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ABSTRACTS

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SOCIETÀ ITALIANA DI NEUROLOGIA
Background: In the last decades various studies have showed that cognitive and behavioural disorders are frequent in ALS.

Objectives: To delineate a specific cognitive and behavioural profile in patients with early ALS. Materials and Methods: sixteen patients with probable or definitive ALS disease, according to El Escorial (1998) criteria, were tested with an extensive neuropsychological testing. We have included patients with 0-2 years history disease. The patients were recruited in order of diagnosis to avoid an inadequate selection. A behavioural scale (The Neuropsychiatric Inventory) and a depression scale (Hamilton Rating Scale for Depression) were administered.

Results: In the 40% of patients at least four tests, especially frontal tests, were impaired. FAB (The Frontal Assessment Battery) was the more sensible test to diagnose a cognitive impairment. All the patients with cognitive impairment had a mild cognitive impairment and two had a dementia. Apathy was the more frequent behavioural disorder.

Discussion and Conclusions: Cognitive and behavioural disturbances are frequent in patients with early ALS disease. Dysexecutive syndrome is the typical cognitive pattern in patients with ALS. The FAB is a sensible cognitive screening instrument in these patients.

References:

VERY SLOW PROGRESSION OF CLINICALLY DIAGNOSED BEHAVIOURAL VARIANT OF FRONTOTEMPORAL DEMENTIA

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Introduction: The behavioural variant of Frontotemporal dementia (bv-FTD) is a neurodegenerative disease characterised by early and progressive changes in personality and behaviour, such as apathy, reduced empathy and lack of the usual social mores, or disinhibition, agitation, irritability and stereotypical and ritualized behaviours (Hodges and Miller, 2001). Imaging changes in frontotemporal regions are supportive but not mandatory for the diagnosis (Neary et al., 1998). Recently, Kipp et al. (YEAR), compared two groups of patient with a clinical diagnosis of bv-FTD. At the onset of disease the first group showed an absence (or mild) brain atrophy and hypometabolism, while the second group showed a definite frontotemporal hypometabolism in FDG-PET analysis. These patient were followed for 6 years, and a lack of progression was observed in the first group. In contrast, patients with significant frontal atrophy of hypometabolism at onset showed a clear-cut deterioration over time. The authors suggest that these findings support the existence of a non-degenerate phenotype of the disease.

Material and methods: We describe three patients who met the criteria for bv-FTD (Neary et al., 1998); no patient had clinical or neuroradiological evidence of cerebrovascular disease, or a possible history of psychiatric disorder or alcohol abuse. CT scan of all patients at the onset of the disease was normal, while SPECT imaging showed a significant hypoperfusion in frontotemporal regions. Neuropsychological evaluation showed a profile characterized by executive function deficits in all patients. Memory and visuo-spatial abilities were also impaired, possibly as a consequence of attentional and executive disorders. All patient were regularly followed over a period of 10 years (2000–2009).

Results and conclusions: Neuroimaging studies showed a significant progression of atrophy and hypoperfusion in frontotemporal areas. Neuropsychological evaluation showed a mild and aspecific progression of the deficits. The clinical and functional status was stable over time. In particular, the behavioural disturbances remained relatively stable in the type and frequency of symptoms, while activities of daily living were spared. These findings suggest the existence of a variant of bv-FTD characterised by very slow clinical progression.

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