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SHORT COMMUNICATION/COMMUNICATION BRÈVE

The presence of spontaneous EMG activity in sternocleidomastoid is associated with ventilatory dysfunction in ALS

La présence d'une activité EMG spontanée dans les muscles sterno-cléido-mastoïdiens est associée à un dysfonctionnement ventilatoire dans la SLA

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Summary We investigated electromyography (EMG) of the sternocleidomastoid (SCM) in 128 patients with amyotrophic lateral sclerosis (ALS) including correlation with forced vital capacity (FVC) and ALS Functional Rating Scale scores. The presence of fibrillation potentials and positive sharp waves in the SCM was significantly more frequently observed in patients with an FVC < 80% (31/49, 63%) than in patients with an FVC ≥ 80% (34/79, 43%). This study suggests that the SCM could be concomitantly involved with primary respiratory muscles in ALS. This is of value since needle EMG is a less invasive procedure in SCM than in the diaphragm or intercostal muscles.

Résumé Nous avons étudié l'activité électromyographique (EMG) des muscles sterno-cléido-mastoïdiens (SCM) chez 128 patients atteints de sclérose latérale amyotrophique (SLA), y compris les corrélations avec la capacité vitale forcée (CVF) et les scores des échelles d'évaluation fonctionnelle de la SLA. La présence de potentiels de fibrillation et de potentiels positifs à front raide dans les SCM était significativement plus fréquente chez les patients avec une CVF < 80% (31/49, 63 %) que chez les patients avec une CVF ≥ 80% (34/79, 43 %). Cette étude suggère que l'atteinte des SCM pourrait être concomitante de celle des muscles respiratoires principaux dans la SLA. Ceci est à souligner car la détection EMG à l'aiguille est une procédure moins invasive dans le SCM que dans le diaphragme ou les muscles intercostaux.

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Introduction

The sternocleidomastoid (SCM) muscles are currently regarded as alternative muscles for investigating the bulbar area in patients with amyotrophic lateral sclerosis (ALS) [8,9]. The SCM muscles are innervated by lower motor neurons of the lower medulla oblongata and the C2—C3 anterior horn [2,7], and the diaphragm is innervated by lower motor neurons of the C3—C5, which are primarily located in the C4 anterior horn [11]. The two regions are tightly adjoined, and their motor neurons may be concomitantly involved in ALS. As axial muscles, the lower motor neurons of the SCM are more medially situated in the anterior horn, similar to those of the diaphragm, compared with those innervating the distal muscles of the limbs [5]. Pinto et al. found that SCM compound muscle action potential (CMAP) amplitudes, diaphragm CMAP amplitudes and many respiratory function measurements, including forced vital capacity (FVC) and ALS Functional Rating Scale (ALSFRS), were significantly different in a subgroup of ALS patients with neck weakness compared with a group without neck weakness. Their findings suggest that there is a parallel loss of motor units in the SCM and diaphragm [12].

To further investigate denervation of the SCM and its potential role in enabling judgment of ventilatory dysfunction and prognosis in Chinese patients with ALS, we studied needle electromyography (EMG) changes of the SCM and their correlation with FVC and ALSFRS.

Patients and methods

We enrolled 128 patients with sporadic ALS from Peking University Third Hospital from 2009 to 2013. All patients were examined with detailed neurological, imaging, electrophysiological, and laboratory investigation, and other diagnoses were excluded. Fifty-two patients were diagnosed with definite ALS and the remaining patients were diagnosed with probable ALS according to the revised El Escorial criteria [3]. The ethical committee of Peking University Third Hospital approved the present study. All patients signed consent forms after the study design was fully explained.

ALSFRS, FVC and EMG measurements were performed on all patients. The ALSFRS and FVC measurements were obtained concurrently with EMG measurements. An FVC value less than 80% was considered abnormal [1,13]. EMGs were recorded from the bulbar, upper limb, thoracic, and lower limb muscles, including the SCM, first dorsal interosseous, rectus abdominis, and tibialis anterior muscles, with a concentric needle electrode using standard settings (Keypoint; Medtronic, Skovlunde, Denmark). Patients lay calmly in a relaxed state while spontaneous activity (fibrillation potentials and positive sharp waves; fib-psw) was investigated. In each muscle, we searched for spontaneous activity at two sites in each of four different insertions. Spontaneous activity was considered as present if observed in reproducible trains after at least 300 ms following needle insertion, and was regarded as pathological only when waves were identified at more than two sites in a muscle. The motor unit action potential (MUAP) configuration was assessed during a slight voluntary contraction. Recruitment patterns of motor units were evaluated during maximal voluntary contraction. The EMG measurements of the SCM were performed in accordance with previous reports [8,9]. Usually SCM was first investigated on the same side as the limb which had more severe weakness or amyotrophy. SCM in the other side would be performed if EMG of the first chosen SCM was normal.

All 128 ALS patients were separated into two groups based on ALSFRS respiratory subset scores. Thirty-three cases of the patients exhibited an ALSFRS respiratory subset score of less than 12. The relationship between fib-psw of the SCM and ALSFRS, FVC and dyspnoea was evaluated.

Values are expressed as means $\pm$ standard deviations (SDs). Independent *t*-tests and chi-square tests were used for continuous variables and Fisher's exact test was used for discrete variables. We considered a value of $P < 0.05$ as statistically significant.

Results

Clinical characteristics

The 128 ALS patients had a mean symptom onset age of 51.6 ± 10.9 years (range 24—80 years) and included 79 males and 49 females with a mean diagnostic delay from symptom onset of 16.6 ± 12.6 months (range 3—81 months). The disease exhibited bulbar onset in 17 patients, upper limb onset in 73 patients, and lower limb onset in 38 patients. The mean FVC was $83.4 \pm 17.1\%$ (range 45—131%) of the predicted value, and the mean ALSFRS score was 40.0 ± 5.8 (range 15—47). FVC values $\geq 80\%$ were found in 79 patients, and FVC values $< 80\%$ were found in 49 patients (Table 1). There were no significant differences in age of onset, diagnostic delay, gender, and first affected anatomical site between the two subgroups. The ALSFRS scores of the former group were significantly different from those of the latter group ($P < 0.001$).

Electromyography

Fib-psw of the SCM were observed in 65 patients (65/128, 50.8%), including 34 patients with an FVC $\geq 80\%$ (34/79, 43.0%) and 31 patients with an FVC $< 80\%$ (31/49, 63.3%) ($P < 0.05$, Table 1). We further compared the FVC values and ALSFRS scores between patients with and without fib-psw of the SCM. The FVC values were lower in patients with compared with those without fib-psw of the SCM ($P = 0.012$). However, there were no significant differences in ALSFRS scores between the two groups ($P > 0.05$). Neurogenic changes in SCM MUAPs were detected in 29 patients, including 17 patients with an FVC $\geq 80\%$ and 12 patients with an FVC $< 80\%$. The frequency of large SCM MUAPs and the increase in MUAP durations and amplitudes did not differ between the groups with and without an abnormal FVC ($P > 0.05$).

Discussion

In this study, we found that the levels of SCM spontaneous potentials were increased in ALS patients with an FVC $< 80\%$ compared with those with an FVC $\geq 80\%$. Our study indicates