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A case of progressive posterior cortical atrophy (PCA) with vivid hallucination: are some ghost tales vivid hallucinations in normal people?

Patients with Parkinson's disease (PD), diffuse Lewy body disease (DLBD), or dementia with Lewy body disease (DLB) unaccompanied by paranoia, delusions, REM sleep behaviour disorder, or an obvious sleep problem sometimes report experiencing extremely vivid visual hallucinations (VH) of ghosts or monsters.[1 2] Here we describe two VH experienced by a patient with progressive posterior cortical atrophy (PCA) which are similar to Japanese folktales and personal ghost stories.

Case report

The patient was a 60 year old man who complained that he had begun to see ghosts frequently. He noted difficulty in writing when he was 57 years old because he could not remember Japanese characters well, and this was accompanied by severe insomnia. He frequently had VH both during the day and at night. Two typical hallucinations are described below.

(1) The patient was at home when suddenly six people broke into his living room and started cooking silently. He asked them why they had come and what they were doing, but no one answered. He questioned them repeatedly and tried to tap one on the shoulder, but as soon as his hand touched the ghost's shoulder, they all vanished instantaneously.

(2) One night, when the patient went to the toilet, he found his wife lying in the corridor stabbed to death and bleeding profusely. Although it was midnight and completely dark, he could see the fresh blood coming out of her neck wound. He was astonished and tried to help her up, but all images vanished as soon as he touched her.

Neurological examination showed Bálint syndrome accompanied by dressing apraxia, constructional apraxia, agraphia, and dyscalculia. The patient's mini-mental state examination (MMSE) score was 17/30. He showed no bradykinesia, muscle rigidity, or finger tremor. He was also seen by a psychologist who found no emotional problems. MRI revealed bilateral parieto-occipital, occipital, and mild left temporal cortical atrophy (fig 1), and cerebral perfusion estimated by xenon CT perfusion showed decreased cerebral blood flow in the bilateral parietal and frontal regions. CSF showed no abnormality. Administration of 10 mg/day of propericyazine ameliorated the VH.

The VH experienced by this patient resembles a ghost story published by the Japanese essayist Shinya Nishimaru nearly 50 years ago.[3] One night when he was 23 years old, Shinya Nishimaru encountered a lady leaning against a concrete wall outside a factory. Although it was a dark, moonless night, he noticed the pattern of her clothes and outlines of her face and eyes precisely. At first, she appeared in the open air, but after 1 year began appear beside his bed. When he tried to touch her or hit her with a club, she disappeared as soon as he touched her clothes. One night, she suddenly looked him in the eye which really frightened him. He could hardly sleep a wink for several days, which made him fear for his life, so he fled the town. (A longer version of this story together with other ghost stories can be found in the supplemental information available at http://www.jnnp.com/supplemental.)

This hallucination is characterised by (i) the ghost appearing without any relation to sleep at first, (ii) purely visual hallucinations (that is, the ghost never talked or tried to touch him, and the image of the ghost was very clear), and (iii) the ghost vanishing when he tried to touch it. He consulted a psychiatrist but was diagnosed as sane. However, he later saw another vivid ghost while he was mountain climbing.

We also found ghost tales resembling the VH of our case in a record of authentic Japanese folklore written almost 100 years ago. *Toh-no Monogatari* (*The 'Toh-no' Folktales*) edited by Kunio Yanagida, a famous Japanese folklorist, evaluated more than 300 short verbatim records of face to face encounters from old Japanese folklore, especially those involving traditional Japanese monsters, goblins, and elves. In *Toh-no Monogatari*, we also found two other stories that referred to VH of ghosts, although both of them were experienced by children.[4]

Discussion

Although neuropathological examinations were not performed, our patient was diagnosed as having PCA with VH and the cause was considered to be DLBD or Alzheimer's disease on the basis of the clinical symptoms.[1 5]

The VH of this patient are quite similar to descriptions in Japanese folktales and personal ghost stories. When we investigated these stories from a neurological point of view, we noted several similarities between the stories and the VH common in patients with DLBD, PD, DLB, or Charles Bonnet syndrome (CBS).[1] VH is usually considered one of the clinical symptoms of DLBD, PD with late

