Respiratory Myoclonus. Report of a Case with Electromyographic Study

Kenji Jinnai, Keiichi Takahashi, Fumiko Shundo, Yukihiro Komine, Kazuko Gotoh and Takuo Fujita

A case of respiratory myoclonus in a 45 year-old male was studied by electromyography. He complained of anorexia, involuntary swinging movement of the thorax and trunk, and epigastric pulsation. Electromyographic studies showed synchronous rhythmic grouped discharges at the rate of 2 to 3 Hz from the bilateral 8th to 10th intercostal muscles, the right iliocostalis lumborum muscle, and the left hemidiaphragm only during the expiratory phase. They disappeared on inspiration and forced expiration. Anesthetic block of the left phrenic nerve abolished both the voluntary and myoclonic movements of the left hemidiaphragm. Abnormal excitation of the central nervous system and possible irritation of the afferent pathways were postulated.

Key Words: Respiratory myoclonus, Diaphragmatic flutter, Myoclonus

Involuntary clonic contraction of the diaphragm with high frequency has been called diaphragmatic flutter, clonic spasm of the diaphragm, diaphragmatic myoclonus and so on. Since Leeuwenhoek, “father of the microscope”, reported the phenomenon occuring on his own diaphragm1, about 50 cases have been reported2~6. Diaphragmatic flutter with involuntary movements of the other respiratory muscles is rarer and has been called “Respiratory myoclonus”3,7. In this paper, we report a case of respiratory myoclonus studied by electromyography.

REPORT OF A CASE

A forty five year old man was admitted to our hospital on November 16, 1979, complaining of anorexia, involuntary swinging movements of his right lumber region and right lower thorax and epigastric pulsations. He had worked as an electrician. Family history was not contributory. He suffered from diphtheria at 4 years of age. At 9 years of age, he suffered from pulmonary tuberculosis and was occasionally given antituberculous drugs until right upper lobectomy in January 1961. In 1969, he started to feel occasional epigastric pulsations and fasciculation like involuntary muscular contractions in his right lower chest, which used to continue for several seconds to a few minutes. These involuntary movements lacking the thoracic swinging were different from the movements on his admission. In September 1974, he fell from a 10 meter ladder and hit the lumber region and buttock without any disturbances of consciousness nor motor function. Three months later, the involuntary muscular contractions gradually increased in magnitude and frequency, so as to produce lateral swinging movements of the trunk synchronous with the epigastric pulsations.

Physical examination revealed a well-oriented, co-operative Japanese male without any mental abnormalities and in no acute distress. He was 158.5 cm tall and weighted 46 kg. Blood pressure was 102/78 mmHg. No involuntary movements were found in and around the oral cavity. There was an operation scar, 30 cm in length, along the
Respiratory Myoclonus

medial edge of his right scapula. Auscultation of the chest and abdomen revealed splashing sounds on the left hypochondriac area synchronous with the epigastric pulsations. Liver was palpated 2 t.f.b. below the right costal margin. During the quiet expiratory phase, i.e. in relaxation of the respiratory muscles, his trunk rhythmically swung at the rate of 2 Hz with the rapid phase to the left. The left side of the epigastric abdominal wall pulsated synchronously with the truncal swinging. These movements could be suppressed voluntarily by inspiration and forced expiration. They disappeared during sleep and were moderately decreased by compression on the point posterior of the middle part of the left sternocleidomastoid muscle. On the other hand, emotional stress and massage on the operation scar in his back increased them. There were very mild hypesthesia and hypalgesia in the lateral side of the right forearm, the medial side of the left forearm and the lateral side of the lower part of the legs. Vibration sense was also slightly decreased on the bilateral external malleoli. Deep tendon reflexes were normal except for moderately decreased PTR and ATR. Any other neurological abnormalities were not shown.

Laboratory examinations showed no abnormalities. Mantoux reaction was positive without Mycobacterium tuberculosis in sputum, urine nor stool. Cerebrospinal fluid study gave normal values. Chest X-ray revealed a lobectomized lung with healed tuberculosis and normal heart silhouette. A brain CT scan showed slight diffuse cortical atrophy. A myelography showed no abnormalities. A fluoroscopic study of the chest and abdomen revealed that the epigastric pulsations were due to up and down oscillation of the left hemidiaphragm accompanied by no limitation on the voluntary movements of the diaphragm. Electrocardiogram showed the periodic small deflexions which were prominent in leads II, III, aVF, V5 and V6 irrelevant with the heart beats. Spirographic study showed the undulations occurring at a rate of 3 to 4 Hz limited in the expiratory phase. (Fig. 1) Percent vital capacity was 89% and one second forced expiratory volume was 78% of predicted normal.

![Fig. 1. Spirogram. Small undulations occurred at the rate of 3 to 4 Hz during the expiratory phase. The frequency of the undulations were more than that on the quiet expiration.](image)

**ELECTROMYOGRAPHIC STUDY**

Electromyographic study was performed by conventional method using DISA EMG 1500 system. Needle EMG showed that synchronous rhythmic grouped discharges were observed in the bilateral 8th to 10th intercostal muscles (the left side muscles were involved 3 months after admission), the right iliocostalis lumborum

