CASE REPORT

Continuous Positive Airway Pressure Therapy and Tracheostomy for Multiple System Atrophy Accompanied by Sleep Apnea-hypopnea Syndrome

Sakuo Hoshi¹; Koichiro Tatsumi²; Tetsuo Yamaguchi¹; Yoshihito Yamada¹; Yuko Matsu¹; Chiyoko Kono¹; Hiroko Amano¹; Hiroshi Kamoshita³; Yuichi Takiguchi²; Takayuki Kuriyama²

ABSTRACT — Background. In multiple system atrophy (MSA) patients, a variety of sleep-related respiratory disturbances, including sleep apnea-hypopnea, are frequently manifested. Both nasal continuous positive airway pressure (CPAP) therapy and tracheostomy have been shown to be effective to treat sleep-related breathing disorders in patients with MSA. Case. A 62-year-old man with MSA accompanied by sleep apnea-hypopnea syndrome and marked obesity was admitted because of gradual worsening daytime sleepiness, fatigue and snoring. Arterial blood gas analysis showed hypoxemia and hypercapnia. Laryngoscopy revealed vocal cord abductor paralysis. He was first treated by CPAP therapy. Next, tracheostomy was performed to avoid sudden death due to vocal cord abductor paralysis. Conclusion. CPAP therapy and subsequent tracheostomy gradually improved daytime hypoxia and hypercapnia as well as sleep desaturation, suggesting that oxygen desaturation due to sleep disordered breathing and obesity may partly contribute to the pathogenesis of alveolar hypoventilation in the present case. (JJSRE. 2004;26:633-638)

KEY WORDS — Multiple system atrophy, CPAP therapy, Tracheostomy, Vocal cord abductor paralysis, Sleep apnea-hypopnea syndrome

INTRODUCTION

Patients with multiple system atrophy (MSA) including olivo-ponto-cerebellar atrophy (OPCA) and Shy-Drager syndrome (SDS) frequently manifest a variety of sleep-related respiratory disturbances, which could be life-threatening.¹² Among them, a common and serious complication is bilateral vocal cord abductor paralysis manifested by inspiratory stridor or snoring during sleep. Nasal continuous positive airway pressure (CPAP) therapy has been shown to be effective for sleep-related respiratory disturbances in patients with MSA.⁵⁴ However, tracheostomy is eventually needed in most cases, because no other effective treatment for vocal cord abductor paralysis has been established.⁵⁸ Here, we describe a case of MSA, in which both nasal CPAP therapy and tracheostomy were effective regarding the degree of sleep desaturation and daytime alveolar hypoventilation.

CASE REPORT

A 62-year-old man was referred to our hospital in February 2001, mainly because of urinary incontinence, which had persisted since August 2000. At that time, MSA was diagnosed based on ataxic dysarthria, ataxic abasia, and neurogenic bladder. His weight increased from 80 to 100 kg in one year, in 2001. He was hospitalized in June 2002 because of gradual worsening daytime sleepiness, fatigue and snoring. Epworth sleepiness score, which evaluates the magnitude of daytime sleepiness, was 14 out of 24. He had smoked 1.5 packs of cigarettes per day for 30 years, but quit smoking 12 years before.

On admission, his height was 164 cm and his...
weight was 105 kg, and his body mass index (BMI) was 39 kg/m². His blood pressure was 120/70 mmHg in a sitting position, and 86/52 mmHg in the standing position. A laboratory analysis revealed an FBS of 96 mg/dl and HbA1C of 6.1%. The %VC was 85.5%, FEV₁% 68.0% and FEV₁/FVC 2.17 liter, suggesting a slight decrease in %VC and slight obstructive airflow limitation (Figure 1). Arterial blood gas analysis showed pH 7.36, partial pressure of oxygen (PaO₂) 46.5 mmHg, and partial pressure of carbon dioxide (PaCO₂) 59.6 mmHg, suggesting a complication of obesity-hypoventilation syndrome was obtained. After hyperventilation a pH of 7.40, PaO₂ of 62.7 mmHg and PaCO₂ of 53.9 mmHg. Chest X-ray film findings were normal except that the CTR was 55%, suggesting that marked obesity may explain an enlarged CTR (Figure 2). Echocardiography indicated no left ventricle dysfunction. Brain MR imaging showed slight atrophy of the cerebellum, which was not different from that of his initial visit (February 2001) (Figure 3). His vocal cord abductor muscles were partially paralyzed and rigid (Figure 4), which was classified by laryngoscopy as stage II (moderate) according to the classification made by Isozaki.⁹ Oxygen inhalation therapy (O₂ 1 l/min) was first started to ameliorate the degree of hypoxemia.