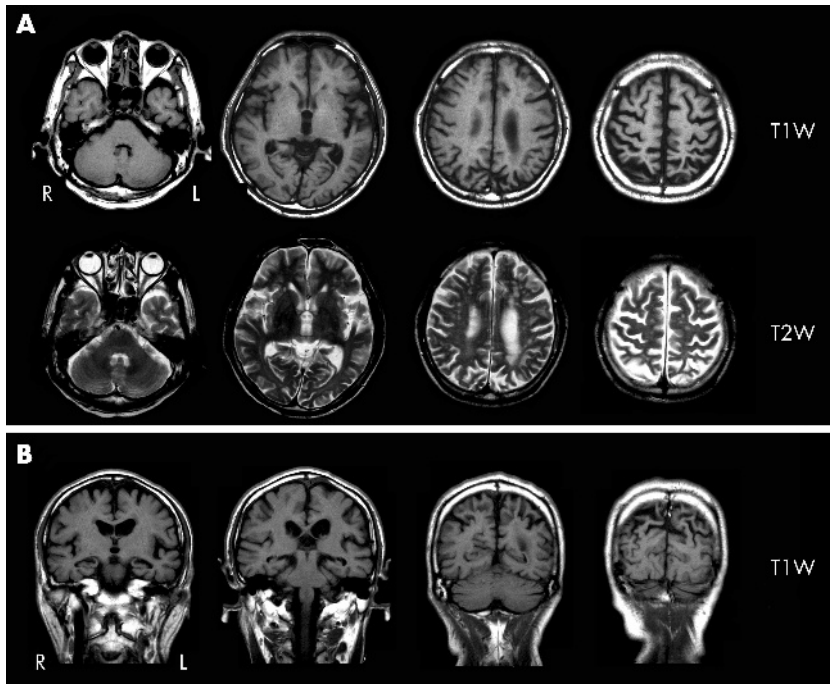

Figure 1 MRI imaging of patient. T1 and T2 images show bilateral parieto-occipital, occipital, and mild left temporal atrophy in horizontal (A) and coronal (B) sections.

developing dementia and rapid cognitive decline, and DLB.[1 2] Similar VH are also reported in CBS, which was originally observed in elderly and visually impaired people without psychiatric or neurological problems. However, a close resemblance between CBS and PD related VH is also suggested.[1]

Recent neuropathological and radiological studies revealed that VH in CBS are associated with hyperperfusion of the temporal cortex, striatum, and thalamus, as well as increased cerebral activation in the ventral extrastriatal region, while DLB patients with VH showed hypoperfusion or hypometabolism of the occipital cortex.[1] It was also reported that PD patients with VH also have alterations in or decreased cerebral activation in the temporo-parietal, parietal, and occipital regions.[1] Our patient showed atrophy of the bilateral parieto-occipital, occipital, and mild left temporal lobes and decreased cerebral perfusion in the parietal and frontal lobes, indicating that the VH in our case may be caused by dysfunction of these regions. Such VH are rarely experienced by normal people, but, once seen, some people may be convinced they are supernatural events. We neurologists pay attention to such reports, and note the similarity in content between ghost tales and VH, and pursue their mechanism of origin.

In conclusion, we propose that some ghost sightings are actually VH in normal people, which occur very rarely but more frequently than we in the medical establishment think.

Electronic-database information

A longer version of the story by Shinya Nishimaru together with other ghost stories can be found in the supplemental information available at http://www.jnnp.com/supplemental.

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Glucose metabolism and dopamine PET correlates in a patient with myotonic dystrophy type 2 and parkinsonism

We describe a 72 year old woman who presented with a 5 year history of progressive gait uncertainty, frequent sudden falls, and difficulty rising from a chair, associated with fatigue. She noticed some degree of distal weakness in her arms when carrying weight compared with previous years. She had no complaints of pain or cramps but noticed stiffness and 'locked' legs when walking. She denied cranial nerve problems and had no general systemic complaints. Past medical history was significant for hypertension. Family history was unremarkable.

Examination revealed normal vital signs and intact cranial nerves. Slow and slightly slurred speech was evident. Her trunk was forcefully bent forwards in a camptocormic attitude, which could be corrected by passive extension of the trunk. Her tone was increased with rigid wrist and hips and brisk (3+) reflexes and an equivocal bilateral extensor toe sign. Her gait was slow, and worsened during the day and over time to the extent that it limited everyday activities and often culminated in sudden falls. Pendular movements were reduced on the left. She had mild weakness in the shoulder abductors (4), finger flexors (4.5), hip flexors (4), and ankle dorsiflexors (4). Sensory examination and coordination were normal.

