Management of Impaired Vocal Fold Movement During Sleep in a Patient with Shy-Drager Syndrome

Hirohito Umeno, MD,* Yoshihisa Ueda, MD,* Kazunori Mori, MD,* Keiichi Chijiwa, MD,* Tadashi Nakashima, MD* and Nasser M. Kotby, MD†

A 46-year-old woman with Shy-Drager syndrome is presented. She has impaired vocal fold abduction during sleep, but has no laryngeal dysfunction while she is awake. In order to reduce laryngeal obstruction during sleep, she initially underwent laterofixation of 1 vocal fold (Ejnell’s method) with little lasting success because of accidental slipping of the ligature. Later, she successively underwent arytenoidectomy with the use of CO₂ laser. Her nocturnal breathing improved markedly after arytenoidectomy.

Case Report

A 46-year-old woman presented to the neurology clinic at the Kurume University Hospital with a 25-month history of urinary incontinence, edema of the lower extremities, and a 12-month history of nocturnal stridor. Neurologic examination showed orthostatic hypotension, atonic bladder, and rectal incontinence. The diagnosis of Shy-Drager syndrome was suspected. Because marked nocturnal stridor is associated with a high possibility of sudden nocturnal death, she was referred to the otolaryngology clinic at the Kurume University Hospital. Laryngoscopy revealed no vocal fold palsy while she was awake. However, during sleep, both vocal folds approximated during inspiration (Fig 1A) and abducted during expiration (Fig 1B). Marked inspiratory stridor was noted and her arterial O₂ saturation rate fell to 89%. Her voice was normal at all instances.

Laryngeal electromyography during phonation showed normal activity in the left thyroarytenoid muscle (Fig 2A) and low-frequency firing in the right thyroarytenoid muscle (Fig 2B).

She underwent tracheostomy and laterofixation of the right vocal fold (Ejnell’s method) under general anesthesia to reduce laryngeal obstruction.

Although her voice deteriorated somewhat after this surgical treatment (G2R0B2A0S0), no aspiration was noted and her nocturnal stridor improved dramatically. Thirteen days
after the surgery, her tracheostoma could be closed completely.

However, 16 days after the surgery, she suffered from a sudden episode of dyspnea possibly attributable to laryngospasm for which a tracheostomy was done. The real cause of this spasm is not known. It may be attributable to laryngeal irritation by the threads used in the laterofixation to anchor the vocal fold. Laryngoscope done after the subsidence of this laryngospasm showed that the thread that had abducted the right vocal fold had broken, thereby resulting in a small glottal gap during respiration and reappearance of severe nocturnal stridor. Twenty-one days after the initial surgical treatment, she underwent arytenoidectomy with the use of CO₂ laser under direct laryngoscopy. The vaporized area extended to the tip of the vocal process anteriorly, to the laryngeal ventricle laterally, and to the posterior edge of the arytenoid cartilage posteriorly (Fig 3A), resulting in a maximal posterior glottis of 5 mm in width. Immediately after the surgery, marked dysphonia was noted without any aspiration, and nocturnal stridor was again eliminated dramatically. The vaporized wound epithelialized completely about 2 weeks after the laser surgery (Fig 3B).

Table 1 shows the results of the polysomnography before the first operation, 8 days after the laterofixation of the vocal fold, and 8 days after the laser surgery. They were measured with the use of Nihon Koden Neurofax EEG-5400 (Nihon Koden Co, Ltd, Tokyo, Japan). After the laterofixation, central apnea decreased and the number of apnea attacks during sleep decreased, although the incidence of hypopnea increased in comparison with those before the operation. However, all these parameters showed improvement after the laser surgery.

Table 2 shows the results of the voice examinations before the first operation, 6 days after the surgery, her tracheostoma could be closed completely.

Fig 1. (A) Fiberoptic finding of the larynx during sleep, both vocal folds approximated during inspiration. (B) Fiberoptic finding of the larynx sleep, both vocal folds abducted during expiration.

Fig 2. (A) Laryngeal electromyography during phonation showed normal activity in the left thyroarytenoid muscle. (B) Laryngeal electromyography during phonation showed low-frequency firing in the right thyroarytenoid muscle.
the laterofixation, and 27 days after the laser surgery. Five objective parameters were used for the evaluation of the vocal function: (1) maximum phonation time (MPT), (2) mean airflow rate during phonation over comfortable duration (MFRc), (3) pitch perturbation quotient (PPQ), (4) amplitude perturbation quotient (APQ), and (5) normalized noise energy for 0-4 kHz (NNEa). The details of each parameter have been reported elsewhere.12,13

MPT and MFRc were measured with the use of the Nagashima PS 77H (Nagashima Medical Instruments Co, Ltd, Tokyo, Japan),13 and PPQ, APQ, and NNEa were measured with the use of the Rion SH 10 voice analyzer (Rion Co, Ltd, Tokyo, Japan).12 MPT decreased after the laterofixation and further decreased after the laser surgery. MFRc increased slightly after the laterofixation and further increased after the laser surgery. In addition, PPQ and APQ increased after the laterofixation and further deteriorated after the laser surgery. However, NNEa improved after the laser surgery in comparison with that after the laterofixation. As a whole, voice after the laser surgery was slightly worse (G2R1B2A0S0) than that after the laterofixation (G2R0B2A0S0).

