Myocardial MIBG Scintigraphy in Amyotrophic Lateral Sclerosis

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Introduction

Iodine-123 metaiodobenzylguanidine (MIBG) scintigraphy has been widely used in cardiac diseases and neurological diseases\(^3\). No uptake of MIBG in the myocardium (cardiac image defect) has been noted in Lewy body disease (Parkinson’s disease, dementia with Lewy bodies) and has been used to differentiate Lewy body disease from the other neurological diseases\(^2,3\).

We had a case of amyotrophic lateral sclerosis with frontotemporal dementia (ALS-D) which showed no uptake of MIBG in the myocardium. We report the case and discuss the mechanism of no uptake of MIBG in the myocardium.

Case Report

The patient is a 68-year-old man who visited our clinic with chief complaints of progressive dysarthria, dysphagia, and muscle weakness. His past history and family history were unremarkable. He had not been on any medication.

On neurological examination, atrophy and fasciculation of the tongue (Fig. 1), weakness and atrophy of the four extremities, bilateral hyperreflexia, and bilateral positive Babinski and Chaddock signs were noted.

Mini-mental state examination was 20/30 points. There were no signs nor symptoms of autonomic disturbance or parkinsonism.

Blood tests and electrocardiography were normal. Electromyography showed positive sharp waves and high-amplitude long-duration motor unit potentials in all the extremities. Cerebrospinal fluid examinations and gallium scintigraphy were normal. Sympathetic skin response\(^4\) and coefficient of variation of R-R interval were normal.

The head magnetic resonance imaging (Fig. 2) revealed mild atrophy of the frontal lobes and temporal lobes. We made the diffusion tensor image (Fig. 3) using the magnetic resonance diffusion tensor analysis software “dTV”, which was developed in the Department of Radiology, Tokyo University School of Medicine\(^5\). Single photon emission computed tomography showed blood flow decrease in bilateral prefrontal areas\(^8\). Superoxide dismutase 1 gene was not examined in the present case.

We made a diagnosis of amyotrophic lateral sclerosis with frontotemporal dementia\(^9-12\). In MIBG scin-
Fig. 3  The diffusion tensor images of the head magnetic resonance imaging in the present case (left) and in a healthy person (right). A decrease in the number of nerve fibers traveling through the pyramidal tract is noted in the present case.

Fig. 4  MIBG scintigrams in the present case (upper) and in a healthy person (lower). Cardiac image defect is noted in the present case.

Fig. 5  Thallium scintigram in the present case. Myocardial thallium uptake is normal.

tigraphy, early images were obtained 20 minutes after intravenous injection of 111 MBq of MIBG (Daiichi Radioisotope Laboratories, Tokyo, Japan) and delayed images were obtained 3 hours after the injection by planar method\(^1\). Myocardial MIBG scintigraphy (Fig. 4) showed no uptake of MIBG in the myocardium. Myocardial thallium scintigraphy (Fig. 5) was normal. He died of aspiration pneumonia one year and seven
months after the onset. Autopsy was not performed.

Discussion

Cardiac image defect in MIBG scintigraphy has been reported in a case of familial amyotrophic lateral sclerosis and a case of amyotrophic lateral sclerosis/parkinsonism-dementia complex of the Kii peninsula. The present case is the first report of cardiac image defect in MIBG scintigraphy in amyotrophic lateral sclerosis with frontotemporal dementia. Decreased uptake in myocardial MIBG scintigraphy has been reported in early stages of amyotrophic lateral sclerosis.

The mechanisms of cardiac image defect in MIBG scintigraphy include (1) denervation of postganglionic cardiac sympathetic nerve and (2) disturbance of norepinephrine transporter. In the present case, autonomic dysfunction such as orthostatic hypotension was absent. We consider that cardiac image defect in MIBG scintigraphy in the present case is due to disturbance of norepinephrine transporter by oxidative stress.

Cardiac image defect in MIBG scintigraphy is also seen in 1-methyl-4-phenyl-1,2,3,6-tetrahydroxypridine (MPTP)-induced parkinsonism in mice. In MPTP-induced parkinsonism, the mechanism of cardiac image defect in MIBG scintigraphy is not denervation of postganglionic cardiac sympathetic nerve but disturbance of norepinephrine transporter. The mechanism of cardiac image defect in the present case and in MPTP-induced parkinsonism suggests that the primary cause of cardiac image defect (no uptake) in MIBG scintigraphy in Lewy body disease (Parkinson’s disease, dementia with Lewy bodies) is not denervation of postganglionic cardiac sympathetic nerve but disturbance of norepinephrine transporter.

Conclusion

We reported the first case of amyotrophic lateral sclerosis with frontotemporal dementia that showed cardiac image defect in MIBG scintigraphy. We consider that the cardiac image defect in MIBG scintigraphy in the present case is caused not by denervation of postganglionic cardiac sympathetic nerve but by disturbance of norepinephrine transporter.

References