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Case Report OMICS International

Delusional Misidentification Syndrome with Response to Donepezil and Behavioral Intervention in a Patient with Dementia

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Abstract

Introduction: Delusional Misidentification Syndrome (DMS) encompasses a group of disorders in which a person persistently believes the identity of people, places, or objects are altered. Historically, described in psychotic disorders, DMS prevalence is 15.8% in Alzheimer's disease (AD) and 16.6% in dementia with Lewy bodies (DLB). We present a case of DMS in a patient with dementia that incorporates elements of mirrored self-misidentification and phantom boarder syndrome and therapeutic response to a combination of a behavioral intervention and donepezil.

Case: 75-year-old white female presented with a four months history of DMS and visual hallucinations. Patient perceived her own reflection in picture glass as an older lady who was trying to steal her "boyfriends." Her "boyfriends" were three pictures of soldiers in her apartment. MMSE was 27/30 (WORLD) and 23/30 (Serial 7s). MRI showed biparietal and right hippocampal atrophy. NPT showed impaired language, spatial abilities, memory, and executive control. She scored <1 percentile on category word fluency, judgement of line orientation, raw complex figures and Beery VMI. Patient was diagnosed with probable AD, using NINCDS-ADRDA and findings on neuropsychological testing (NPT) and MRI. DLB was excluded using McKeith's criteria. After a failed trial of risperidone, she received donepezil and family was instructed to remove photographs. MMSE stable with resolution of the mirrored selfmisidentification at 4 months follow up.

Conclusion: Patient's poor response to risperidone is consistent with previous studies suggesting limitations of antipsychotic treatment for psychotic symptoms in AD. Removal of potential symptom trigger along with an acetylcholinesterase inhibitor resulted in remission for up to 4 months. The potentiating effect of donepezil on the cholinergic component of the visuo-amygdaloid pathway/dorsal visual pathway may account for these changes.

Keywords: Delusional misidentification syndrome; Dementia; Poserior cortical atrophy; Alzheimer's disease; Cholinesterase inhibitors

Introduction

Delusional misidentification syndromes (DMS) encompass a group of delusional disorders in which patients persistently believe that people, places, objects, or events have somehow been altered. They are usually associated with schizophrenia spectrum, but also reported with organic disorders like dementias, epilepsy, traumatic brain injury, sub-arachnoid hemorrhage [1] and stroke [2]. The prevalence in Alzheimer's dementia (AD) and dementia with Lewy Bodies (DLB) is estimated at 16%, with the prevalence somewhat higher in DLB [3]. We present a case that incorporated elements of mirrored self-misidentification (delusion that one's reflection in the mirror is another person) and phantom boarder (delusion that someone uninvited is living in one's house), resolved with combination of behavioral intervention and donepezil.

Case Report

Ms. H was a 75 year old female who presented with a four months history of visual hallucinations (VH) and DMS. She reported living in her apartment with 3 boyfriends and recently noticed an older woman trying to steal the boyfriends from her. Ms. H called the police several times because of the older woman. According to her daughter, the "boyfriends" were photographs of three soldiers, pictured from waist up (no legs visible in the pictures). Ms. H believed that those men lost their legs in a war. She carried those pictures around, believing they men couldn't walk and needed her help. She acknowledged that they were photographs but believed that they had material bodies with certain needs. One of the pictures were of her late husband in his youth but patient didn't acknowledge it. Ms. H also reported that she could see the reflection of the older woman trying to climb onto her boyfriends which was in fact her own reflection in the glass of picture frames. She claimed that old woman in reflection always wore the same jewelry and clothes as her which she must had stolen from her as well.

