

Unusual visual illusions in Dementia with Lewy Body: A report of two clinically distinct cases

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December 23, 2024

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Data availability statement:

The data supporting this study's findings are available from the corresponding author upon reasonable request.

Funding and acknowledgments:

Funding resources are needed for this study.

Disclosure of potential competing interests:

Dr.M.Almasi reports no financial relationships with commercial interests.

Patient consent statement:

Written informed consent was obtained from the patients to publish this report anonymously by the journal's patient consent policy.

Author contributions:

Mahsa Sepahvand: Data curation, Writing original draft, writing-review & editing; Mostafa Almasi-Dooghaee: Project administration, Supervision, writing-review & editing, Babak Zamani: Conceptualization, Supervision, Kimia Kogani: Writing original draft

Key Clinical message:

In patients with parkinsonism, the presence of unusual visual illusions, such as the room tilt illusion and palinopsia, highlights the diagnosis of Dementia with Lewy Bodies. A detailed, goal-directed neurological examination enhances the clarity of diagnosis and promotes early treatment initiation.

Keywords: Dementia with Lewy Bodies, Visual illusion, Visual hallucination, room tilt illusion, palinopsia

Introduction:

Dementia with Lewy Bodies (DLB) is the second most prevalent form of dementia in older adults, behind Alzheimer’s dementia.(1) The presence of parkinsonism, visual hallucinations, cognitive fluctuations, and Rapid Eye Movement (REM) sleep disturbances hallmarks dementia with Lewy bodies (DLB). (2) The Associated visual hallucinations and illusions exhibit a high level of complexity, are well-developed, and last over time.

The room tilt illusion (RTI) or Tortopsia often occurs in individuals with vestibular system disorders, resulting in a misalignment of visual representations in the brain. (3). In the extreme form, there is a complete 180° rotation of the environment, known as the ”upside-down” illusion.(3) The ”clock drawing test” may be used to determine the existence of Tortopsia.(4) Visuospatial dysfunction due to occipitotemporal hypoperfusion may contribute to a room tilt illusion and palinopsia in patients with DLB.(5) We also propose that early aggregation of Alpha Synucleins in the vestibular nucleus of the brainstem may explain the vestibular-visual mismatch and associated illusions along with early recurrent falls in the early phase of DLB.(6)

To our knowledge, this is the first report on room tilt illusion in DLB.

Case History/Examination:

Case 1:

The first individual was a 73-year-old male from Iran who had received 16 years of formal schooling and was employed as an English instructor at a language institution. He was fluent in five languages: Persian, English, and Turkish, and had an intermediate degree of proficiency in Dutch and French. He came to our specialized dementia clinic because of slight forgetfulness during the previous two years. He has had episodic visual hallucinations for the last year. He said, ”I observe the objects on the ceiling descending towards the floor.” Postural instability has been apparent over the previous eight months. He had a mild symmetric resting tremor in his hands since 1.5 months ago. His partner saw violent activity in the second half of his sleep. She said he ”appears to be struggling with his dreams.” His medical background includes a history of hypertension, and he was prescribed Aspirin, Valsartan, Fluoxetine, Buspirone, Haloperidol, and Trihexyphenidyl. He refused any familial background of mental disorders.

The neurological examination determined that the Montreal Cognitive Assessment (MoCA) scores were 15 out of 30 during the first appointment. The MoCA sub-scores were as follows: visuospatial/executive function 1 out of 5, Naming 3 out of 3, Attention 2 out of 6, Language 1 out of 3, Abstraction 2 out of 2, Recall 0 out of 5 with Memory Index Scale (MIS) 6 out of 15, and Orientation 6 out of 6. The Clock Drawing exam has an estimated tilt of 30 degrees (Figure 1, A). Additional observations during the examination were pronounced hypokinesia, tactile-sensitive myoclonic-like motions in the upper limbs, stiffness on the left side, and retraction of both eyelids.

The electroencephalogram (EEG) revealed intermittent delta waves in the posterior alpha background. The Brain MRI showed slight enlargement of both ventricles, bilateral grade 2 medial temporal lobe atrophy (MTA), and areas of increased signal intensity in T2/FLAIR images in the cerebral white matter (Fazekas grade 2) (Figure 1, C). The cerebrospinal fluid (CSF) and blood examination revealed no notable findings.

The diagnosis of probable Dementia with Lewy body (DLB) was established based on the presence of recurring visual hallucinations, parkinsonism REM-sleep behavioral disorder, and fluctuating cognition.

Case 2:

A 75-year-old woman, who is a homemaker and has completed 12 years of schooling, was referred to the neuropsychiatric clinic because she has been experiencing memory loss for events and often asked the same questions since 1.5 years ago. She had profound sadness and frequently cried over the previous year after the passing of her spouse. The presence of RBD was detected. In the past one year ago, she has been experiencing intense visual hallucinations. For example, she sometimes perceives the presence of two children in her home and engages in conversations with them. Once, she had an unusual visual hallucination known as an after-image, referred explicitly to as Palinopsia. She saw a canine on the road, and after three hours, she reencountered it while traversing a different street, ten kilometers further. Previous medical history encompasses ischemic heart disease, hypertension, and a record of thyroid surgery. Currently, a psychiatrist has recommended a daily dosage of 200mg of Pregabalin, 25mg of Nortriptyline, and Ginkgo biloba.

