table delusions of misidentification. Table summarizing the most common delusions of misidentification and related delusions. (DOCX 483 kb)
Abbreviations
DLB: Dementia with Lewy bodies; MRI: Magnetic resonance imaging; RBD: Rapid eye movement sleep behavior; SPECT: Single-photon emission computed tomography

Acknowledgements
None.

Funding
No funding was obtained for this study.

Availability of data and materials
All the relevant data of this article have been presented in the main paper of the article.

Authors’ contributions
PS contributed to the conception and design of the paper, to the acquisition, to the analysis and interpretation of data, wrote the first draft of the manuscript and was involved in its final drafting. GM contributed to the design of the paper, to the acquisition, to the analysis and interpretation of data, wrote the first draft of the manuscript and was involved in its final drafting. AC contributed to the design of the paper, wrote the first draft of the manuscript and was involved in its final drafting. MGC contributed to design of the paper, revised critically the manuscript and was involved in its final drafting. GO revised the manuscript and was involved in its final drafting. All authors read and approved the final manuscript, and were involved in the decision to submit the manuscript for publication.

Competing interests
The authors declare that they have no competing interests.

Consent for publication
Informed consent was obtained by the patient.

Ethics approval and consent to participate
Because this is a retrospective case report, ethical approval was waived. However, informed consent was obtained by the patient in order to publish the case in accordance with the declaration of Helsinki.

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Received: 4 July 2016 Accepted: 17 March 2017
Published online: 19 April 2017

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