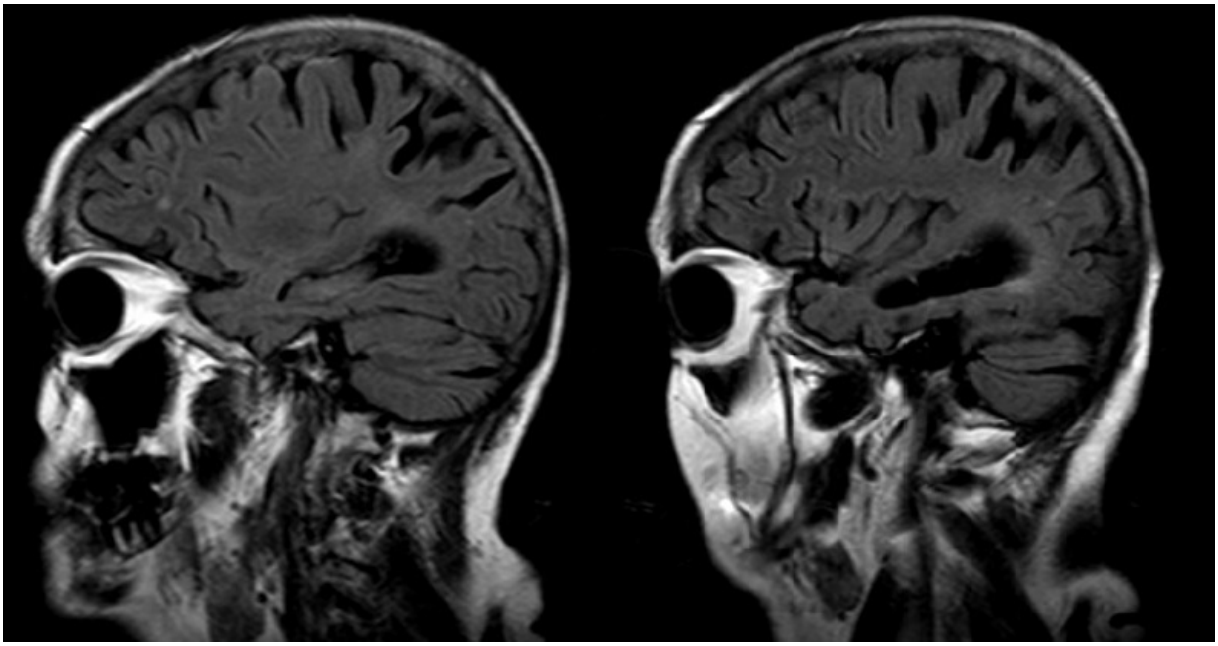

Figure 1: T1 sagittal views demonstrating bi-parietal atrophy.

She scored 27 on MMSE WORLD and 23 with Serial 7s. Neuropsychological testing (NPT) showed impaired language, spatial abilities, immediate memory and executive control (Table 1). Dalyed recall was borderline abnormal. MRI showed bi-parietal, occipital and right hippocampal atrophy (Figures 1-3). She presented to us after a failed trial of risperidone. Given that her presentation and workup was more consistent with AD, she was started on donepezil 5 mg, titrated to 10 mg/day. The family was also instructed to put the photographs away to remove the presumed trigger.

