INTERNAL MEDICINE

□ CASE REPORT □

Progressive Bálint's Syndrome in a Patient Demonstrating Dementia with Lewy Bodies

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Abstract

We herein report a 65-year-old man demonstrating dementia with Lewy bodies who first presented with Bálint's syndrome. Two years later, a mild cognitive impairment was noted. From three years after onset, he developed progressive parkinsonism, visual hallucination, and autonomic dysfunction, in line with the deterioration of the cognitive function. Single photon emission computed tomography with a ^{99m}Tc-ethylcysteinate dimer performed two years after onset revealed hypoperfusion in the restricted area of the bilateral superior parietal lobule, which extended to the lateral occipital cortices within two years. It is suggested that the pathological process can extend from the parietal to occipital lobes.

Key words: Bálint's syndrome, dementia with Lewy bodies, dorsal simultanagnosia, optic ataxia, dysmacroscopia, posterior cortical atrophy

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Introduction

Dementia with Lewy bodies (DLB) is characterized by parkinsonism, fluctuating attention and alertness, and recurrent visual hallucinations (1). Visuospatial impairments, such as constructional disorder and elementary visuoperceptual disturbances, are also noted (2). On the other hand, Bálint's syndrome (oculomotor apraxia, optic ataxia, and visual inattention) due to a bilateral parieto-occipital junction has rarely been reported in association with DLB. This is probably because the parieto-occipital area is not preferentially damaged, at least in the early stage of DLB.

We herein report a patient with DLB who first showed symptoms of Bálint's syndrome, and discuss the progression of cortical degeneration in DLB.

Case Report

A right-handed, 65-year-old man, a university graduate and retired president of a self-owned business, became aware that he had lost his sense of perspective in 2008. His wife reported that he had screamed during his sleep a few years previously. He could not pick up food with chopsticks, and searched for food on the table. He put on his clothes backwards, had difficulty identifying his wife when outside of his home, and went in the wrong direction to the bathroom in the middle of the night. Also, he was unable to put a Go stone in the designated place on a Go board. The patient was accompanied by his wife to the Department of Psychiatry of our hospital at two years after onset. The Mini-Mental State Examination (MMSE) score was 24/30: he lost scores for place orientation, mental arithmetic, sentence writing, and figure copying. On copying a "double pentagon", he overdrew the figure. MRI showed slight atrophy of the bilateral superior parietal lobule (Fig. 1a). Single photon emission computed tomography with a ^{99m}Tc-ethylcysteinate dimer (ECD-SPECT) revealed hypoperfusion of the bilateral posterior superior parietal area, and the right middle and inferior occipital areas in three-dimensional brain surface images, which were constructed with the easy Z-score Imaging System (eZIS) (3) version 3.4 and Statistical Parametric Mapping (4) version 2 (SPM 2) (Fig. 2; two-sample t-test, patient vs. healthy men aged over 70 years old, uncorrected $p<0.001$). ^{123}I-meta-iodobenzylguanidine (^{123}I-MIBG) myocardial scintigraphy showed a lowered early (1.39) and delayed (1.35) heart-to-mediastinum (H/M) ratio [normal mean (SD): early, 2.1 (0.16); delayed, 2.42 (0.30)],

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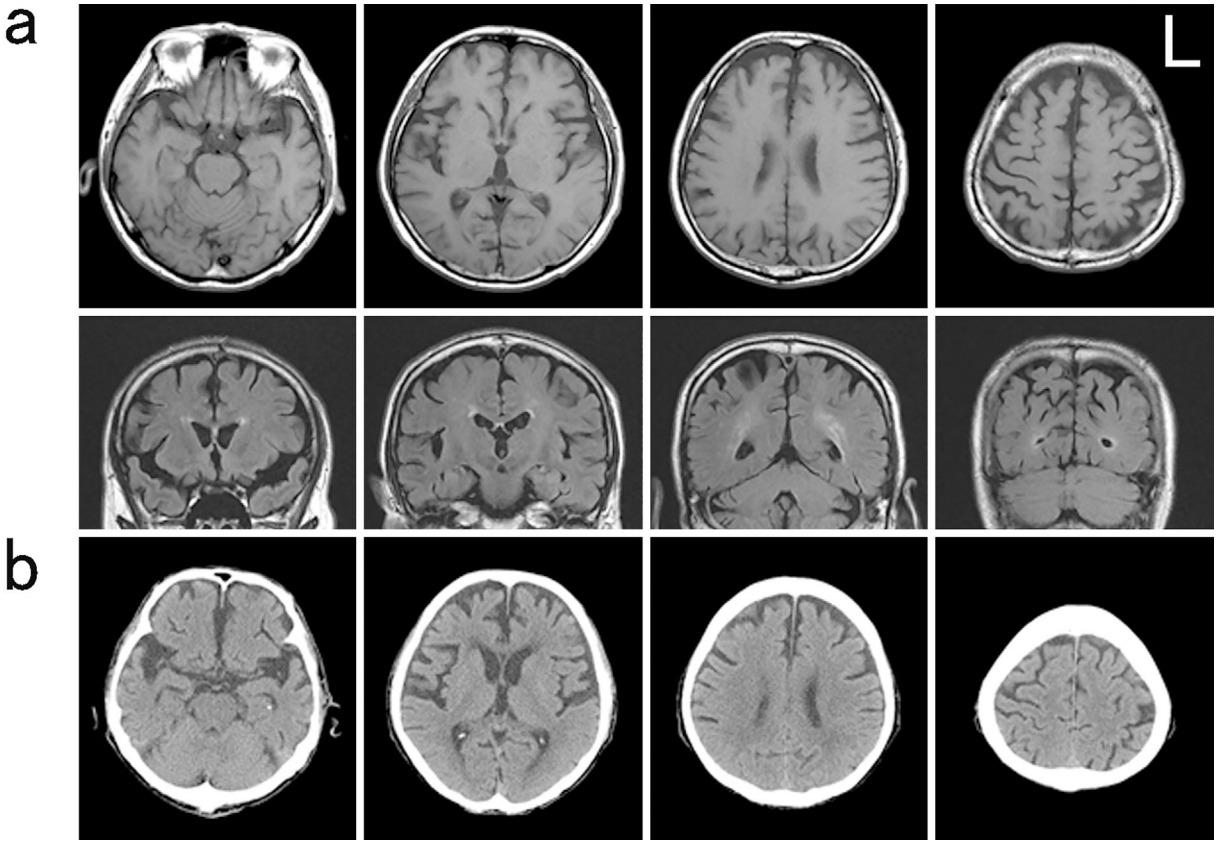