Obstructive sleep apnea-hypopnea syndrome (SAHS) was diagnosed because his apnea/hypopnea index was 54.6 by polysomnography (Figure 5). No central sleep apnea was observed during sleep. When awake, his chest and abdominal movements were synchronized. As he did not accept tracheostomy, nasal CPAP therapy was first adopted using an Autoset Portable II kit (Teijin Ltd., Tokyo, Japan). Ten cmH₂O was his maximum tolerable pressure in CPAP therapy with oxygen inhalation (O₂ 1 l/min). The degree of sleep desaturation ameliorated, and daytime sleepiness and fatigue improved 10 days after the induction of CPAP therapy (Table 1).

About one month after the induction of CPAP therapy during sleep, he suddenly suffered from severe dyspnea once during daytime, probably due to sudden deterioration of vocal cord abductor paralysis and rigidity. Therefore tracheostomy was performed to avoid sudden death. After tracheostomy, daytime sleepiness and fatigue further improved and he stopped snoring. The degree of sleep desaturation and arterial blood gas analysis was re-evaluated 1.5 years after tracheostomy. His body weight had not markedly changed during the follow-up period (body weight 102 kg 1.5 years after tracheostomy). Though the degree of sleep desaturation did not clearly ameliorate soon after tracheostomy, it was more improved 1.5 years after tracheostomy (Table 1). His Epworth sleepiness score improved from 14 to 3. Arterial oxygenation and hypercapnia improved (Table 2) and he was finally weaned from oxygen inhalation during daytime. Though brain MR imaging (Figure 3) and the symptoms of MSA such as ataxic dysarthria, ataxic abasia, and neurogenic bladder gradually deteriorated, the improvement of arterial oxygenation was still observed 1.5 years after tracheostomy.
Figure 3. Brain MR imaging showed slight atrophy of the cerebellum on his second visit, similar to his initial visit. However, the atrophy of the cerebellum deteriorated 1.5 years after his second visit. a, b. Brain MR imaging on the initial visit (April 27, 2001). c, d. Brain MR imaging on the second visit (July 31, 2002). e, f. Brain MR imaging on the third visit 1.5 years after tracheostomy (January 24, 2004).
Figure 4. Laryngoscopy performed during daytime showed severe vocal cord abductor paralysis at inspiration, exhibiting a deterioration at one year. a. Expiration on admission. b. Expiration after one year. c. Inspiration on admission. d. Inspiration after one year.

DISCUSSION

In MSA patients, a variety of sleep-related respiratory disturbances, including sleep apnea-hypopnea, are frequently manifested, and may cause sudden death. A vocal cord abductor paralysis, which is usually associated with stridor or snoring during sleep, is thought to be the most serious complication. Nasal CPAP therapy has been shown to be effective for sleep-related breathing disorders in patients with MSA. However, Bannister et al. have recommended that tracheostomy should be performed at early stages of MSA, if there is any central respiratory disorder (i.e. histological or functional changes in the nucleus ambiguus).

To the best of our knowledge, only a small number of cases, including this patient, have been reported in Japan regarding the effectiveness of nasal CPAP therapy in MSA patients. Sleep apnea-hypopnea and oxygen desaturation have been successfully treated with 5 to 10 cmH2O pressure of CPAP, including our case. In most cases, the duration of MSA was longer than 5 years, although in the present case it was only 2 years.

In reported MSA cases treated with tracheostomy, including our case, the duration of the disease varied without any relation to respiratory symptoms. It has not been determined when and which therapy, nasal CPAP therapy or tracheostomy, should be first performed. Isozaki et al. recommended that tracheostomy should be taken into consideration according to the stage of vocal cord abductor paralysis. The comparison of CPAP therapy and tracheostomy in the present case suggests that both CPAP and tracheostomy were effective to improve subjective symptoms and ameliorate the degree of
MSA Accompanied by SAHS—Hoshi et al

Figure 5. Obstructive apnea in stage II vocal cord abductor paralysis. Polysomnography showed sleep apnea accompanied by oxygen desaturation. Thor. move.: thoracic movement, Abd. move.: Abdominal movement.

