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CASE REPORT

Difficulty differentiating a case of posterior cortical atrophy from a psychogenic disturbance of vision

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Abstract

Differentiating posterior cortical atrophy (PCA) from other diseases can be difficult and time-consuming, and there is a particularly high possibility of misdiagnosis when psychiatrists diagnose complaints related to visual perception. Here, a case of PCA involving prominent visual perceptual disorders is reported; PCA was difficult to distinguish from psychogenic disturbance of vision in this case. For a year, a 59-year-old woman had had visual perceptual disorders, including a distorted view and prosopagnosia. She underwent examinations at multiple clinical departments at several medical institutions before receiving a definitive diagnosis of PCA. This PCA diagnosis was based on clinical symptoms, including Gerstmann syndrome, Bálint's syndrome, and transcortical sensory aphasia, and hypoperfusion in the occipital lobe observed on single-photon emission computed tomography. This case was initially misdiagnosed as a psychogenic disease partly because characteristic clinical manifestations of PCA include visual agnosia with a disjunctive component. This patient displayed a disordered perception of stationary objects but an intact perception of moving objects. For example, she had to grope her way through a room at home, but she could visit a familiar hair salon on foot without hindrance. Behaviours like claiming to be blind while inexplicably moving without colliding with surrounding objects may lead to the misdiagnosis of PCA as a psychogenic or dissociative disorder involving histrionic or neurologically irrational symptoms with an expectation of sympathy or personal gain. It is critical to make every effort to exclude organic diseases, even in cases provisionally diagnosed as psychogenic disease. Despite its low prevalence, PCA should be considered a syndrome caused by Alzheimer's disease, dementia with Lewy bodies, or other dementias.

Key words: *Bálint's syndrome, dementia with Lewy bodies, Gerstmann syndrome, posterior cortical atrophy.*

INTRODUCTION

Posterior cortical atrophy (PCA) is a syndrome caused by degenerative diseases or other dementias, and it is characterized by visuospatial dysfunction.[1] Benson *et al*. described five cases of PCA with functional decline (as seen in Gerstmann and Bálint's syndrome), transcortical sensory aphasia, and predominant parieto-occipital atrophy.[2] Those patients had relatively preserved memory, insight, and judgement until late in the disease course. Several clinical criteria or detailed inclusion criteria for identifying PCA have been described (1–5 in Appendix S1), and all have highly consistent definitions of PCA. PCA has also been recognized and described in consensus criteria for typical and atypical Alzheimer's disease (6, 7 in Appendix S1).

The differential diagnosis between PCA and other disorders can be difficult and time-consuming.[3] There is a particularly high possibility of misdiagnosis when the characteristic vision-related complaints are diagnosed by psychiatrists. Here, a case of PCA that involved prominent visual perceptual disorders and

that was difficult to distinguish from psychogenic disturbance of vision is reported.

CASE PRESENTATION

A female patient had no relevant family or medical history. She had married at the age of 25 and spent over a decade as a full-time housewife. From the age of 40 to 59 years, she was a public employee in a job related to school-provided lunches.

She began to recognize visual perceptual disorders at age 58, in November of Year X. She stated that she had started to see objects as distorted—for example, a tomato no longer looked round, or the right and left sides of an object would appear to differ in size. She additionally reported difficulty recognizing the faces of entertainers on TV, trouble holding food with chopsticks, and abnormality in her colour vision. She also had become unable to drive a car.

In the year after the onset of symptoms, disease exacerbation rendered the patient unable to recognize family members by their faces, and she began to distinguish people by differences in their voices. She experienced deteriorating cognitive and motor functions, including an inability to mobilize independently, which severely limited her ability to perform activities of daily living. Impairments in language understanding and fluency were also noted. With regard to psychosocial background, the patient experienced a rift with her mother-in-law and stress in relations with co-workers, although there was no definite temporal causal link.

