Sleep Apnea and Palatal Myoclonus in a Patient with Neuro-Behçet Syndrome

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A 50-year-old man with neuro-Behçet syndrome showed truncal imbalance, dementia, palatal myoclonus, snoring and rhythmic inspiration synchronized with palatal myoclonus. Magnetic resonance imaging showed hypertrophy of the bilateral inferior olivary nuclei with slight pontine atrophy. A polysomnographic recording disclosed sleep apnea during non-REM sleep: 6% central type, 46% mixed type, and 48% obstructive type. We speculate that lesions of the respiratory center or related structures in the brainstem resulted in sleep apnea and that laryngeal myoclonus also affected the apnea.

Key words: respiration, magnetic resonance imaging (MRI), polysomnography, inferior olivary nucleus

Introduction

Multiple areas of the central nervous system are involved in neuro-Behçet syndrome (NBS), the most common site of involvement being the brainstem (1, 2). Facial palsy, nystagmus, ophthalmoplegia (1) and palatal myoclonus (3, 4) have been reported as clinical manifestations of brainstem lesion. Sleep apnea, however, has not been described in NBS. We here report a patient with NBS who presents palatal myoclonus and sleep apnea.

Case Report

A 50-year-old man admitted to our hospital on December 12, 1988 had a two-year history of gait imbalance and tinnitus of the left ear. He was a truck driver and had fallen from his truck twice because of unsteady gait.

He was a well-nourished, euphoric person. No ocular abnormalities were found by slit-lamp and other ophthalmologic examinations. Close inspection of the skin and mucosa showed follicular facial eruptions, oral aphthous ulcers, and “punched out”-appearing scrotal ulcerations. Neurologic examination disclosed slight dysarthria, dysdiadochokineses and dysmetria of the upper limbs; truncal imbalance; and moderately ataxic gait. Deep tendon reflexes were slightly hyperactive in all limbs without pathologic reflexes. Rhythmic myoclonus of 2-3 Hz was continuous at the soft palate. He snored severely during sleep but did not complain of insomnia or daytime somnolence. Neither obesity nor micrognathia was present.

Peripheral blood counts were normal except for slight leukocytosis. The erythrocyte sedimentation rate was 50 mm/hr; C-reactive protein 4.2 mg/dl (normal range: 0.4 mg/dl<); serum sialic acid 98 mg/dl (50-65 mg/dl); IgA 482 mg/dl (168-306 mg/dl); and the total complement level 53.8 U/ml (30-45 U/ml). Serologic tests for syphilis, antinuclear antibody, rheumatoid factor, and needle reaction were negative. In the cerebrospinal fluid, protein was 29 mg/dl, glucose 49 mg/dl, and the cell count 40/3 mm³ (all lymphocytes). Chest and skull roentgenograms and electroencephalography were all normal. The hearing threshold of the left ear to click stimuli was 40 dB higher than that of the right ear. Auditory brainstem response showed normal interpeak latency between the I, III and V waves for the right ear stimulation but was poorly elicited for the left ear. His WAIS was borderline; total IQ, 78. Biopsy specimens of the scrotal ulcerations showed nonspecific acute inflammation with no vasculitic component.

A CT scan of the brain showed slight enlargement of the ventricular system. Magnetic resonance imaging
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showed slight atrophy of the pons on T1-weighted imaging and increased signal intensity of the enlarged inferior olivary nuclei (ION) on proton-weighted imaging (Fig. 1). Electromyographic recordings of the palatopharyngeal and sternocleidomastoid muscles showed rhythmic discharges of about 3 Hz. Rhythmic small chest movements were synchronized with the palatal myoclonus (Fig. 2A). The flow volume showed multiple small peaks on the V/V curve during inspiration and delayed peaks during expiration (Fig. 2B). A fiberscopic examination of the larynx showed a 2–3 Hz rhythmic spasm of the vocal cord throughout the inspiratory and expiratory phases. No abnormal movements were found by electro-oculography.

On the polysomnographic recording, palatal myoclonus persisted during non-REM sleep but decreased during REM sleep. Sleep apneas were frequent during stage II non-REM sleep but did not occur during REM sleep (Fig. 3). Most apnea periods lasted 20–40 seconds, with 138 apnea periods during the 7.3 hours of his overnight sleep. The apnea index was 18.9. Total sleep apnea consisted of 6% of the central type, 46% of the mixed type (Fig. 4), and 48% of the obstructive type. There was no stage I REM with tonic EMG.

Oral colchicine was administered, but did not improve his neurological status, including the palatal myoclonus.

Discussion

Palatal myoclonus has been occasionally reported in NBS (3, 4). Enlargement of the ION related to palatal myoclonus is thought to be transneuronal degeneration caused by interruption of Guillain-Mollaret’s triangle (5). Only one case of NBS with palatal myoclonus in which MRI showed enlargement of the ION has been reported (6).

Palatal myoclonus often is accompanied by synchronous contractions of the pharynx, larynx, and extraocular muscles that originate from the branchial arches, and is called palato-pharyngo-laryngo-ocular myoclonus (5). Respiratory changes in palatal myoclonus, however, have only rarely been described (3). Nagaoka and Narabayashi (3) recorded rhythmic movements of the thorax during the inspiratory phase, similar to those seen in our patient. They considered that myoclonus of the sternocleido-

Fig. 1. Magnetic resonance imaging with proton-weighted imaging showing increased signal intensity and enlargement of inferior olivary nuclei (arrow). (TR = 2,500 msec, TE = 30 msec).

Fig. 2. Palatal myoclonus and chest movements (A). Electromyographic recording of the palatal muscle showing irregular discharges of about 3 Hz. The small fluctuations in chest movement have the same frequency as the palatal myoclonus. The polygraphic recording shows electromyograms of the palatopharyngeal muscle and chest movements recorded by a transducer (Actgram). The V/V curve of the flow volume (B) shows multiple small peaks during inspiration and delayed peaks during expiration.
Fig. 3. Sleep stages and episodes of apnea. Frequent sleep apneas are present during stage II, but not during REM sleep. Most apnea periods lasted 20–40 seconds.

Fig. 4. Mixed apnea during stage II non-REM sleep. Air flow and chest movements are arrested simultaneously, chest movements recovering earlier. Myoclonic discharges of the submental muscles are continuous. The polygraphic recording shows the EEGs (top 3 channels), electro-oculogram (EOG), electromyogram of the submental muscles (EMG), nasal and oral mixed air flow (Thermist.), diaphragmatic movements (Actgram), and electrocardiogram (ECG).

Sleep apnea in multiple system atrophy in which direct involvement of the medullary respiratory neurons that affect the pattern and generation mechanism may induce central sleep apnea. Moreover, the involvement of the respiratory and non-respiratory motor neurons in the brainstem (e.g., the nucleus ambiguus and hypoglossal nucleus) may cause upper-airway-obstructive apnea. These respiratory neurons mainly are located in the reticular formation of the brainstem.

Polysomnographic recording showed no evidence that the abnormal thoracic movements associated with palatal myoclonus increased just before sleep apnea. More than 50% of our patient's apnea periods were of the central and mixed types. Lesions in the respiratory center or related structures in the brainstem of the NBS, which
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could not be shown by MRI, may have caused the sleep apnea. Laryngeal myoclonus did not induce sleep apnea directly but affected the apnea by increasing of airway resistance. Respiratory abnormalities in patients with NBS, in particular sleep apnea, must be evaluated carefully because sudden death occasionally occurs.

References