Figure 1. **Brain MRI and CT images of the patient. a: MRI T1-weighted axial (upper) and fluid-attenuated inversion recovery (FLAIR) coronal (lower) images obtained two years after onset revealed slight atrophy of the bilateral superior parietal lobule, b: Brain CT images obtained seven years after onset showed bilateral atrophy of the frontal lobe with enlargement of the anterior horn of the lateral ventricles and Sylvian fissures.**

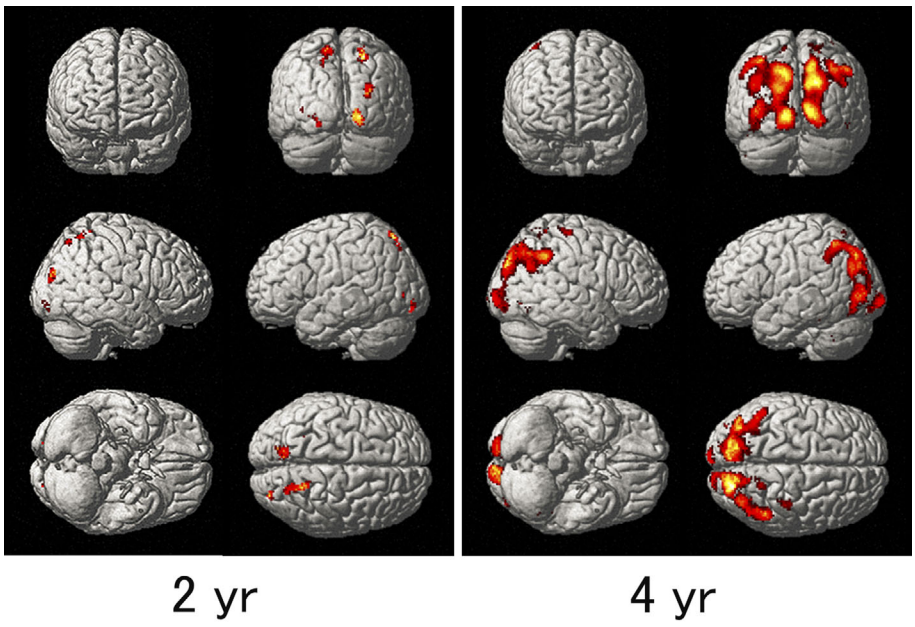

Figure 2. **^{99m}Tc- ECD-SPECT images at two and four years after onset. Hypoperfusion of the bilateral posterior superior parietal and right middle and inferior occipital areas was noted at two years after onset (left). Over the following two years, hypoperfusion extended to the lateral occipital gyri (right). Areas of significant hypoperfusion were determined with the easy Z-score Imaging System (eZIS) version 3.4 and Statistical Parametric Mapping version 2 (SPM 2). Significance was based on the two-sample t-test (patient vs. healthy men, aged over 70 years old, n=20, uncorrected p<0.001).**

thus suggesting either Parkinson disease or DLB (5). Based on these findings, he was diagnosed with DLB. The oral administration of donepezil hydrochloride was thereafter started.

The patient had a history of severe myopia, cataract surgery, and normal pressure glaucoma, and had been administered eye-drops at the Department of Ophthalmology of our hospital. Goldmann perimetry performed two years after onset showed peripheral visual field constriction in the left and right horizontal meridian of the left eye. An examination by a neuro-ophthalmologist at three years after onset revealed poor visual fixation, the absence of smooth pursuit, and exotropia during accommodation.

