Dropped Head Syndrome Preceding the Onset of Dementia with Lewy Bodies

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Abstract

A 67-year-old woman developed dropped head. Her neck was severely flexed, with prominent cervical paraspinal muscles, although no parkinsonism was observed. Brain MRI showed no significant findings. We considered dystonia as the cause of the dropped head and administered trihexyphenidyl, an anticholinergic. After 10 years of follow-up, remarkable psychotic symptoms, including hallucinations regarding insects, appeared. Following the discontinuation of trihexyphenidyl, the psychotic symptoms decreased but still remained. $^{123}$I-N-isopropyl-p-iodoamphetamine single-photon emission computed tomography ($^{123}$I-IMP SPECT) revealed hypoperfusion in the bilateral occipital lobes. We diagnosed the patient with dementia with Lewy bodies (DLB). This case suggests that dropped head syndrome may precede the onset of DLB.

Key words: dropped head syndrome, dementia with Lewy bodies, anticholinergic drugs, trihexyphenidyl

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Introduction

Dropped head syndrome is characterized by the forward flexion of the head and neck that is more pronounced than expected relative to the flexed posture of the trunk. It results from neck extensor weakness or disproportionate tonus of the neck muscles (1). Reported etiologies include amyotrophic lateral sclerosis, myasthenia gravis and several myopathies and extrapyramidal disorders (1). Among extrapyramidal disorders, multiple system atrophy (MSA) frequently presents with head drop (2-4). This feature is considered to be relatively rare in patients with idiopathic Parkinson’s disease (PD) (4, 5).

The incidence of dropped head syndrome among PD patients is reported to be 6.0% in Japan, and dropped head may precede the onset of PD (5). We herein describe the case of a patient with dropped head syndrome that preceded the onset of dementia with Lewy bodies (DLB) by 10 years.

Case Report

A 67-year-old woman developed dropped head over the course of one week. There was no history of rapid eye movement sleep behavior disorder or constipation requiring medication. She had no diplopia, dysarthria, dysphagia, limb weakness or fatigability. On a physical examination, her neck was severely flexed with slight rotation toward the left. The neck flexion was more noticeable than the concomitant camptocormia. The neck extensor muscles were quite prominent and very firm on palpation (Fig. 1A). In particular, the right levator scapulae muscle exhibited marked hypertrophy (Fig. 1B), a finding supported by muscle computed tomography (Fig. 1C). The patient did not have weak neck flexor or extensor muscles. The sternocleidomastoid muscle did not show abnormal contractions, and there was no muscular atrophy, fasciculation, rigidity, rest tremors, bradykinesia or orthostatic hypotension. The serum creatine kinase level was 130 IU/L (normal range, 20-180 IU/L). No anti-acetylcholine receptor antibodies were detected. Repetitive nerve stimulation of the trapezius and hypothenar muscles was normal. Electromyography of the left sternocleidomastoid muscle was minimally abnormal with occasional polyphasic motor unit potentials of normal amplitude. The recruitment was normal, and no spontaneous activity was detected. Head magnetic resonance imaging (MRI) did not
show any atrophy of the brain stem or cerebellum or signal abnormalities of the basal ganglia, and cervical MRI did not reveal significant compression of the cord or roots or any specific signal changes in the paraspinal muscles. We considered disproportionate tonus of the neck muscles as a cause of the patient’s dropped head syndrome, although the definitive etiology could not be determined. We initiated treatment with trihexyphenidyl (4 mg/day), and she experienced a slight improvement. Since that time, we have found no significant progression of the dropped head syndrome, torticollis or camptocormia or the emergence of symptoms of parkinsonism, including muscular rigidity, rest tremors and bradykinesia. Although we prescribed levodopa instead of trihexyphenidyl for six months, the dropped head syndrome worsened. The readministration of trihexyphenidyl again improved the patient’s symptoms.

After 10 years of follow-up, we increased the dose of trihexyphenidyl from 4 to 8 mg/day because the patient was not satisfied with the treatment effect of trihexyphenidyl. Subsequently, remarkable psychotic symptoms gradually developed over the next several months, beginning with visual hallucinations of insects and then progressing to Capgras syndrome and phantom boarders (6). Following the discontinuation of trihexyphenidyl, the patient’s psychotic symptoms decreased; however, they did not disappear. She continued to suffer from visual hallucinations of insects, although the severity reduced. Her primary caregivers noted daytime sleepiness lasting over hours to days. The Mini-Mental State Examination score was 24 of 30, with more deficits in attentional processing than those in memory. We were unable to validate the presence of parkinsonism. We diagnosed the patient as suffering from probable DLB, according to the consensus guidelines for the clinical diagnosis of DLB (7). 

Discussion

We herein presented the case of a 67-year-old woman with dropped head syndrome that preceded the onset of DLB by 10 years. In this patient, the mechanism underlying
been no reports of similar patients, we assumed that the pre-
gates called Lewy bodies (9). Therefore, although there have
upright position. Although no dystonic contractions of the
tic marked contraction of the neck extensor muscles in the
tonus of the neck muscles. The patient exhibited characteris-
cated dystonia (12). In contrast, avoiding medications that exacer-
into the sternocleidomastoid muscles has been reported to be
hibit neck muscle weakness.
appropriate treatment for dropped head syndrome has not
yet been established (1, 8). The injection of botulinum toxin
into the sternocleidomastoid muscles has been reported to be
ineffective, with complications of significant
dysphagia (1-3), and the responses to levodopa have gener-
ally been reported to be disappointing (1-5). Physiotherapy
may alleviate symptoms in some patients (8). Tri-
hexyphenidyl, an anticholinergic agent, is a potential therapy
in patients whose disease mechanism is assumed to involve
dystonia (12). In contrast, avoiding medications that exacer-
bate hallucinations and other perceptual disturbances is im-
portant in patients with DLB. Such medications include an-
ticholinergic agents (13).
In conclusion, this case suggests that dropped head syn-
drome may occur before the clinical features of DLB be-
come evident. The mechanism underlying the development
of dropped head in our patient appeared to involve dispro-

Figure 2. (A) 123I-N-isopropyl-p-iodoamphetamine single-photon emission computed tomography (123I-IMP SPECT) with a three-dimensional stereotactic surface projection (3D-SSP) analysis (Symbia T16, Siemens, Erlangen, Germany; total acquisition time: 28 minutes). The color coding represents the Z-score of the decrease in the regional blood flow (normal controls: age±SD, 64.2±8.2 years; 10 men and 19 women). Hypoperfusion in the bilateral parietal, temporal, occipital, posterior cingulate and precuneus cortices was observed. (B) An early-phase image of 123I-meta-iiodobenzylguanidine (123I-MIBG) myocardial scintigraphy with an H/M ratio of 2.72 (normal range, 2.14-3.38) and (C) a delayed phase-image with an H/M ratio of 2.11 (normal range, 2.31-3.71). The global MIBG washout was 34.9% (normal range, 17.0-38.4%). 123I-MIBG myocardial scintigraphy was performed with Symbia T16 (Siemens). The total acquisition time was four minutes in both phases. H/M ratio: heart-
to mediastinum ratio
portionate tonus of the neck muscles.

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

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References