The patient underwent ophthalmological examinations and magnetic resonance imaging at several ophthalmology and cerebral neurosurgery clinics, which revealed no abnormalities. Therefore, she was referred to a psychosomatic clinic. Some hospitals accused her of fabricating her disorder because she had groped her way through a room at home but had been able to visit a familiar hair salon on foot without hindrance. Upon visiting a neighbouring psychiatric clinic, the patient was diagnosed with a somatoform disorder based on organic brain disturbances and the presence of psychological stressors in her environment (i.e. a rift with her mother-in-law and stress in relations with co-workers). The patient was treated with an antidepressant (fluvoxamine 100 mg/day) and an anxiolytic (ethyl loflazepate 1 mg/day) and received supportive psychotherapy from a clinical psychotherapist, but no effect was observed.

Over a year after symptom onset, the patient visited the neurology department of Juntendo University Shizuoka Hospital for a second opinion. Head computed tomography and magnetic resonance imaging revealed no abnormalities; thus, she was diagnosed with a psychogenic disturbance of vision. In April of Year X +1, the patient visited multiple eye clinics and underwent repeated ophthalmological examinations, but none found any abnormality. She gradually began to exhibit delusion-like ideation of persecution from her husband and mother-in-law—for example, she often thought that someone was talking badly about her. She additionally showed jealous delusion-like ideation as represented by statements such as 'my husband was talking to another woman', and she occasionally became unreasonably emotional and irritated. This presentation of paranoia and jealous delusions based on a misunderstanding of her family's conversations was most likely related to prosopagnosia, impaired language understanding, and impaired perceptual spatial processing of voices or auditory inputs, in addition to impaired perception of visual objects.

Seeking another opinion, the patient visited the psychiatric clinic of Juntendo University Shizuoka Hospital in May of Year X+1. Because other hospitals had failed to find causative organic factors, the clinical evaluation focused on psychogenic factors. Given the symptom of delusional ideation, it was suspected that personal misidentification was associated with a pathological mechanism, such as Capgras syndrome, although no definitive diagnosis was achieved. Repeated consultation confirmed that memory impairment and diminished language understanding and output had begun around the same time as the visual abnormalities. On the Mini-Mental State Examination, the patient scored 9 points out of 30 points. Visual inspection of magnetic resonance images did not reveal significant atrophy (Fig. 1), but modest atrophy in the occipital lobe was observed using the voxel-based specific regional analysis system for Alzheimer's disease (8 in Appendix S1) (Fig. 2). Therefore, single-photon emission computed tomography was performed to rule out dementia with Lewy bodies and other dementias. Hypoperfusion in the occipital lobe was observed via statistical analysis of single-photon emission computed tomography images (3-D stereotactic surface projection) using N-isopropyl-p-[123I]iodoamphetamine (9 in Appendix S1) (Fig. 3). Yet, metaiodobenzylguanidine myocardial scintigraphy was negative for dementia with Lewy