The neurological examination indicated the presence of stiffness on the left side, slowness of movement in the upper limbs, and slight resting tremor. The overall MoCA score was 15, with an additional point for 12 years of schooling. The sub-scores were as follows: visuospatial/executive function 1 out of 5, Naming 2 out of 3, Attention 3 out of 6, Language 1 out of 3, Abstraction 1 out of 2, Recall 0 out of 5 (MIS= 6 out of 15), and Orientation 6 out of 6.

Investigations and treatment:

Following the discontinuation of Haloperidol and Trihexyphenidyl in case one, we started Donepezil (5mg/day) and Valporate sodium (for myoclonic-like jerks). During the one-month follow-up, the myoclonic jerks ceased, and the hallucinations were reduced. A little dosage of Levodopa was prescribed to alleviate symptoms of parkinsonism.

After nine months, the subsequent MoCA test scored 18 out of 30, with significant enhancement in the Attention sub-score (5 out of 6) and the Recall sub-score (1 out of 5, with a Modified Item Score of 6 out of 15). However, there was no change in the visuospatial/executive function score (1/5), and the Clock Drawing was more straightforward than the prior one (Figure 1, B).

In our second case, laboratory testing results, including thyroid function assessments and EEG, were within the normal range. The brain MRI revealed parietooccipital atrophy, but the medial temporal region remained intact.

The patient received a diagnosis of likely DLB and was prescribed Donepezil at a daily dosage of 10mg, along with a modest dose of Levodopa. The use of Duloxetine and quetiapine resulted in an enhancement in her mood and a reduction in hallucinations over the two months of observation. The score on the MoCA exam, eight months later, was 19.

Discussion:

We provide findings on two uncommon illusions seen in individuals diagnosed with DLB.

The first patient reported experiencing a phenomenon known as the room tilt illusion, characterized by the perception of items flowing downwards towards the floor and a distorted sense of the surrounding environment being skewed to one side. (7)

Room tilt illusion occurs when there is a mismatch between the visual and vestibular inputs that reach the visuospatial cortex. The vestibular afferents may be disrupted due to the defect in the sensory inputs (from the peripheral nervous system or spinal cord) (8), brainstem, or cerebellar lesions. (9) When evaluating patients with Room Tilt Illusion, we can categorize them as having a direct pathology in the vestibular system and its sensory afferents or in connection with the cortex. (3) In our patient, The episodic pattern of the illusion was against subjective visual vertical tilt, in which the constant environment vertical torsion

is the result of otolithic dysfunction. (10) We excluded infratentorial structural abnormalities and epileptic episodes by Brain MRI and EEG, respectively. The absence of vertigo, tinnitus, and hearing loss was against a previous vestibular pathology. To our knowledge, our case is the first reported Room Tilt Illusion in DLB. It has been reported previously in a case of Parkinson's disease and Multiple system atrophy. (3, 11) In 2009, the brain single photon emission computed tomography (SPECT) of a Parkinson-affected patient with Room tilt illusion revealed hypoperfusion in bilateral occipital cortices and the posterior part of the right intraparietal sulcus. This finding became a helpful imaging biomarker of DLB in 2024.(11, 12)

We suggest that the Room tilt illusion in DLB is the result of cortical disintegration. Therefore, it is plausible to indicate that the presence of visuospatial dysfunction early in the disease course of patients with Parkinsonism may be a diagnostic clue to the DLB.

Our second patient's primary complaint was palinopsia, a visual disorder characterized by the persistence of afterimages even after the removal of visual stimuli. Palinopsia is classified into two kinds based on its content (hallucinatory and illusionary) and the time gap between the original picture and its afterimage (immediate and delayed forms, occurring within a few minutes or hours). (13) A distinct and detailed afterimage with a high level of clarity characterizes hallucinatory palinopsia. (14) Our patient had delayed hallucinating palinopsia.

Palinopsia may occur in people with brain lesions, mainly in the Post-geniculate cortex, medications (Topiramate), seizures, psychiatric illness (schizophrenia), and migraine. (15, 16) A 2015 case report discussed a case of palinopsia due to unilateral vestibular deafferentation after a large cholesteatoma surgery. The authors explained that palinopsia occurs when the vestibular input delays mismatches of visual inputs reaching the cortex. (17) Our brain MRI findings showed no evidence of structural abnormalities. There was no record of any previous head injury. The EEG revealed no epileptic activity. The patient's medication record did not indicate the likelihood of drug-induced palinopsia. Ultimately, the patient's symptoms did not satisfy the criteria for any significant mental condition.

Research has shown that DLB is distinguished by a consistent decrease in metabolic activity in the parieto-occipital area, as seen in Brain SPECT. (5) There is a correlation between decreased blood flow on both sides of the occipitotemporal cortex, more intense hallucinations, and worse visual processing. (18) In a study conducted in 2024, palinopsia was categorized as a minor visual hallucination that occurred when a partial defect in white matter connected dorsal and ventral attention networks and visual areas.(19)

In addition to the role of occipitotemporal hypoperfusion, we propose that early aggregation of Alpha Synucleins in the vestibular nucleus of the brainstem may explain the vestibular-visual mismatch and associated illusions along with early recurrent falls in the early phase of DLB. (6, 20)

Figures:

Figure 1 . Clock Drawing test (A and B) and Brain MRI (C) of Case 1.

The position of the number on the clock is somewhat diagonal, with the number 6 (arrow) inclined roughly 30 degrees clockwise. B) The subsequent Clock Drawing assessment revealed a more streamlined clock design over nine months. (C) Axial FLAIR and Diffusion-Weighted MRI scans showed increased brightness in the subcortical white matter (Fazekas grade 2) and slight enlargement of the ventricles, with no signs of restricted diffusion.

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