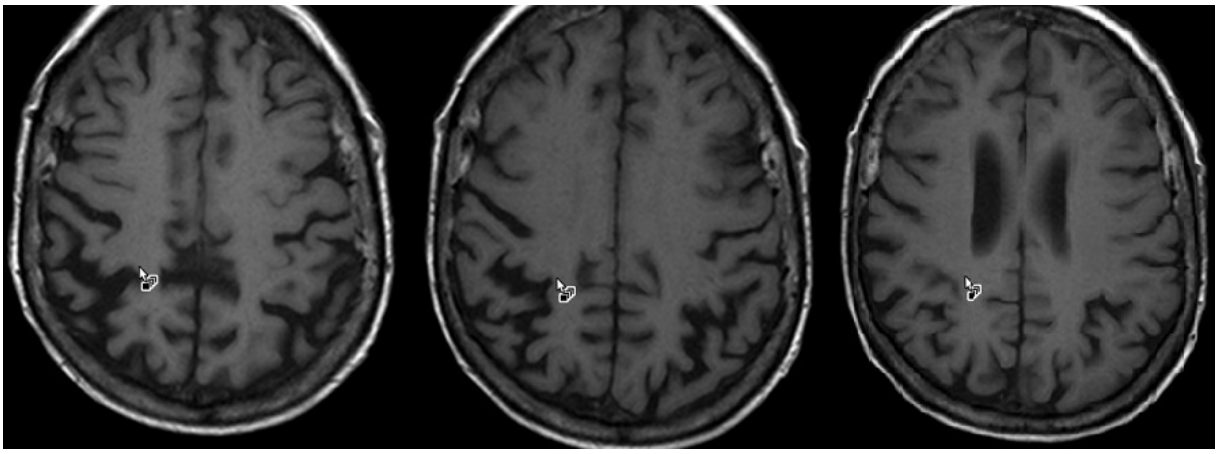

Figure 2: T1 axial views demonstrating bi-parietal atrophy.

At 4 months follow-up, she demonstrated marked improvement stating, "That old lady is finally gone!" The photographs were put back after 2 weeks, as per her request, without recurrence of DMS. She still acknowledged the presence of her soldier friends but it was not distressful. Her MMSE remained stable.

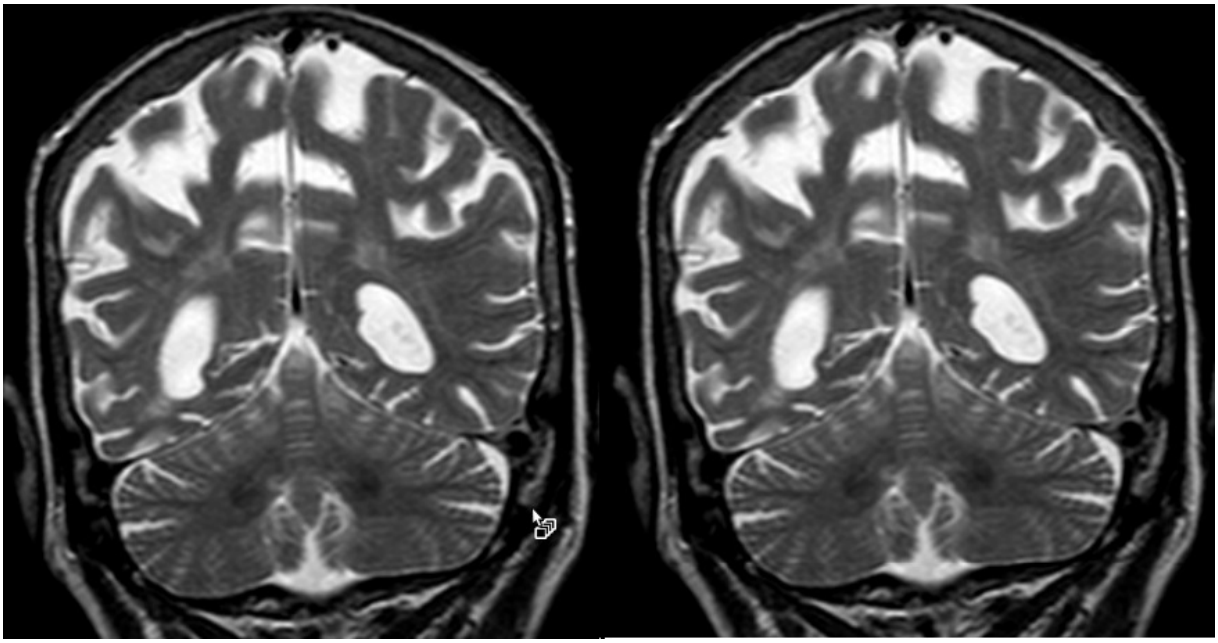

Figure 3: T2 coronal views demonstrating the loss of brain volume.

