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An 81-year-old woman mimicking PSP syndrome with decreased uptake of MIBG myocardial scintigraphy

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General Note
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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

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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; 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).

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; Rascol 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 heal 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.

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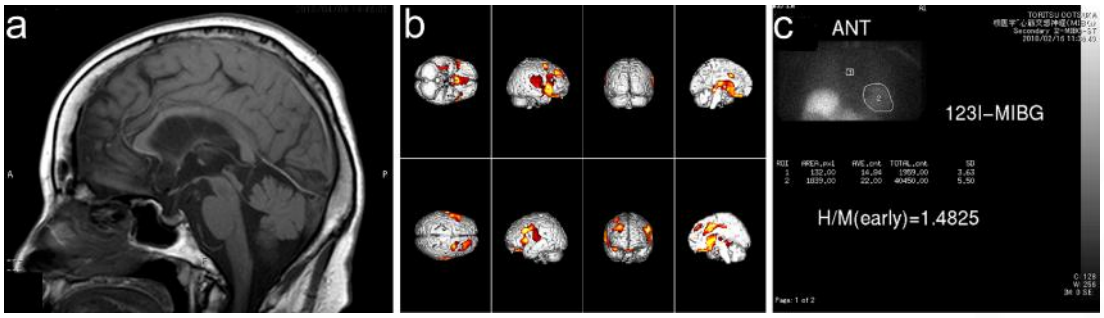

Figure 1

Magnetic Resonance Images (MRI), easy Z-score imaging system of ethyl cysteinate dimer-single photon emission computed tomography (eZIS ECD-SPECT), and ^{123}I-metaiodbenzylguanidine (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.

Reporter	Age/gender	Parkinsonism	Dementia	Diagnosis
Lewis et al.	76/male	+	+	DLB: pathological
Fearnley et al.	71/male	+	+	DLB: pathological
de Bruin et al.	70/male	+	+	DLB: pathological
Seno et al.	76/male	+	+	PD+AGD: pathological
Nakashima et al.	66/male	+	+	DLB: pathological
Clerici et al.	72/male	+	+	DLB: clinical
Kasahata et al.	71/female	+	+	PD: pathological
Lagarde et al.	74/female	+	+	DLB; pathological
Present patient	81/female	+	-	PD: clinical

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

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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 (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; Rascol and Schelosky, 2009; Chung et al., 2009; Nagayama et al., 2010). Therefore, deceased uptake of MIBG suggests diagnosis of 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

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REFERENCES

1. Kasahata N, Hagiwara M, Kato H, Nakamura A, Uchihara T. An 85-year old male with levodopa-responsive parkinsonism followed by dementia and supranucelar ophthalmoplegia caused by Alzheimer-type pathology without Lewy bodies. *J Alzheimer Dis* 2014a, 39(3), 471-476 doi: 10.3233/JAD-131508
2. Kasahata N, Hagiwara M, Kato H, Miyazawa Y, Kamei T, Makita Y, Uchihara T. How Progresssive Supranuclar Palsy (PSP) is Mimicked and What is Mimicking PSP? *J Alzheimer Dis* 14 Apr, 2014b
3. Lewis AJ, Gawel MJ. Diffuse Lewy body disease with dementia and oculomotor dysfunction. *Mov Disord* 1990, 5, 143-147
4. Fearnley JM, Revesz T, Brooks DJ, Frackowiak RSJ, Lees AJ. Diffuse Lewy body disease presenting with a supranuclear gaze palsy. *J Neurol Neurosurg Psychiatry* 1991, 54, 159-161

