PATIENT VIGNETTES

Patient 1: A 55-year-old woman was admitted to our hospital because of progressive parkinsonism. She was diagnosed with striatonigral degeneration. Because of nocturnal snoring, a fiberoptic laryngoscopy was performed during wakefulness, showing a mild abduction restriction of the vocal cords. Arterial blood gas analysis was normal. Over the next year, she developed inspiratory stridor during wakefulness, especially while talking. A second fiberoptic laryngoscopy during wakefulness showed a narrower glottic aperture as compared with the previous examination. Arterial blood gas analysis showed only mild hypoxemia: pH = 7.44, pCO2 = 43 Torr, PO2 = 72 Torr. At this point, she had no dyspnea and could still speak and eat. Only 3 weeks after admission, she was found in a cardiopulmonary arrest in her bed at 8 PM, only 15 minutes after she was heard snoring as usual.

A diagnosis of multiple system atrophy (MSA) was confirmed on postmortem neuropathological examination. The posterior cricoarytenoid muscle—the laryngeal abductor—showed severe neurogenic atrophy. Neither pneumonia nor intratracheal secretions were present to explain her sudden death.

Patient 2: A 74-year-old man, who was diagnosed with MSA 9 years before, was readmitted to our hospital in 1999 because of pneumonia. A fiberoptic laryngoscopy showed no laryngeal abnormalities during wakefulness and diazepam-induced sleep (stage 0). In February 2000, he developed nocturnal snoring. On fiberoptic laryngoscopy, moderately severe vocal cord abductor paresis (VCAP) with abduction restriction during wakefulness and paradoxical movement during sleep was seen (stage 2). The posterior glottis could not be observed well. On an overnight recording of percutaneous arterial blood oxygen saturation (SpO2), no desaturation less than 90% was demonstrated. Arterial blood gas analysis on room air was normal. He was discharged on August 14, 2000, when he was still able to eat, and his nocturnal snoring was not so loud as to disturb other patients in the same room. Only 1 week later, he was readmitted to our hospital because of increasing snoring. Arterial blood gas analysis on oxygen inhalation with 2 l/min when awake showed pH = 7.39, pCO2 = 51 Torr, and pO2 = 88 Torr. On physical findings, his suprasternal recess became hollow during
every inspiration. On August 23, 2000, a fiberoptic laryngoscopy during wakefulness

INTRODUCTION

In 1976, Holinger analyzed 389 patients with vocal cord abductor paresis (VCAP) in various diseases including poliomyelitis, Parkinson’s disease (PD), cerebrovascular accidents, Guillain-Barré syndrome, and multiple sclerosis (1). Spinocerebellar degeneration was not included in his list, and the concept of multiple system atrophy (MSA) was not established at that time. Investigators reported that patients with MSA, including olivopontocerebellar atrophy, striatonigral degeneration, and Shy-Drager syndrome, developed laryngeal complications such as velopharyngolaryngeal paralysis (2), upper airway obstruction (3), and vocal cord palsy (4). In 1981, Bannister reported three necropsied MSA cases in which the posterior cricoarytenoid muscles showed neurogenic atrophy, whereas the nucleus ambiguus, innervating the abductor muscle, demonstrated no neuronal loss (5). Selective neurogenic atrophy of the abductor muscle, among all the intrinsic laryngeal muscles, has been confirmed histologically (2,5,6) and electromyographically (7) in MSA. The myelinated nerve fibers of the recurrent laryngeal nerve (which innervates all the intrinsic laryngeal muscles) are decreased in number (8). However, it is controversial whether the nucleus ambiguus is involved (2,6) or not (5,9). Electromyographical studies support laryngeal dystonia (10,11) or dyskinesia (12) as possible mechanisms of VCAP.

MECHANISM

Although the pathophysiology of VCAP in MSA has not been fully clarified, we propose the following hypothesis (Fig. 1): neurogenic atrophy of the posterior cricoarytenoid muscle, the sole abductor of the vocal cords, is caused by neuronal loss in the nucleus ambiguus. In addition to weakening of the abductor, initiation of abduction becomes delayed. During normal inspiration, the laryngeal abductor muscles contract first, and then the diaphragm contracts to avoid upper airway collapse. However, in patients with MSA and VCAP, inspiratory negative pressure caused by diaphragm contraction occurs concurrent with or even before full opening of the vocal glottis, because of the delay in abductors. This results in laryngeal collapse (13). Paradoxical movement of the vocal cords occurs, with inspiratory adduction and expiratory abduction (14). Sleep enhances VCAP because it increases upper airway resistance (15). In the early stage of VCAP, the stenotic breathing from obstruction in the upper airway is recognized as a snoring only during sleep. Then, in the advanced stage, audible daytime inspiratory stridor occurs, often on talking. This daytime inspiratory stridor can be misdiagnosed as pseudo-steroid resistant asthma (16).