Discussion

Patient was diagnosed with probable AD, using NINCDS-ADRDA which was further supported on NPT and MRI [4]. Patient did not meet the Mckeith's Criteria for probable DLB as no cognitive fluctuations, parkinsonism or REM sleep behavioral disorder were observeded [5]. Possible DLB still remained in differential given prominent visuospatial deficits, visual hallucinations (VH), exectutive dysfunction and the fact that AD and DLB frequently overlap. Further diagnositic modalities including transcranial magnetic stimulation study Posterior cortical atrophy (PCA) variant was also a consideration given prominent space and object percepion deficits, constructional dysparaxia seen on NPT and marked parieto-occipital and temporal atrophy seen on MRI. However, our patient lacked other frequently seen PCA features including Balint's syndrome (simultanagnosia, oculomotor apraxia, optic ataxia) or Gerstmann's syndrome (acalculia, agraphia, finger agnosia, left/right disorientation) [6].

Cognitive Test	Description	(Raw scores & Interpretation)	Percentiles adjusted for age	Percentiles adjusted for age, sex & Education	Interpretation
MMSE	Used as baseline to assess general cognitive ability	World 27, serial seven 23	-	-	-
NAART IQ	Estimated premorbid intelligence	110	-	-	High Average
Boston naming test	Confrontational word retrieval	46	5	-	Borderline
CIFA letter word fluency	Language and academic skills	26	42	58	Normal
CIFA category word fluency	Language and academic skills	21	1	1	Defect
Beery Developmental test of visual-motor integration	Visual-motor construction & Integration	18	1	-	Defect
Clock drawing test	Visual-motor construction, visual-motor planning and organization	-	-	-	-

Benton judgement of line orientaion test	Line and angle discrimintaion	0	<1	-	Defect
Benton facial recognition test	Facial recgonition, Capgras-like visual decomposition of face	17	-	-	Defect
Rey complex figure copy	Visual-motor construction and Organization	5	<1	<1	Defect
Hopkins verbal learnig test (trials 1-3)	Verbal short term memory	14	8	14	Borderline
Hopkins verbal learning test (delayed recall)	delayed verbal memory and recall (delay 20-25 mins)	6	24	34	Normal
Hopkins verbal learning test retention (%)	Verbal memory retention	100	84	86	Normal
Hopkins delayed recognition index	-	6	2	4	Borderline
WMS-R logical memory trial-1	Logical learning and memory	11	2	2	Defect
WMS-R logical memory trial-2	Delayed logical memory and recall (delay: 20-25 mins)	9	8	12	Borderline
Wms-R Visual Reproduction Trial-1	Visual learning and memory	22	18	21	Normal
Wms-R Visual Reproduction Trial-2	Delayed visual memory and recall (delay: 20-25 mins)	3	10	12	Borderline
Wais iii digit span forward	Short-term memory, working memory and recall	5	16	18	Normal
Wais iii digit span backward	Short-term memory, working memory and recall	3	5	4	Borderline
Wais iii total digit span (forward + backward)	-	8	7	7	Borderline
Trail making test-a (secs to complete)	Visual attention and executive control	49	34	38	Normal
Trial making test-b (secs to complete)	Visual attention and executive control	d/c	-	-	Defect
DKEFS sorting test - correct sorts	Verbal and spatial concept formation and executive control	2	2	-	Defect
DKEFS sorting test - description score	Verbal and spatial concept formation and executive control	6	2	-	Defect
DKEFS sorting test - repeated sorts	Verbal and spatial concept formation and executive control	1	50	-	Normal
Geriatric depression scale	-	7	-	-	Mild
Purpose in life scale	-	3	-	-	Average
Neuropsychiatric inventory	-	32	-	-	Moderate
Lawton brody adl/iadl, Self report	Self-reported activities of daily living	2	8	7	Borderline

Lawton brody adl/iadl, infromant report	Informant reported activities of daily living	4	3	3	Borderline

Table 1: Neuropsychiatric test results.

