P6

Application of the MDS diagnostic criteria to the large cohort of Italian PSP patients: results from the PSP-NET

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Introduction: Supranuclear Progressive Palsy (PSP) is a rare, rapidly progressive, neurodegenerative disease characterized by dysfunction in four core domains including ocular motor function, postural instability, akinesia, and cognition. PSP-NET is the Italian registry of PSP patients promoted by the LIMPE Foundation.

Objectives: The aim of the present study is to describe the preliminary motor data collected until early March 2022.

Methods: We analyzed the demographic and clinical features of the PSP patients enrolled. Difference between PSP subtypes were computed with ANOVA test and post hoc analysis.

Results: A total of 261 PSP patients were evaluated (75 from the North; 47 from the Center and 139 from the South of Italy). One-hundred and thirty-two (50.6%) were women, mean age was 59.67 (13.09) and mean disease duration was 2.89 years (2.05).

Applying previous National Institute of Neurodegenerative Disorders and Stroke-PSP criteria (NINDS-PSP), that recognize PSP-RS as the only form of disease, 229/261 (87,7%) fulfilled PSP diagnosis of whom 104 (39.84%) reached a probability and 125 (47.89%) a possibility level of diagnostic certainty.
According to the Movement Disorder Society-PSP (MDS-PSP) criteria, 236 subjects qualified for probable PSP (90.4%), 6 for possible PSP (2.29%) e and 7 for suggestive of PSP (2.68%). Two-hundred and twelve (81.2%) were PSP-Richardson Syndrome (PSP-RS), 19 (7.27%) were PSP with predominant parkinsonism (PSP-P) and 19 (7.27%) were among the other variants of the disease (vPSP). Mean total PSP-rating scale (PSP-rs) was 39.38 (18.53). PSP-rs was higher in PSP-RS compared with vPSP ($p<0.01$) but not with PSP-P ($p=0.65$).

Conclusions: The MDS-PSP criteria enlarge the number of patients fulfilling PSP diagnosis compared to previous criteria. As expected, the most prevalent phenotype was PSP-RS [1] followed by PSP-P with the remaining phenotypes accounting for only 7%. Finally, PSP-RS showed a greater disease severity compared to all other phenotypes but not to PSP-P.

References:

[1] K Peikert, J Linn, MD Brandt and A Hermann. MDS criteria for the diagnosis of progressive supranuclear palsy overemphasize Richardson syndrome. Ann Clin Transl Neurol. 2020 Sep; 7(9): 1702–1707.