Naoki Kasahata, Naoaki Dan

Department of Medicine, Tokyo Metropolitan Ohtsuka Hospital, 2-8-1 Minamiohtsuka, Toshima-ku, Tokyo 170-8476, Japan

Corresponding author: Division of Neurology, Department of Medicine, Tokyo Metropolitan Ohtsuka Hospital, 2-8-1 Minamiohtsuka, Toshima-ku, Tokyo 170-8476, Japan; e-mail address: naoki_kasahata@tmhp.jp, telephone number: +81 3 3941 3211, fax number: +81 3 3941 9557

Publication History
Received: 23 September 2016
Accepted: 15 October 2016
Published: November-December 2016

Citation
Naoki Kasahata, Naoaki Dan. An 81-year-old woman mimicking PSP syndrome with decreased uptake of MIBG myocardial scintigraphy. Medical Science, 2016, 20(82), 208-212

Publication License
This work is licensed under a Creative Commons Attribution 4.0 International License.

General Note
Article is recommended to print in recycled paper.

ABSTRACT
An 81-year-old woman with 10-year history of PSP-like symptoms developed falling tendency. Neurological examination showed supranuclear gaze palsy and parkinsonism. MIBG myocardial scintigraphy revealed decreased early H/M ratio 1.4825. Levodopa
treatment improved her symptoms. Parkinson's disease can cause supranuclear gaze palsy. Parkinson's disease should be considered as a differential diagnosis of PSP syndrome.

**Keywords:** supranuclear gaze palsy, Parkinson's disease, metaiodobenzylguanidine (MIBG) myocardial scintigraphy, PSP syndrome

**Abbreviations:** PSP-progressive supranuclear palsy

1. **INTRODUCTION**

Progressive supranuclear palsy (PSP) pathology usually develops PSP syndrome. However, Alzheimer disease mimics PSP syndrome (Kasahata et al., 2014 a, b). Lewy body disease such as dementia with Lewy bodies (DLB) and Parkinson's disease also mimics PSP syndrome. Almost previous cases, however, are autopsy proven cases (Lewis and Gawel, 1990; Fearly et al., 1991; de Bruin et al., 1992; Seno et al., 2000; Nakashima et al., 2003; Clerici et al., 2005; Kasahata et al., 2012, Lagarde et al., 2015).

Decreased $^{123}$I-metaiodobenzylguanidine (MIBG) myocardial scintigraphy uptake seems to represent Lewy body pathology in cardiac sympathetic nerves (Yoshita et al., 1997; Yoshita, 1998; Orimo et al., 1999; Druschky et al., 2000; Orimo et al., 2001; Orimo et al., 2004; Lee et al., 2006; Shin et al., 2006; Rascoc and Schelosky, 2009; Chung et al., 2009; Nagayama et al., 2010). Therefore, decreased MIBG myocardial scintigraphy is used as diagnostic findings of DLB or Parkinson's disease.

We encountered 81-year-old woman presented with supranuclear gaze palsy and levodopa responsive parkinsonism. She also showed decreased uptake of MIBG myocardial scintigraphy. Here, we describe the clinical characteristics of the patient.

2. **CASE REPORT**

An 81-year-old woman admitted to the hospital because of falling tendency. She had been well until approximately 10 years before admission, when she was diagnosed as progressive supranuclear palsy at a certain university hospital. Several years earlier, she developed difficulty in walking and she used walker. Several years earlier, she developed retinitis pigmentosa, thereafter, she developed difficulty in seeing objects. She was able to see person, but she was unable to recognize faces. A few years earlier, she developed syncope with falling. She also developed bradykinesia. Her family thought that she developed dementia. Two years earlier, she was able to walk using walker. She fell several times, and she became difficult to walk and bedridden gradually. She developed falling tendency since last autumn. She moved last December. She neither developed rigidity nor tremor.

Neurological examination showed as follows: she was oriented regarding time and place. She was able to perform easy subtraction. Her auditory comprehension and repetition were good. She showed neither ideomotor apraxia nor limb kinetic apraxia. She was able to name objects. She was able to differentiate hand movement. Her visual field was normal. She was unable to move her eyes vertically. Oculocephalic reflex was able to move her eyes, but vertical eye movements were insufficient. Her masseter tonus was good and facial sensation was normal. Her facial muscle was normal. Her auditory acuity was normal. She was able to elevate her uvula well. Her tongue did not show atrophy and did not deviate at protrusion. She showed neither muscle weakness nor muscle atrophy. Her muscle tonus was normal except neck rigidity. She showed no involuntary movement such as tremor. Her biceps and brachioradial reflexes were bilaterally lively. Her patellar tendon reflexes and Achilles tendon reflexes were absent. She showed no pathological reflex. Plantar responses were indifferent. She showed normal coordination: finger nose tests, rapid alternative movements and heel shin tests were normal. Her sensory system was normal: touch, pinprick, vibration, and position senses were preserved.