At three years after onset, the character lines he made when writing slanted upward, and he could not read headline characters more clearly than text in a newspaper (This "headline" symptom may be called dysmacroscopia). He complained of diplopia and loss of visual acuity. Recurrent visual hallucinations were observed: he saw an unknown old woman or children in a room. Also, he mistook an electric pole for a person. Action tremor, bradykinesia, short steps and festination on walking, and impotence appeared. Follow-up ECD-SPECT performed four years after onset revealed hypoperfusion that extended from the bilateral posterior parietal to lateral occipital gyri (Fig. 2). The blood flow of the posterior cingulate cortex and precuneus appeared to be preserved. L-dopa/carbidopa administration was started at four years after onset. The MMSE score at this time was unchanged (24/30). However, at five years after onset, it decreased to 19/30: time orientation, delayed memory, sentence comprehension, and copying of a "pentagon" were additionally impaired. He developed severe constipation. Because of the progression of parkinsonism, the patient was referred to the Department of Neurology of our hospital at six years after onset. On neurological examination, resting tremor of the hands, neck and limb rigidity (cogwheel rigidity in the upper limbs and lead-pipe rigidity with spasticity in the lower limbs), bradykinesia, and retropulsion were noted (Hoehn and Yahr stage III). Furthermore, oculomotor apraxia (slowness in shifting gaze and difficulty of horizontal pursuit), optic ataxia (causing a reaching disturbance in both hands, while performance in the right upper quadrant field was relatively well preserved), and visual extinction were observed. The patient pointed to a clock on the wall, and a fire extinguisher on the floor, but could not find a personal computer on the desk in front of him. Thus, visual inattention (dorsal simultanagnosia) (6) was suspected. He was diagnosed with DLB with Bálint's syndrome. Ropinirole hydrochloride was added to the prescription. However, he stopped taking the drug because he started to fall down. Hypotension (blood pressure, 110/80) was noted: he showed a "pale" face when getting out of bed. Also, urinary incontinence appeared.

At seven years after onset, he began to fall easily, could not walk, and became bedridden throughout the day. Brain CT revealed bilateral atrophy of the frontal lobe with enlargement of the anterior horn of the lateral ventricles and Sylvian fissures (Fig. 1b).

Discussion

The patient first developed suspected rapid eye movement (REM) sleep behavior disorder (screaming during sleep), optic ataxia (inability to pick up food with chopsticks and incorrect placement of a Go stone) and visual inattention (difficulty finding food on a table or a person in a crowd, and difficulty reading headline characters relative to text in a newspaper [dysmacroscopia]), and dressing apraxia. Two years later, mild cognitive impairment with preserved memory was evident. Three years after onset, oculomotor apraxia was confirmed. Over the next few years, parkinsonism, recurrent hallucination, and autonomic dysfunction developed and became aggravated, in parallel with the deterioration of the cognitive function.

The case fulfils the clinical diagnostic criteria for probable DLB (1). Characteristic neuropsychological features of DLB compared with Alzheimer's disease are pronounced visuoperceptual and attentional impairments with a preserved declarative memory (7). As described in the Introduction, DLB presenting with Bálint's syndrome has only rarely been reported, probably because the bilateral parieto-occipital area, a responsible lesion site for Bálint's syndrome, is not initially involved in DLB.

On the other hand, Bálint's syndrome is often observed in posterior cortical atrophy (8). The neuropathological substrate is mostly associated with Alzheimer's disease. However, in a series of 21 individuals who were clinically diagnosed with progressive posterior cortical dysfunction syndrome and had neuropathologic examinations, two cases with part of Bálint's syndrome had an Alzheimer's disease plus nigral and limbocortical Lewy body pathology, and one case with part of Bálint's syndrome had an Alzheimer's disease plus nigral Lewy body pathology (9). It was unclear whether or not these patients suffered from parkinsonism. Our patient suggests that progressive Bálint's syndrome precedes parkinsonism in DLB.

With respect to this point, the present ECD-SPECT demonstrated hypoperfusion of a restricted area of the bilateral posterior superior parietal lobule which extended to widespread areas of the lateral occipital gyri over the course of two years. This finding suggests that cortical degeneration progressed from the superior parietal lobule to the lateral occipital cortices, and this is compatible with the fact that Bálint's syndrome first occurred and recurrent hallucination followed within two years. In this regard, it is known that visual hallucination in DLB is associated with the bilateral parietal and left ventral occipital degeneration (10).

In conclusion, our patient with DLB suggests that cortical dysfunction can emerge in the bilateral parieto-occipital area and then spread posteriorly to the occipital cortices. This pathological process is atypical for DLB in which the occipital area is usually the preceding site for degeneration.

This condition should be differentiated from posterior cortical atrophy, for which Alzheimer's disease is mostly the cause.

The authors state that they have no Conflict of Interest (COI).

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