

Central sleep apnoea in multiple system atrophy

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Abstract

Central sleep apnoea (SAS) is caused by multiple system atrophy (MSA) due to a functional disorder of the brainstem, and sudden death is caused. Respiratory disorders during sleep are evaluated using an examination by polysomnogram (PSG) all night. It is a problem that this testing is complicated, and an evaluation of simpler and easier testing is desirable. We determined whether an evaluation of the SAS can use auditory brainstem response testing (ABR) which is one of the brainstem function tests. The subjects included patients with MSA, hospitalized in Tokushima National Hospital from April 1, 2010 to March 31, 2013. There were two subjects.

The MSA disease severity of the two patients was high. The disease duration period of case 1 was 27 years. With the PSG, the apneic hypopnea index (AHI) intervalsit was normal (0.8 times/h). The contralateral III and V waves of the stimulation were inarticulate in ABR. Case 2 had a shorter disease history by two years. With the PSG, AHI was 30.5, indicating SAS. There was no clear abnormality in ABR. Functional disorders of the brainstem in the MSA can be detected in ABR. Examination of further cases is necessary to elucidate any association between abnormal findings and SAS of ABR.

Keywords : Central sleep apnoea, SAS, multiple system atrophy, MSA, brainstem, polysomnogram

Introduction

Multiple system atrophy (MSA) is an unidentified slow progress-related neurodegenerative disease. This disease occurs in the 50-60 years old age with autonomic nervous system disorders such as cerebellar ataxia, Parkinsonism, orthostatic hypotension, urination disorders, and constipation [1]. Although medical treatment is provided for each symptom, mean length of life after the onset is 8-10 years. With MSA, the frequency of sudden death is than 10-30%, which is very much higher compared with other neurodegenerative diseases [2,3].

Sudden death often occurs during sleep at night. Central sleep apnoea (Sleep Apnea Syndrome:SAS) and vocal cord extorsion paralysis due to a disorder of the apneustic center of the brainstem is suggested. Polysomnography (PSG) is used for diagnosis of SAS. In PSG, some problems requiring hospitalization and multiple testing are noted. Auditory brain stem response ABR is used for evaluation of the brainstem function in the action potential of the brainstem acoustic pathway which occurs in response to sonic stimulation from the outside. This testing can be superior to detection of dysfunction of the hindbrain

bridge joint - midbrain where the apneustic center is located [4,5]. The examination for ABR is conducted at awakening and is very much simpler and easier than PSG. In this study, ABR and PSG are gone the MSA patients for the same period, and central sleep apnoea obtained with the PSG was weighed against the findings of ABR. If the central apnea of the MSA patients could be evaluated using ABR, it is thought that it would be useful in respiratory care and in improving convalescence of MSA patients.

Materials and methods

The subjects were hospitalized in Tokushima National Hospital from April 1, 2010 to March 31, 2013 or it is the hospitalized patients.

They were possible or probable MSA cases and patients who had a diagnosis based on the diagnostic criteria of Gilman et al. [6]. For the patients who were able to acquire consent form, clinical evaluations of daytime sleepiness and the MSA were performed according to JESS and UMSARS. The examination for ABR was conducted based on the evoked response measurement guidance plan of the Japanese clinical neurophysiology society (1997 revision). The PSG examination was consigned to the Philips company, and the presence or absence of sleep apnoea, and the kind and severity of the disease were determined.

Results

The patients hospitalized from April 1, 2010 to March 31, 2013 were 15 people (eight men, seven women, average age 69.1±8.2 years old). The age of the patients at onset was 59.1±7.4 years old. The disease duration period was 9.2±5.9 years. As for the clinical type, nine people were MSAp, and five people were MSAc. Because one person had an autonomic nerve symptom to a foreground set, Shy-Drager syndrome was diagnosed. Because a respirator was attached, 11 people were excluded among ten hospitalized patients by a subject of this study. From the four patients who remained, two agreed to participate in this study.