$^{123}$I-metaiodobenzylguanidine (MIBG) myocardial scintigraphy revealed decreased early heart mediastinum (H/M) ratio 1.4825 (Figure 1c), and slight accumulation of heart muscle was observed. The easy Z-score system (eZIS) ethyl cysteinate dimer (ECD) single photon emission computed tomography (SPECT) revealed decreased blood flow in frontotemporal lobes (Figure 1b). Magnetic resonance imaging revealed preserved midbrain tegmentum without atrophy, chronic ischemic change, and mild atrophy of medial temporal lobes (Figure 1a).

We prescribed levodopa for deferential diagnosis of parkinsonism and treatment of the present patient. Thereafter, her standing and walking disturbance were improved. She was able to walk independently. Levodopa treatment also seemed to improve her supranuclear gaze palsy.
CASE STUDY

ARTICLE

Figure 1
Magnetic Resonance Images (MRI), easy Z-score imaging system of ethyl cysteinate dimer-single photon emission computed tomography (eZIS ECD-SPECT), and 123I-metaiodobenzylguanidine (MIBG) myocardial scintigraphy findings.

a) A T1-weighted median sagittal section MRI revealed preserved midbrain tegmentum without atrophy.

b) eZIS ECD-SPECT revealed decreased blood flow in frontotemporal lobes.

c) 123I-MIBG myocardial scintigraphy revealed decreased early heart/mediastinum (H/M) ratio 1.4825, and slight accumulation of heart muscle was observed.

3. DISCUSSION
Summary of present patient included as follows: almost normal cognitive state; gaze palsy: predominantly supranuclear and partially nuclear about vertical eye movement; areflexia of lower extremities; standing and walking disturbance; decreased uptake of MIBG myocardial scintigraphy; and improvement her parkinsonism by levodopa.

Table 1
Previously reported DLB or Parkinson’s disease patients presented with supranuclear gaze palsy
All patients presented with supranuclear gaze palsy. 6 patients were autopsy confirmed DLB or Parkinson’s disease. Clerici’s patient was diagnosed by clinical features and positron emission tomography.

<table>
<thead>
<tr>
<th>Reporter</th>
<th>Age/gender</th>
<th>Parkinsonism</th>
<th>Dementia</th>
<th>Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lewis et al.</td>
<td>76/male</td>
<td>+</td>
<td>+</td>
<td>DLB: pathological</td>
</tr>
<tr>
<td>Fearnley et al.</td>
<td>71/male</td>
<td>+</td>
<td>+</td>
<td>DLB: pathological</td>
</tr>
<tr>
<td>de Bruin et al.</td>
<td>70/male</td>
<td>+</td>
<td>+</td>
<td>DLB: pathological</td>
</tr>
<tr>
<td>Seno’ et al.</td>
<td>76/male</td>
<td>+</td>
<td>+</td>
<td>PD+AGD: pathological</td>
</tr>
<tr>
<td>Nakashima et al.</td>
<td>66/male</td>
<td>+</td>
<td>+</td>
<td>DLB: pathological</td>
</tr>
<tr>
<td>Clerici et al.</td>
<td>72/male</td>
<td>+</td>
<td>+</td>
<td>DLB: clinical</td>
</tr>
<tr>
<td>Kasahata et al.</td>
<td>71/female</td>
<td>+</td>
<td>+</td>
<td>PD: pathological</td>
</tr>
<tr>
<td>Lagarde et al.</td>
<td>74/female</td>
<td>+</td>
<td>+</td>
<td>DLB: pathological</td>
</tr>
<tr>
<td>Present patient</td>
<td>81/female</td>
<td>+</td>
<td>-</td>
<td>PD: clinical</td>
</tr>
</tbody>
</table>

Abbreviations DLB: dementia with Lewy bodies, PD: Parkinson’s disease, AGD: argyrophilic grain disease