Neuroanatomical correlations for VH and DMS include right mesiotemporal atrophy or involvement of ventral and dorsal visual streams. Ismail Z., in his review reported that delusions in AD are predominantly a right hemispheric phenomenon with medial temporal lobes specifically involved in DMS [7]. A disconnection from right temporo-limbic regions may result into lack of insight into the visual stimuli and hence the wrong emotional responses attached to them.

Study	# of Patients	DMSs Type	Underlying Pathology	Treatment	Outcome
Gil-Ruiz, et al.	1	Mirror Sign	Probable DLB	Behavioral Intervention	Mirror Sign replaced by Imposters/ another DMS
Oulis P, et al.	1	Capgras	Paranoid Schizophrenia complicated by Vascular Dementia	Donepezil, Olanzapine	Resolved
Peritogiannis V, et al.	1	Capgras	Vascular Dementia	Donepezil	Resolved
Reimers, et al.	2	Capgras	DLB	Donepezil	Resolved
Roane, et al.	3	Capgras, Reduplication	PDD	Clozapine	Resolved in 2/3 cases
Shiotsuki	1	Capgras	PDD	Failed trial with Donepezil, Increased dose of levodopa/ carbidopa	
Sutton	1	Capgras	Dementia NOS	Quetiapine	Improved
Marantz	1	Capgras	DLB		Spontaneous resolution with worsening of the disease
Pagonabarraga J, et al.	5		PDD	A) Quetiapine in 3 patients. B) Ziprasidone in 1 patients C) Riviastigmine initiated in 1 patient 4/5 patients were already on it	Improvement with Quetiapine in 2/3 patients. Improvement with Ziprasidone 1/1 patient. Marked improvement was observed in the one in which therapy was initiated with rivastigmine

Table 2: Cases reporting successful management of DMS in neurodegenerative disorders

The involvement of ventral (occipito-temporal) and dorsal (occipito-parietal) visual streams can also explain these symptoms. These pathways, through connections to the limbic system, are involved in processing facial recognition and generating affective responses, based on the familiarity to the stimulus. Any disruption along these pathways can lead to a false interpretation of the visual stimulus [8]. Reeves et al. suggested a role of ventral visual pathway based on the poor performance on rapid visual processing tests that have documented association with these pathways [9]. Diffusion tensor imaging studies also showed the involvement of an indirect occipito-temporal pathway (inferior longitudinal fasciculus) [10]. Furthermore, post-mortem studies have shown increased tax burden along these pathways as well as in right hippocampal and parahippocampal regions [11-13].

Several studies have described the role of cholinesterase inhibitors (ChEI) in managing neuropsychiatric symptoms of AD, DLB, and Parkinon's disease dementia (PDD) [14-16]. ChEI have also been reported to be beneficial in managining DMS in cases of DLB [17], vascular dementia [18] and PDD [19]. In addition, antipsychotics and simple behavioral interventions have been used for DMS with some success, although antipsychotics are not without risk and side effects in such patients [20,21] (see Table 2). In our case, a combination of behavioral modification with donepezil was effective. Further double-blind studies are needed to evaluate the role of ChEI in treating DMS associated with neurodegeneration.

References

1. Cipriani G, Vedovello M, Ulivi M, Lucetti C, Di Fiorino A, et al. (2013) Delusional Misidentification Syndromes and Dementia A Border Zone Between Neurology and Psychiatry. Am J Alzheimer's Disease Dementias 28: 671-678.
2. Villarejo A, Martin VP, Moreno-Ramos T, Camacho-Salas A, Porta-Etessam J, et al. (2011) Mirrored-self misidentification in a patient without dementia: evidence for right hemispheric and bifrontal damage. Neurocase 17: 276-284.
3. Harciarek M, Kertesz A (2008) The prevalence of misidentification syndromes in neurodegenerative diseases. Alzheimer Disease & Associated Disorders 22: 163-169.

