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 ⁹⁹mTc-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, dysmacropsia, 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 ⁹⁹mTc-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). ¹²³I-meta-iodobenzylguanidine (¹²³I-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)],
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. ⁹⁹mTc- 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 ad-
ministration of donepezil hydrochloride was thereafter
started.

The patient had a history of severe myopia, cataract sur-
gery, and normal pressure glaucoma, and had been adminis-
tered eye-drops at the Department of Ophthalmology of our
hospital. Goldmann perimetry performed two years after on-
set 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 head-
line 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 re-
vealed hypoperfusion that extended from the bilateral poste-
rior 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 de-
creased to 19/30: time orientation, delayed memory, sen-
tence comprehension, and copying of a “pentagon” were ad-
ditionally impaired. He developed severe constipation. Be-
cause of the progression of parkinsonism, the patient was re-
ferred 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 rigid-
ity 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 horizon-
tal pursuit), optic ataxia (causing a reaching disturbance in
former parietal to lateral occipital area (10).

Bálint’s syndrome first occurred and recurrent hallucination
was confirmed. Over the next few years, parkinsonism, re-
current hallucination, and autonomic dysfunction developed
and became aggravated, in parallel with the deterioration of
the cognitive function.

The case fulfills the clinical diagnostic criteria for prob-
able DLB (1). Characteristic neuropsychological features of
DLB compared with Alzheimer’s disease are pronounced
visuoperceptual and attentional impairments with a pre-
served declarative memory (7). As described in the Intro-
duction, 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 syn-
drome, is not initially involved in DLB.

On the other hand, Bálint’s syndrome is often observed in
posterior cortical atrophy (8). The neuropathological sub-
strate is mostly associated with Alzheimer’s disease. How-
ever, in a series of 21 individuals who were clinically diag-
nosed with progressive posterior cortical dysfunction syn-
drome 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 dis-
ease 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 pre-
cedes parkinsonism in DLB.

With respect to this point, the present ECD-SPECT dem-
strated hypoperfusion of a restricted area of the bilateral
posterior superior parietal lobule which extended to wide-
spread 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 oc-
cipital 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 oc-
cipital 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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