Vol. 118, n. 2 (Supplement): 91, 2013

## Neurons but Motor Neurons in Motor Neuron disease

<u>Francesco Fornai</u><sup>1</sup>, Paola Lenzi<sup>1</sup>, Michela Ferrucci<sup>1</sup>, Stefano Ruggieri, Loredana D'Este<sup>4</sup>, Antonio Pellegrini<sup>1</sup>, Lorenzo Fumagalli<sup>5</sup>, Francesco Giannessi<sup>1</sup> and Antonio Paparelli<sup>1</sup>

<sup>1</sup>Human Anatomy Department of Translational Research and New Technologies in Medicine and Surgery, University of Pisa, 56126 Pisa, Italy

<sup>2</sup> Neuronal Sciences, Sapienza University of Rome, Rome, Italy

<sup>3</sup>I.R.C.C.S. Neuromed, Pozzilli, Italy

<sup>4</sup> Laboratory of Immunohistochemistry Tindaro G. Renda, Department of Anatomic, Histologic, Forensic and Locomotor Apparatus Sciences, Sapienza University of Rome, Rome, Italy

<sup>5</sup> Department of Anatomy, Histology, Forensic Medicine and Orthopedics, Sapienza University of Rome, Rome, Italy

The occurrence of motor neuron death is the milestone of amyotrophic lateral sclerosis (ALS). Therefore, morphological analysis along decades focussed on motor neuron loss as the sole marker to score disease severity.

Recently, non autonomous cell death took a prominent role to explain the need for additional cell types to induce motor neuron degeneration in ALS. This concept encompasses a variety of inflammatory cells including resident astroglia and microglia.

Despite tremendous clinical and basic research activity the role of inflammation remains controversial. On the other hand very recently non autonomous cell death creped back in considering neuronal cell types differing from motor neurons.

In the present communication evidence is provided demonstrating that interneurons and other neuronal cell types participate to degeneration. Remarkably, this multi-neuronal non-autonomous motor neuron degeneration extends to mutiple models of motor neuron disorders. Interneurons degenerate earlier and more than motor neurons, thus suggesting a key role in the pathophysiology of motor neuron disease. Additionally, these findings portray ALS as a whole spinal cord disorder rather than a disease affecting solely motor neurons. Novel concepts of abnormal cell to cell communication are likely to underlie multi-neuronal disease spreading.

This work was supported by research grant PRIN 2010-2011 FF

Key words

Non authonomous cell death, motor neuron Ranshow cell, ALS.