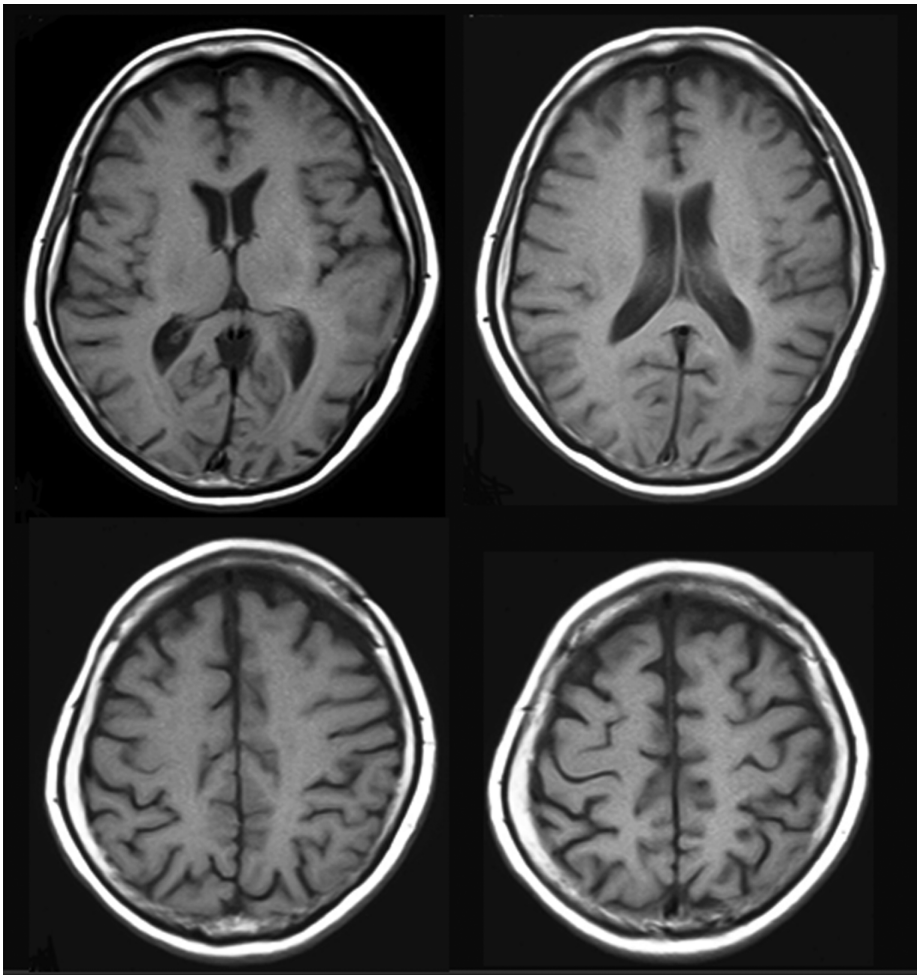

Figure 1 Magnetic resonance imaging (T_1-weighted image) showing no atrophy or ischemic change.

bodies, as no reduction of accumulation was detected (Fig. 4). Tau protein was detected in the cerebral spinal fluid (total tau: 3338 pg/mL; 14-3-3 protein: 1803.7 μg/mL). However, because of the lack of equipment in our hospital, we could not determine cerebrospinal fluid amyloid levels or detect amyloid deposition with positron emission tomography.

Neurological reassessment revealed Gerstmann syndrome based on optic ataxia (i.e. an inability to grab scissors held by an examiner, due to a poor sense of distance, or trying to grab the hand holding the scissors instead of the scissors themselves) and psychic gaze palsy (i.e. an inability to maintain attention in a definite direction). It was also determined that the patient had Bálint's syndrome (based on finger agnosia, right–left disorientation, agraphia, and acalculia) and transcortical sensory aphasia (she would mishear, for example, 'ball' as 'bowo' or 'chosho' as 'tosho').

Ultimately, the patient was diagnosed with PCA based on her clinical symptoms, including Gerstmann syndrome, Bálint's syndrome, and transcortical sensory aphasia, and hypoperfusion in the occipital lobe observed on single-photon emission computed tomography. This diagnosis met the criteria advocated by Benson *et al*. and Tang-Wai *et al*.[2,4] The patient may have already been in the middle phase of PCA rather than the initial phase when she first visited the psychiatric clinic of Juntendo University Shizuoka Hospital.

The patient was treated with yokukansan (5.0 g/day), which relieved her anxiety, delusion-like ideation of persecution, and irritability. Further treatment with 3–5-mg/day donepezil led to a trend of recovery of verbal fluency.

