THE ELECTROMYOGRAPHIC, PHYSIOPATHOGENETIC EVALUATION

OF AMYOTROPHIC LATERAL SCLEROSIS

P. Pinelli

1st Neurological Clinic, University Medical School
S. Paolo Hospital
Milan, Italy

In our present state of knowledge, we lack a specific definition and clinical classification of idiopathic ALS. Its borders vanish among different forms of spinal muscular atrophy (SMA) or lateral sclerosis[1]; moreover its differentiation from hereditary ALS, phenocopies and exogenous similar syndromes is hardly possible except by the identification of the respective etiologies. A pathognomonic marker of primary sporadic ALS seems to be represented by the increase in c.s.f. of thiamine/thiamine monophosphate (T/TMP). On the other hand a goal-directed electromyographic investigation complete with histochemical, neurobiological and radiological examinations can allow a precise study of the single motor neuron in its whole life-span during the course of the disease. If such a longitudinal investigation could be carried out in patients affected by different types of motor neuron pathology, specific aspects of the neurobiological processes occurring in ALS could be better defined and their relationship with the physiopathogenesis of the disease could probably be discussed.

The aim of the present study is to identify the main disorders in motor unit size and their recruitment occurring in ALS by means of EMG investigations carried out over a 3 year period in 2 groups of ALS patients with increased T/TMP in c.s.f. and normal T/TMP, respectively[2].

Subjects and Methods

Follow-up EMG investigations were performed in 68 muscles of 20 ALS patients with T/TMP in c.s.f. > 1 and in 15 muscles of 4 ALS patients with T/TMP < 1 (that is, the normal value of T/TMP). Four patients with spinal chronic amyotrophy (SMA), 2 with Friedreich's disease, 1 with Strumpell spastic paraplegia, 1 with Mills hemiplegia, 1 with lead poisoning, and 1 with parainfective myelitis (all with normal T/TMP) underwent the same investigations. Ten normal subjects were also investigated with the same technique.

We used a Medelec electromyograph (MS 92a) and the action potentials were recorded with surface skin electrodes (DISA) or with Adrian concentric needle electrodes. Changes in tension for single motor units (MU) and for whole muscle were recorded through transducers and measured according to previously defined methods[3].
Fasciculations were recorded as mechanical displacement of the muscle and their site of origin investigated with previously reported methodologies[4]. Criteria for identifying synchronism of different MUs during voluntary recruitment were analyzed in a previous study[3]. The trophic state of a group of muscles was determined by means of the amount of water displaced in containers where the distal segment of the limb was placed; the most systematical research was carried out for the forearm; preliminary research with CAT of the muscle are programmed.

Results

1. Time of Occurrence of Amyotrophy

The measurements of forearm volume in clinically normal arms were carried out in 4 ALS patients who underwent the examination when other muscular districts were clinically affected: two patients presented with polyneuropathic type of Patrikios and two other patients with the bulbar type. The extensor and flexor muscles of wrist and finger showed normal parameters by clinical and electromyographical investigation carried out over a period of six months. In this period the forearm volume was measured twice; at the second investigation we found a decrease of 20% in three and 30% in the fourth case (Patrikios syndrome); this last result was not associated with any decrease in the body weight of the patient. Unfortunately the T/TMP ratio could not be measured.

2A. Spontaneous MU Activity in Clinically Normal Muscles

(i) ALS patients with c.s.f. T/TMP > 1. In 26 muscles of 13 patients with normal strength during a 2 year period preceding the first stage of paresis, we found true spontaneous activity of MU a.p.s with amplitude often (14 among 32) exceeding 2 mV with frequency of discharges between 2 - 6 seconds independent of any kind of reflex activation. The ectopic site of origin, taking into account the relation of fasciculations with M evoked impulse and the Roth's F response of spontaneous discharges, could probably be localized at a proximal point of the motor neuron.

(ii) ALS patients with c.s.f. T/TMP < 1. In clinically normal muscles spontaneous MU activity was found only once in 1 out of 6 muscles during a 1 year period of study. The frequency of discharge was less than 1/second and their amplitude never exceeded 2 mV; no mechanical equivalent of the MU a.p.s. were detected with the clinical observation.

2B. Fasciculations in Paretic Muscles (period of 1 year)

The frequency of discharges was less than 1/second. In both groups of ALS patients in moderately weak muscles, fasciculations were evident at clinical observation; the corresponding action potentials were characterized by an amplitude > 2 mV or by several linked potentials. In a few moderately weak muscles of SMA patients (2 out of 11) a peculiar finding was obtained in the occurrence of spontaneous activity of giant MU with a distal site of origin, with repetitive discharges at a rather high rate of frequency (14-20/second).

3. Decrease in the Number of Recruitable MUs

Decrease in muscular strength with corresponding decrease in M mx response was found in moderately weak muscles without any sign of denervation in examinations carried out at two to six month intervals. Since the decrease in M amplitude and duration was also observed with stimulation of the most distal point of the nerve, a block of conduction affecting the