Table 1. Degree of Sleep Desaturation

<table>
<thead>
<tr>
<th></th>
<th>Before therapy (O₂ 1/min)</th>
<th>CPAP (10 cmH₂O, O₂ 1/min)</th>
<th>Tracheostomy (room air)</th>
<th>Tracheostomy after 1.5 years (room air)</th>
</tr>
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<tbody>
<tr>
<td>% time of SaO₂ ≤ 95%</td>
<td>66</td>
<td>57</td>
<td>44</td>
<td>41</td>
</tr>
<tr>
<td>% time of SaO₂ ≤ 85%</td>
<td>38</td>
<td>7</td>
<td>14</td>
<td>5</td>
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<td>% time of SaO₂ ≤ 75%</td>
<td>37</td>
<td>6</td>
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<td>% time of SaO₂ ≤ 65%</td>
<td>35</td>
<td>6</td>
<td>1</td>
<td>3</td>
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</table>

Table 2. Arterial Blood Gas Analysis

<table>
<thead>
<tr>
<th></th>
<th>Before therapy (room air)</th>
<th>Before therapy (O₂ 1/min)</th>
<th>CPAP (O₂ 1/min)</th>
<th>Tracheostomy (room air)</th>
<th>Tracheostomy after 1.5 years (room air)</th>
</tr>
</thead>
<tbody>
<tr>
<td>pH</td>
<td>7.36</td>
<td>7.33</td>
<td>7.38</td>
<td>7.41</td>
<td>7.41</td>
</tr>
<tr>
<td>PaO₂ (mmHg)</td>
<td>46.5</td>
<td>58</td>
<td>75.1</td>
<td>76</td>
<td>70.1</td>
</tr>
<tr>
<td>PaCO₂ (mmHg)</td>
<td>59.6</td>
<td>67.4</td>
<td>55.2</td>
<td>44.6</td>
<td>40.9</td>
</tr>
<tr>
<td>SaO₂ (%)</td>
<td>81.7</td>
<td>89.3</td>
<td>95.7</td>
<td>95.4</td>
<td>95.3</td>
</tr>
</tbody>
</table>

sleep desaturation. However, we consider that tracheostomy should be selected for the treatment of MSA patients to avoid sudden death, when a vocal cord abductor paralysis appears to manifest or worsen.

In this case, arterial blood gas analysis showed hypoxemia and hypercapnia, which slightly, but not fully, returned to normal range by hyperventilation maneuvers, suggesting that respiratory muscle weakness may partly contribute to alveolar hypoventila-
tion. Our patient was obese with a BMI of 39, and also had vocal cord abductor paralysis and rigidity, which was classified as Isozaki stage II. The complication of MSA and obesity-hypoventilation syndrome could be considered as pathogenetic factors on sleep disordered breathing in the present case. These observations suggested that upper airway obstruction due to both obesity and vocal cord abductor paralysis, may partly contribute to the desaturation during sleep in this patient. The degree of sleep desaturation did not completely improve soon after tracheostomy, but it was more ameliorated 1.5 years after tracheostomy. This indicates that sleep-disordered breathing, including obstructive sleep apnea hypopnea and respiratory dysrhythmia, may have contributed to sleep desaturation in the present case.

Nasal CPAP therapy improved oxygenation and hypercapnia during daytime, and tracheostomy yielded further improvement. This could contribute to the amelioration of the degree of sleep desaturation, because baseline oxygen saturation levels increased before sleep. Ameliorated oxygenation may improve the function of the medulla respiratory-related motor-neurons, which could contribute to the decrease of sleep-disordered breathing. However, improvement of oxygenation may not be involved in the pathogenesis of vocal cord abductor paralysis.

Nasal CPAP therapy is probably beneficial, because it is non-invasive and does not affect the daily life of a patient. However, compliance for nasal CPAP is not necessarily good. About 50-80% of patients are able to continue nasal CPAP for a long time but some patients withdraw from nasal CPAP in a short time. The present patient complained of discomfort with nasal CPAP, but did not complain of invasion by tracheostomy. It may be important that the severity of vocal cord abductor paralysis does not correlate with the period of MSA. Kurisaki reported that the average life expectancy of MSA patients after tracheostomy was 2.5 years and partial pressure of oxygen gradually decreased to about 60 mmHg. However in those cases, partial pressure of oxygen seemed to be maintained over 70 mmHg for at least 1.5 years. Tracheostomy should therefore be considered in early stage patients with MSA, who show snoring and sleep desaturation partly due to vocal cord abductor paralysis.

ACKNOWLEDGEMENTS

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REFERENCES