DISCUSSION

This case was initially misdiagnosed as a psychogenic disorder partly because of the presentation of visual agnosia. Patients with PCA experience a gap in their perception between stationary and moving

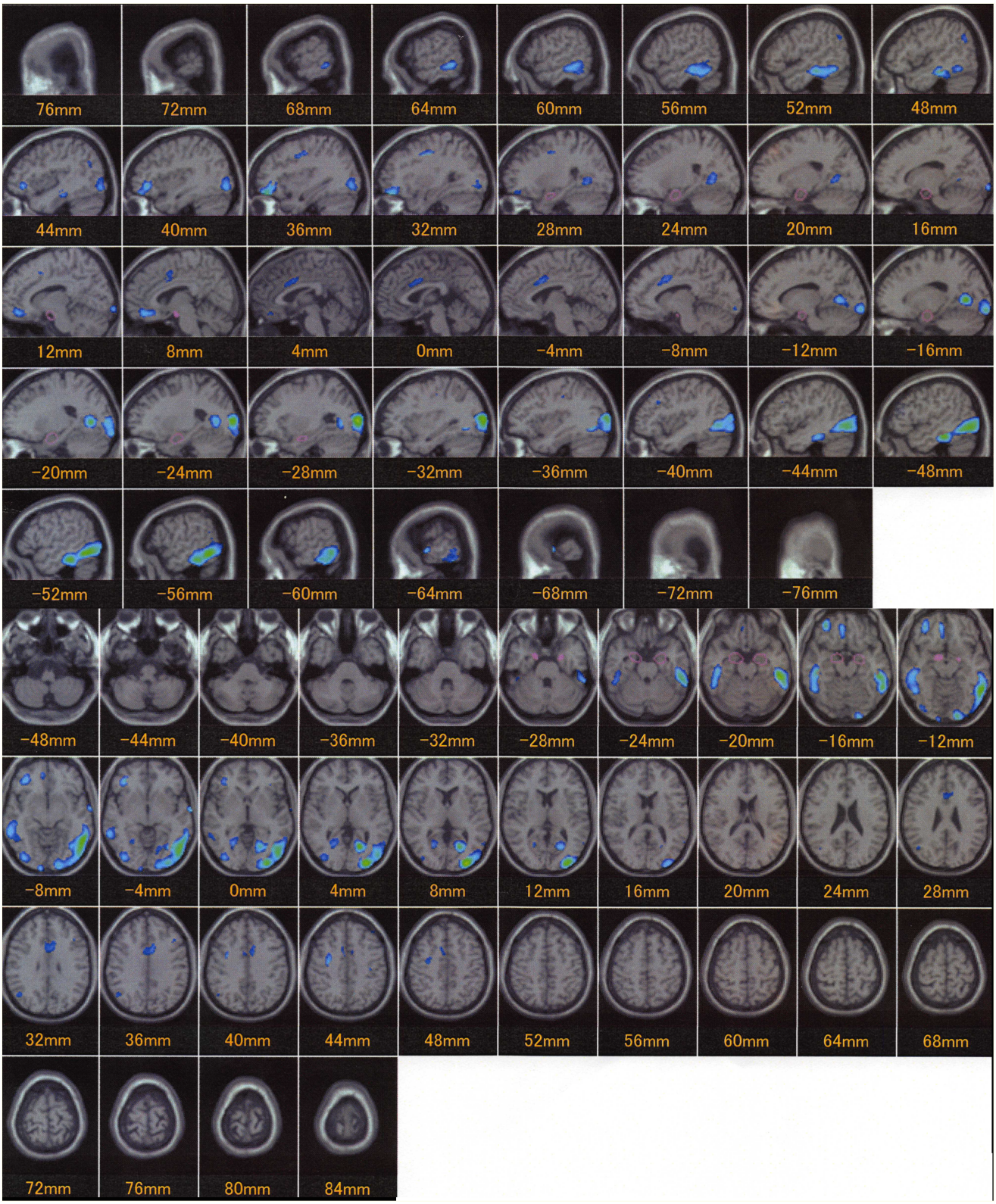

Figure 2 Voxel-based specific regional analysis system for Alzheimer's disease revealed mild atrophy (blue portions) in the occipital lobe, whereas remarkable atrophy was not observed upon visual inspection. The background magnetic resonance image shows a standard brain, not the patient's brain. The labels on each slice represent standard brain coordinates.