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An 81-year-old woman mimicking PSP syndrome with decreased uptake of MIBG myocardial scintigraphy,
Medical Science, 2016, 20(82), 208-212,
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5. de Bruin VMS, Lees AJ, Daniel SE. Diffuse Lewy body disease presenting with supranuclear gaze palsy, parkinsonism, and dementia: a case report. *Mov Disord* 1992, 7, 355-358
6. Seno H, Kobayashi S, Inagaki T, Yamamori C, Miyaoka T, Horiguchi J, Wada M, Harada T. Parkinson's disease associated with argyrophilic grains clinically resembling progressive supranuclear palsy: an autopsy case. *J Neurol Sci* 2000, 178, 70-74
7. Nakashima H, Terada S, Ishizu H, Tanabe Y, Yokota O, Ishihara T, Takata H, Ihara Y, Hayabara T, Kuroda S. An autopsied case of dementia with Lewy bodies with supranuclear gaze palsy. *Neurol Res* 2003, 25, 533-537
8. Clerici F, Ratti PL, Pomati S, Maggiore L, Del Sole A, Chiti A, Lucignani G, Mriani C. Dementia with Lewy bodies with supranuclear gaze palsy: a matter of diagnosis. *Neurol Sci* 2005, 26, 358-361
9. Kasahata N, Uchihara T, Nakamura A, Miyazawa Y, Kamei T. A 71-year-old woman with PSP-like symptoms. *Brain Pathol* 2012, 22, 733-736
10. Lagarde J, Hamelin L, Hahn V, Habert MO Seilhean D, Duyckaerts C, Sarazin M. Progressive supranuclear pasly syndrome and semantic dementia in neuropathologically proven Lewy body disease: a report of two cases. *J Alzheimer Dis* 2015, 47, 95-101 doi: 10.3233/JAD-150203
11. Yoshita M, Hayashi M, Hirai S. Iodine 123-labeled metaiodobenzylguanidine myocardial scintigraphy in the cases of idiopathic Parkinson's disease, multiple system atrophy, and progressive supranuclear palsy. *Rinsho Shinkeigaku* (Article in Japanese) 1997, 37, 476-482
12. Yoshita M. Differentiation of idiopathic Parkinson's disease from striatonigral degeneration and progressive spuranuclear palsy using iodine-123 metaiodobenzylguanidine myocardial scintigraphy. *J Neurol Sci* 1998, 155, 60-67
13. Orimo S, Ozawa E, Nakade S, Sugimoto T, Mizusawa H. (123)I-metaiodobenzylguanidine myocardial scintigraphy in Parkinson's disease. *J Neurol Neurosug Psichiatry* 1999, 67, 189-194
14. Druschky A, Hilz MJ, Platsch G, Radespiel-Tröger M, Druschky K, Kuwert T, Neundörfer B. Differentiation of Parkinson's disease and multiple system atrophy in early disease stages by means of I-123-MIBG-SPECT. *J Neurol Sci* 2000, 175, 3-12
15. Orimo S, Ozawa E, Oka T, Nakade S, Tsuchiya K, Yoshimoto M, Wakabayashi K, Takahashi H. Different histopathology accounting for a decrease in myocardial MIBG uptake in PD and MSA. *Neurology* 2001, 57, 1140-1141
16. Orimo S, Amino T, Ozawa E, Kojo T, Uchihara T, Takahashi A, Tsuchiya K, Wakabayashi K, Takahashi H. A useful marker for differential diagnosis of Parkinson's disease-MIBG myocardial scintigraphy. *Rinsho Shinkeigaku* (Article in Japanese) 2004, 44, 827-829
17. Lee PH, Yeo SH, Kim HJ, Youm 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
18. Shin DH, Lee PH, Bang OY, Joo IS, Huh K. Clinical Implications of Cardiac-MIBG SPECT in the Differentiation of Parkinsonian Syndromes. *J Clin Neurol* 2006, 2, 51-57
19. Rascol O, Schelosky L. 123I-metaiodobenzylguanidine scintigraphy in Parkinson's disease and related disorders. *Mov Disord* 2009, 24 Suppl 2, S732-741
20. Chung EJ, Lee WY, Yoon WT, Kim BJ, Lee GH. MIBG scintigraphy for differentiating Parkinson's disease with autonomic dysfunction from Parkinsonism-predominant multiple system atrophy. *Mov Disord* 2009, 24, 1650-55

Nagayama H, Ueda M, Yamazaki M, NishiyamaY, Hamamoto M, Katayama Y. Abnormal cardiac [(123) I]-metaiodobenzylguanidine uptake in multiple system atrophy. *Mov Disord* 2010, 25, 1744-47

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,
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