Routine blood investigation was unremarkable except for red blood cells (3.84×10^6), haemoglobin (11.2 mg/dl) and haematocrit (34%) in the low normal range. Gammaglobulins were 7% of total protein levels. Electrocardiography revealed first degree arteriovenous block. Brain computed tomography scan demonstrated diffuse white matter vascular hypodensity and cortical atrophy. A diagnosis of parkinsonism was made. Treatment with Sinemet CR tablets (carbidopa/levodopa 25/100 mg twice daily) was started. However, despite treatment, the patient had not improved 5 months later. Muscle pain was present. Repeat examination showed severe neck flexor weakness (Medical Research Council scale <3).

The patient was unable to turn on her side in bed, or to lift her trunk from the bed when lying. Laboratory examination was unchanged except for a two fold increase in creatinine kinase (CK) levels. Needle electromyography (EMG) demonstrated diffuse myotonic discharges. Posterior iridescent initial cataract was demonstrated by slit lamp examination in the lens of the left eye. Fluorescent in situ hybridisation on muscle biopsy of the left biceps brachii demonstrated preferential nuclear clump type II fibre atrophy and ribonuclear inclusions, consistent with the diagnosis of myotonic dystrophy type 2 (DM2). $(CTG)_n$ expansion at the *DMPK* gene was normal. The DM2 mutation was confirmed and diagnosis of DM2 was subsequently made.

Neuropsychological tests were administered as previously described.[1] These included a screening test for dementia (Mini Mental State Examination) and tests of nonverbal reasoning (Raven's progressive coloured matrices), auditory language comprehension (Token test), verbal fluency with phonemic and semantic cues, verbal and spatial short term memory (Digit span forward and Spatial span), verbal and spatial long term memory (Story recall and Rey recall), constructional abilities (Rey's complex copy), attention and executive function (Trail Making A and B, Alertness and divided attention, Tower of London test, Wisconsin Card Sorting test). The patient showed an impairment of spatial long term memory (Rey recall: raw score 1 out of 36, cut off of normative data 9.97 out of 36) and of attention and executive function (Divided attention: raw score 892 ms; cut off 811.7 ms), and Tower of London test (percentage error 193%, cut off 100%). In the other tests, the patient's performances were normal.

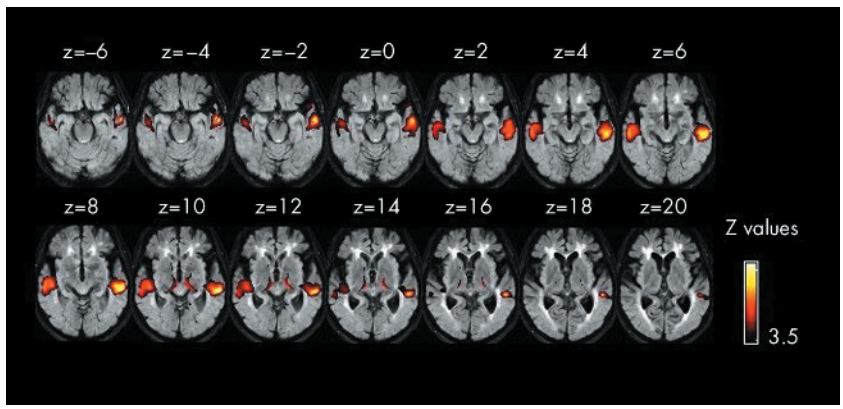

Figure 1 Statistical parametric maps of 18F-FDG PET showing significant metabolic reductions ($p<0.05$ corrected) bilaterally in superior temporal gyrus and parietal operculum (Montreal Neurological Institute stereotactic coordinates of local maxima for suprathreshold clusters x, y, z = 64, −28, 10, Z = 6.01; −62, −22, 6, Z = 5.26), and thalamus (18, −22, 14, Z = 5.06; −8, −16, 12, Z = 4.23), superimposed on the T2 weighted MRI of the patient. The areas indicated in shades of yellow and red both indicate areas of hypoperfusion; yellow represents areas of maximum hypoperfusion.