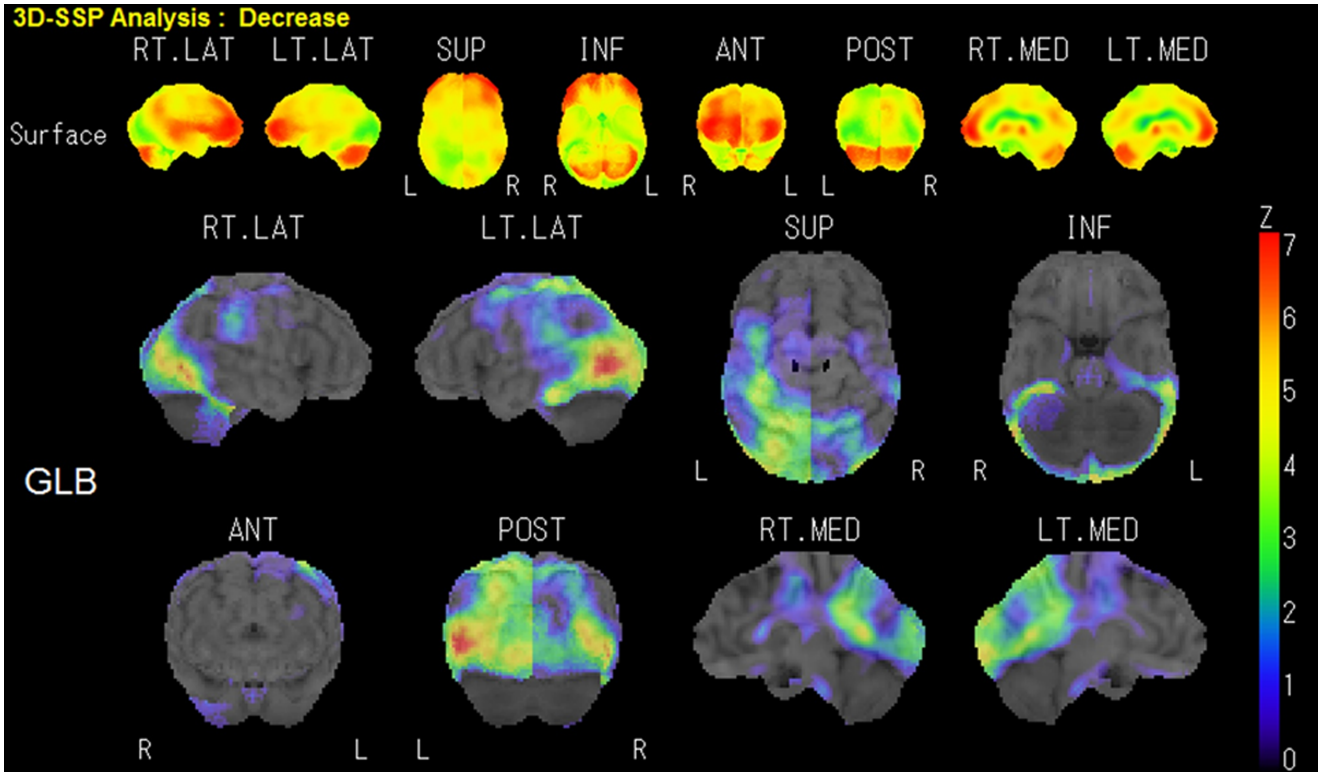

Figure 3 Single-photon emission computed tomography images using 3-D stereotactic surface projection revealed hypoperfusion in the occipital lobe and posterior cingulate gyrus.