No granuloma has been observed at the surgical site. Her nocturnal stridor was eliminated for these 6 months after the laser surgery.

**COMMENT**

MSA is one of the clinicopathological disease categories that groups together the degenerative diseases including SDS, olivo-ponto-cerebellar atrophy (OPCA), and striatonigral degeneration (Parkinsonisms). Patients with MSA, by definition, show varying symptoms with features of autonomic, extrapyramidal, and cerebellar affections. Although there have been many reports1-7 of bilateral defective laryngeal abduction during sleep in MSA, including SDS, its etiology is still obscure.

Ota et al2 have reported that EMG studies of the laryngeal muscles showed that both adductor and adductor muscles showed continuous firing throughout inspiration and they attributed the cause of vocal folds fixation during sleep to muscle rigidity in the larynx. Isozaki et al1 have analyzed laryngoscopic observations during sleep in 6 MSA patients and

**TABLE 1. Results of the Polysomnography**

<table>
<thead>
<tr>
<th>Apnea Index</th>
<th>Before</th>
<th>After Laterofixation</th>
<th>After Laser Arytenoidectomy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Obstructive Type</td>
<td>—</td>
<td>8.3</td>
<td>0.6</td>
</tr>
<tr>
<td>Mixed Type</td>
<td>—</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>Central Type</td>
<td>27.8</td>
<td>6.2</td>
<td>6.5</td>
</tr>
<tr>
<td>Hypopnea</td>
<td>19.6</td>
<td>38.6</td>
<td>15.7</td>
</tr>
<tr>
<td>Total (apnea)</td>
<td>27.8</td>
<td>14.5</td>
<td>7.1</td>
</tr>
<tr>
<td>Total (apnea + hypopnea)</td>
<td>47.4</td>
<td>53.1</td>
<td>22.8</td>
</tr>
</tbody>
</table>
postulated that vocal fold adduction during inspiration and abduction in expiration during sleep would be attributable to a hypotonic change of laryngeal muscles promoted by sleep. In addition, Tajima et al.\(^3\) have reviewed previous literature and suggested a selective degeneration of the posterior cricoarytenoid muscle as the cause of nocturnal stridor in patients with MSA. In this present case, our clinical observations did not suggest rigidity of the laryngeal structures. On the contrary, a hypotonic change of laryngeal muscles caused by sleep was assumed to be the cause of the suction and approximation of the flabby vocal folds during inspiration, thereby causing the nocturnal stridor. Further investigations, however, are necessary in order to clarify the true nature of this paradoxical movement of the vocal folds during sleep in patients with SDS. It is important, however, to consider the condition of MSA as a differential diagnosis of sleep apnea syndrome because nocturnal stridor is an important presenting system of MSA. Therefore, it is recommended that, when otolaryngologists see a patient who complains of snoring, the possibility of MSA should be taken into consideration.

The treatment modalities for bilateral vocal folds palsy may be divided into 2 categories: one is laterofixation of vocal fold, such as that reported by Woodman et al.\(^4\) or Ejnell et al.\(^8\) and the other is direct volume reduction in the airway, such as endoscopic laser arytenoidectomy\(^9\) or transverse cordotomy.\(^15\)

There is an additional option of using a tracheostomy with an expiratory check valve mechanism, thus releasing the dyspnea and keeping the voice completely unaffected. In the present case, as the vocal fold movement was normal, she refused a permanent tracheal fenestration when she was awake. One of the advantages of Ejnell’s less invasive laterofixation technique is that, when the clinical outcome of this procedure is not so effective, it is possible to cut the thread, thereby making the vocal fold return to the previous condition. In fact, Ejnell et al.\(^8\) reported a case involving one patient who, 3 months after the operation, successfully underwent release of a little of the thread loop to obtain a better voice. Therefore, we initially chose Ejnell’s method for this patient. Unfortunately, after this procedure, the thread broke because of an unexpected laryngospasm, suggesting that laterofixation of vocal fold by the lateral traction with the use of nylon thread may not be suitable for patients with good mobility of vocal folds. For these patients, laser arytenoidectomy would be safer, more reliable, and preferable.

Postoperative polysomnography showed that laser surgery was more effective for stridor treatment than laterofixation. Postoperative aerodynamic and acoustic examination of voice revealed that arytenoidectomy resulted in a wider glottis than the laterofixation. Voice quality, however, was worse after the arytenoidectomy. These results suggested that, for patients with SDS, laser arytenoidectomy should be used as the first choice rather than the laterofixation. Intensive follow-up of the patient is now being done, and she is doing very well in her routine life.

**ACKNOWLEDGMENTS**

This article is supported in part by grants in aid from Ishibashi foundation and for scientific research from the Ministry of Education of Japan.

**REFERENCES**