Patient 1

81-year-old man. He was the patient with MSAc which occurred with an ataxic gait at the age of 54. The disease duration period was 27 years. Snoring while sleeping at night had not been noted. There are cerebellar ataxia, orthostatic low blood pressure, and sleeplessness. The score of UMSARS was: Part I (Historical Review) 35/48 points, Part II (Motor Examination) 32/56 points, Part III (Autonomic examination) orthostatic hypotension, and in Part IV (Global Disability Scale) 4, JESS was nine points. In the PSG examination, the apneic hypopnea index (AHI) was 0.8 times/h of intervals. Mean SpO₂ was 97%. The sleeping breathing state was almost stable. In ABR, the I wave was 1.58 msec for the top latency of the left stimulation side. III wave, 3.67msec; V wave, 5.85msec. The right top latency of I wave was 1.46 msec, III wave 3.8 msec, V wave 5.9 msec. The latency between the left tops was within the normal range; I-III 2.09msec, III-V 2.2, I-III 2.34, III-V 2.1.

Patient 2

62-year-old man. He experienced trunkal ataxia and dysarthria at the age of 60. He had MSAc, and the disease duration period was two years. It had been noted that he had snored while sleeping since before cerebellum ataxia developed. He went to our hospital at 62 years old. An examination for simple PSG was performed, and we made a diagnosis of SAS. (AHI 30.5, mean SpO₂ level 95%, lowest SpO₂ level 86%). Continuous positive airway pressure (a CPAP) therapy was started promptly. UMSARS was: Part I (Historical Review) 40/48 points, Part II (Motor Examination) 31/56 points, Part III (Autonomic examination) orthostatic hypotension, and Part IV (Global Disability Sale) 5 points, and JESS was one point. Because CPAP therapy had already been introduced, an examination for PSG could not be conducted. In ABR, the top latency of the left stimulation side was normal; Left I wave 1.56 msec, left III wave 4.03 msec, left V wave 5.83msec, right I wave 1.56 msec, III wave 3.88 msec, V wave 5.57 msec. The latency between tops was normal; left I-III 2.27 msec, left III-V 1.8 msec, right I-III 2.35, and right III-V 1.69 msec. The stimulation and the contralateral response were not recorded.

Discussion

In the MSA, the frequency of sudden death is much higher than in other neurodegenerative diseases such as Alzheimer's disease or Parkinson's disease [2,3]. Sudden death often occurs during sleep at night, and the involvement of a respiratory disorder during sleep is suggested. The sleep-disordered breathing of MSA occurs as a result of neurodegeneration of the brainstem, and onset occurs through two mechanisms. One is central hypoventilation / apnea caused because of neurodegeneration of the apneustic center which is present in pons-oblongata. The other is laryngeal stenosis / obstructive apnea caused by an extorsion disorder of the vocal cords which grow as a result of neurodegeneration of the bulbar ambiguous nucleus [7]. The presence of excessive daytime sleepiness and snoring while sleeping suggests the presence of these respiratory disorders. For MSA patients with the suit, the enforcement of a PSG examination and a laryngoscopy is necessary [8,9]. Based on the results, the presence or absence of sleep-disordered breathing, disease severity and onset mechanism are analyzed, and treatment such as a CPAP or tracheostomy is provided. The two MSA cases enrolled in this study in Tokushima National Hospital were MSAC together, and the disease severity was equivalent. Although the disease duration period was very long at 27 years, case 1 did not have excessive daytime sleepiness. Snoring while sleeping had not been noted either. AHI in the PSG examination was in the criteria range. On the other hand, as for case 2, the disease duration period was in sleeping snoring with two years though we were very short. Because the AHI was 30.5d, we made a diagnosis of serious SAS after a PSG examination. The findings for The present patient were as follows. The sleep-disordered breathing of MSA does not have a strong relation to the disease duration period or the disease severity of the MSA. This may develop from the early stage. A sleeping snore is an important signature of sleep-disordered breathing. By the examination for ABR, I of the stimulation side, III, the top latency of the V wave and the latency between tops were normal. Top latency and the latency between tops of I

wave, III wave and V wave were normal. In case 1, the contralateral IIIIV wave was inarticulate. These abnormal findings suggest a disorder in the intersection fiber appearing from the cochlear nuclei in a bridge and the bulbar border. However, it is unknown whether the disorder of this part is associated with SAS.

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