Naoki Kasahata and Naoaki Dan,
An 81-year-old woman mimicking PSP syndrome with decreased uptake of MIBG myocardial scintigraphy,
Medical Science, 2016, 20(82), 208-212,
www.discoveryjournals.com
It is well known that supranuclear gaze palsy was caused by Lewy body disease such as diffuse Lewy body disease, dementia with Lewy bodies (DLB), or Parkinson’s disease (Lewis and Gawel, 1990; Fearnly et al., 1991; de Bruin et al., 1992; Seno et al., 2000; Nakashima et al., 2003; Clerici et al., 2005; Kasahata et al., 2012, Lagarde et al., 2015). Previously, 8 patients with supranuclear gaze palsy due to Lewy body disease have been reported. Seven patients were autopsy proven patients (Table 1) (Lewis and Gawel, 1990; Fearnly et al., 1991; de Bruin et al., 1992; Seno et al., 2000; Nakashima et al., 2003; Kasahata et al., 2012, Lagarde et al., 2015). One patient was diagnosed by fluorine-18-labeled fluorodeoxyglucose (18F-FDG) positron emission tomography (PET) findings and clinical characteristics: hallucination and fluctuations in cognition and behavior (Clerici et al., 2005). Six patients were diffuse Lewy body disease or DLB (Lewis and Gawel, 1990; Fearnly et al., 1991; de Bruin et al., 1992; Nakashima et al., 2003; Clerici et al., 2005; Lagarde et al., 2015). One patient was Parkinson’s disease and argyrophilic grain disease (Seno et al., 2000). One patient was Parkinson’s disease (Kasahata et al., 2012). Six reported patients were male and 2 was female (Lewis and Gawel, 1990; Fearnly et al., 1991; de Bruin et al., 1992; Seno et al., 2000; Nakashima et al., 2003; Clerici et al., 2005; Kasahata et al., 2012, Lagarde et al., 2015). In contrast, the patient of present study was female, diagnosed as Parkinson’s disease. Rivastigmine improved visual hallucination of the reported DLB patient (Clerici et al., 2005). Levodopa improved parkinsonism of the present patient.

Supranuclear gaze palsy of vertical gaze has been caused by lesion in rostral interstitial nucleus of medial longitudinal fasciculus (riMLF) or nucleus of Cajal. Supranuclear gaze palsy of these DLB or Parkinson’s disease patients has been caused by Lewy body involvement in riMLF, nucleus of Cajal, or oculomotor nucleus (Lewis and Gawel, 1990; Nakashima et al., 2003; Kasahata et al., 2012). Decreased uptake of MIBG myocardial scintigraphy has corresponded to Lewy body pathology (Lewis and Gawel, 1990; Nakashima et al., 2003; Clerici et al., 2005; Kasahata et al., 2012, Lagarde et al., 2015) Parkinson’s disease or DLB. It is reasonable that we perform MIBG myocardial scintigraphy when we suspect Parkinson’s disease or DLB.

4. CONCLUSION
Parkinson’s disease can cause supranuclear gaze palsy. We should consider supranuclear gaze palsy not only as the sign of progressive supranuclear palsy but also as a sign of Lewy body disease. If we suspect underlying Lewy body disease in patients with supranuclear gaze palsy, MIBG myocardial scintigraphy and levodopa treatment are helpful for diagnosis and therapy.

SUMMARY OF RESEARCH

1. Lewy body disease such as dementia with Lewy bodies (DLB) or Parkinson’s disease mimics PSP syndrome. Almost previous cases, however, are diagnosed pathologically, consequently, autopsy proven cases.
2. We clinically diagnosed Parkinson’s disease mimicking PSP syndrome: supranuclear gaze palsy and parkinsonism. It seemed the second case diagnosed clinically.
3. MIBG myocardial scintigraphy was useful to diagnosis of underlying Lewy body disease.

FUTURE ISSUES
I believe that accumulation of clinically diagnosed cases will be able to differentiate clinical features of PSP mimics due to Lewy body disease from PSP. It may be a certain clinical syndrome due to Lewy body pathology.

DISCLOSURE STATEMENT
There is no special financial support for this research work from the funding agency.

REFERENCES
3. Lewis AJ, Gawel MJ. Diffuse Lewy body disease with dementia and oculomotor dysfunction. Mov Disord 1990, 5, 143-147

Naoki Kasahata and Naoaki Dan,
An 81-year-old woman mimicking PSP syndrome with decreased uptake of MIBG myocardial scintigraphy,
Medical Science, 2016, 20(82), 208-212,
www.discoveryjournals.com
17. Lee PH, Yeo SH, Kim HJ, Youn HY. Correlation between cardiac 123I-MIBG and odor identification in patients with Parkinson’s disease and multiple system atrophy. Mov Disord 2006, 21, 1975-77