![Fig. 2. Synchronous rhythmic grouped discharges were recorded by needle electrodes from the rt. 8th to 10th I.C.M. and rt. I.C.L.M., but not from the 7th I.C.M. and P.V.M. These discharges were not superimposed on the inspiratory discharges. I.C.M.; intercostal muscle, I.C.L.M.; iliocostalis lumborum muscle, P.V.M.; paravertebral muscle.](image)
Fig. 3. Electromyogram of the diaphragm recorded by needle electrodes. In the left hemidiaphragm, rhythmic grouped discharges occurred during the expiratory phase synchronously with that of the right iliocostalis lumborum muscle, but not in the right hemidiaphragm. The discharges were not superimposed on the activities of the respiratory muscles during the inspiratory and forced expiratory phase. (Fig. 2, 3) Motor unit action potentials of these muscles and the interference patterns were all normal. Nerve conduction velocities (MCV and SVC of the bilateral N. ulnaris and N. medianus, MCV of the bilateral N. tibialis posterior and SCV of the bilateral N. suralis) were normal except for MCV of the bilateral peroneal nerves (right 37.6 m/s, left not evoked). The anesthetic block of the left phrenic nerve in the supraclavicular area by 2% procaine hydrochloride effectively stopped both the voluntary and involuntary movements of the left hemidiaphragm for about 30 minutes. The latency time of the left phrenic nerve was 8.1 ms (normal value; 8.2 ± 0.72 ms)\(^8\). The distance between the stimulating point and the diaphragm was 300 mm.

**COURSE AND THERAPY**

In January 1980, similar synchronous movement appeared in the left intercostal muscles. He was given diphenylhydantoin, phenobarbital, and diazepam with little effect for the involuntary movements. Oral administration of carbamazepine (400 mg/day) was considerably effective for the involuntary movements of the intercostal and iliocostalis lumborum muscles, but not for that of the diaphragm. The effect decreased several months later and he complained of anorexia because of the epigastric pulsations which had become more frequent and violent. When he was on clonazepam with daily dosis of 6 mg, the myoclonus was alleviated. But this had to be discontinued because of severe sleepiness. Recently, the swinging movements of the trunk disappeared and the epigastric pulsations became less violent without any drugs.

**DISCUSSION**

The different clinical patterns of myoclonus may be divided into two categories. In the one the contractions are irregular in time, intensity and distribution. In the other the contractions are rhythmic, constantly recurring in certain muscles and their intensity is mostly rather regular\(^9\). The latter contains spinal myoclonus, palatal myoclonus, diaphragmatic flutter, respiratory myoclonus and so on. Respiratory myoclonus, which is a combination of diaphragmatic flutter and the synchronous contractions of the intercostal and accessory muscles, is fairly rarer than diaphragmatic flutter about 50 cases of which have been reported. We found only two definite cases of respiratory myoclonus in the literature\(^3\).\(^7\).

Hiccup is easily differentiated from the respiratory myoclonus by the lower frequency and accompanying glottal closure in the former\(^10\). Diaphragmatic flutter is frequently accompanied by passive thoracic movements which may be correctly diagnosed by electromyographic study\(^11\). Palatal myoclonus, which is attributed to the lesion in the triangular area formed by the red nucleus, inferior olive on one side and the contralateral dentate nucleus, is occasionally accompanied by synchronous myoclonus of several muscles containing the diaphragm or rarely the intercostal muscles, called palato-pharyngo-laryngo-oculo-diaphragmatic myoclonus\(^12\). A case of diaphragmatic flutter was reported to be associated with anesthesia of the pharyngolaryngeal region and horizontal nystagmus\(^5\). These cases
Respiratory Myoclonus

suggest a close relationship of the palatal myoclonus to the diaphragmatic flutter of central origin and the respiratory myoclonus. In our cases, no abnormal movement was noted in the palatal region. In the two previously reported cases, the lesions of respiratory myoclonus were assumed to be in the central nervous system, since the case reported by Phillips and Eldridge showed posthyperventilation apnea and the case described by Mochizuki et al was accompanied by ballism like movement. The cause of the lesions was unclear in their cases. Diaphragmatic flutter has been attributed to abnormal excitation of the phrenic nerve, either by disturbance of the central nervous system or by irritation of the phrenic nerve or diaphragm itself. The most common central nervous cause is epidemic encephalitis. The cases with psychogenic origin and electrocution were also reported. The reported peripheral sources of irritation of the phrenic nerve are cervical rib, enlarged mediastinal lymphnodes, pleurisy, peritoneal adhesions, fractured xiphoid process and heart beat as reviewed by Rigatto and Medeiros. In our case, the effective anesthetic block on the phrenic nerve seemed to have excluded the lesion of the peripheral part of the phrenic nerve or the diaphragm itself. Since older times it has been reported that the diaphragm is innervated not only by the phrenic nerve but also by the lower thoracic motor neurons. Recently, Uono et al documented that the diaphragm of the cat was innervated by homolateral motor neurons. In this patient, however, the successful block on the phrenic nerve seemed to have denied participation of such an extraphrenic innervation in the respiratory myoclonus.

Some of the patients with respiratory myoclonus or diaphragmatic flutter were accompanied by lung disease or surgical intervention to the lung, which might cause abnormalities of the afferent system from the lung to the respiratory center. The previous lobectomy could be one of the precipitating factors in this patient. The myoclonus of this case was temporarily suppressed by inspiration and forced expiration. Similar phenomenon was observed in palatal myoclonus and in some cases of diaphragmatic flutter. However, in most cases of diaphragmatic flutter, attempts to suppress the flutter by breath holding were ineffective. In this patient the myoclonus was suppressed to some extent by the pressure on the left side of the neck. Bruni noticed similar suppression by the pressure on the supraclavicular area. Emotional tension and excitement are known to be the most common precipitating factors. The effect of massage on the operation scar in this patient could be either psychogenic or influence on abnormally excited afferent path in a possibility. Fox reported a patient in whom pressure over a dental cyst could start the diaphragmatic flutter.

Though we were unable to identify the lesion or the etiology in this patient, an abnormal excitation in the central nervous system causing irritability of the afferent pathway is probably responsible for this respiratory myoclonus.

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