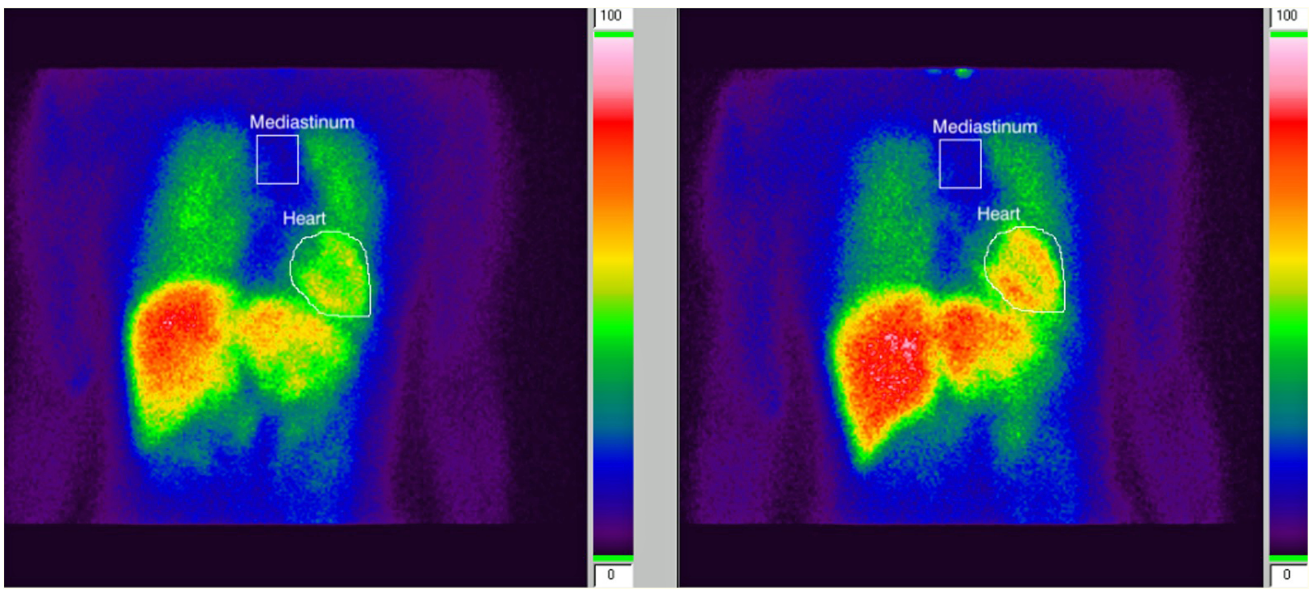

Figure 4 Myocardial scintigraphy (left: 0 min; right: 148.2 min) revealed no reduction in accumulation. The heart-to-mediastinum ratio was 2.60 (0 min) to 3.12 (148.2 min), and the heart washout rate was 5.5%.

objects; for example, they might be unable to recognize a stationary ball but able to catch a thrown ball. Other PCA-associated behaviours can include feeling blind but inexplicably moving without colliding with surrounding objects,[1] feeling blind indoors but still being able to ride a bicycle outdoors,[1] or as in the

present case, having to grope to find one's way indoors but being able to visit a familiar hair salon on foot. These contradictory behaviours can lead to the misdiagnosis of PCA as a psychogenic or dissociative disorder that includes histrionic or neurologically irrational symptoms with an expectation of sympathy or personal gain.

Even in cases that are provisionally diagnosed as psychogenic disorders, as in this case, it is critical to try to exclude organic diseases. Particular attention must be paid to cases in which a patient visits a psychiatric department after the possibility of organic diseases has been excluded by examinations in other clinical departments. It is important to perform a detailed evaluation of the medical history and neuropsychological tests.

Despite its low prevalence, PCA should be considered a syndrome caused by Alzheimer's disease, dementia with Lewy bodies, or other dementias.

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SUPPORTING INFORMATION

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Appendix S1 (Additional REFERENCES)