4. McKhann G, Drachman D, Folstein M, Katzman R, Price D, et al. (1984) Clinical diagnosis of Alzheimer's disease Report of the NINCDS-ADRDA Work Group* under the auspices of Department of Health and Human Services Task Force on Alzheimer's Disease. Neurology 34: 939.
5. McKeith IG, Dickson DW, Lowe J, Emre M, O'Brien JT, et al. (2005) Diagnosis and management of dementia with Lewy bodies: third report of the DLB Consortium. Neurology 65: 1863-1872.
6. Crutch SJ, Schott JM, Rabinovici GD, Murray M, Snowden JS, et al. Consensus classification of posterior cortical atrophy. Alzheimer's & Dementia.
7. Ismail Z, Nguyen MQ, Fischer CE, Schweizer TA, Mulsant BH (2012) Neuroimaging of delusions in Alzheimer's disease. Psychiatry Res 202: 89-95.
8. Ellis HD, Young AW (1990) Accounting for delusional misidentifications. The British J Psychiatry 157: 239-248.
9. Reeves SJ, Clark-Papasavas C, Gould RL, Ffytche D, Howard RJ (2015) Cognitive phenotype of psychotic symptoms in Alzheimer's disease: evidence for impaired visuoperceptual function in the misidentification subtype. Int J Geriatr Psychiatry 30: 1147-1155.
10. Kantarci K, Avula R, Senjem M, Samikoglu A, Zhang B, et al. (2010) Dementia with Lewy bodies and Alzheimer disease Neurodegenerative patterns characterized by DTI. Neurology 74: 1814-1821.
11. Ferman TJ, Arvanitakis Z, Fujishiro H, Duara R, Parfitt F, et al. (2013) Pathology and temporal onset of visual hallucinations, misperceptions and family misidentification distinguishes dementia with Lewy bodies from Alzheimer's disease. Parkinsonism Relat Disord 19: 227-231.
12. Förstl H, Burns A, Levy R, Cairns N (1994) Neuropathological correlates of psychotic phenomena in confirmed Alzheimer's disease. British J Psychiatry 165: 53.
13. Mukaetova-Ladinska E, Harrington C, Roth M, Wischik C (1993) Biochemical and anatomical redistribution of tau protein in Alzheimer's disease. The Am J Pathology 143: 565.
14. Rogers SL, Farlow MR, Doody RS, Mohs R, Friedhoff LT (1998) A 24-week, double-blind, placebo-controlled trial of donepezil in patients with Alzheimer's disease. Donepezil Study Group. Neurology 50: 136-145.
15. Szigeti K, Hafeez MU (2015) Exploring the role of donepezil in dementia with Lewy bodies. Drugs of today (Barcelona, Spain: 1998) 51: 579-590.
16. Oh YS, Kim JS, Lee PH (2015) Effect of Rivastigmine on Behavioral and Psychiatric Symptoms of Parkinson's Disease Dementia. J Movement Disorders 8: 98-102.
17. Reimers K, Emmert N, Shah H, Benedict RH, Szigeti K (2014) Capgras-like visual decomposition in Lewy body dementia with therapeutic response to donepezil. Neurology Clinical Practice 4: 467-469.
18. Oulis P, Dimitrakopoulos S, Konstantakopoulos G, Tsaltas E, Kollias K (2012) Capgras delusion in paranoid schizophrenia complicated by vascular dementia. The J Neuropsychiatry and Clinical Neurosciences 24: 28.
19. Pagonabarraga J, Llebaria G, García-Sánchez C, Pascual-Sedano B, Gironell A, et al. (2008) A prospective study of delusional misidentification syndromes in Parkinson's disease with dementia. Movement Disorders 23: 443-448.
20. Roane DM, Rogers JD, Robinson JH, Feinberg TE (1998) Delusional Misidentification In Association With Parkinsonism. The J Neuropsychiatry And Clinical Neurosciences.
21. Gil-Ruiz N, Osorio RS, Cruz I, Aguera-Ortiz L, Olazaran J, et al. (2013) An effective environmental intervention for management of the 'mirror sign' in a case of probable Lewy body dementia. Neurocase 19: 1-13.