p-309

Friday

Clinicopathological correlations in frontotemporal lobar degeneration: results from a French cohort with deep clinical and behavioural phenotyping

Thibaud Lebouvier, Mélanie Leroy, Baptiste Hallemieux, Grâce François, Vincent Deramecourt, Mira Didic, Daniele Seilhan, Frédérique Etcharry Bouyx, Isabelle Le Ber, Franck Letournel, André Maues de Paula, Claude-Alain Maurage, Sophie Mohr, François Sellal, Marie-Anne Mackowiak, Maxime Bertoux, Florence Pasquier

Sporadic forms of frontotemporal dementia are still the most frequent. In the absence of pathophysiological biomarkers, clinicopathological correlations remain the only tool at our disposal to predict the underlying pathology. In this retrospective clinicopathological study, we performed a deep clinical and behavioural phenotyping of autopsy-proven frontotemporal lobar degeneration (FTLD) patients followed in the French network of tertiary referral memory clinics.

A total of 120 patients were included (73 from Lille-Bailleul, 18 from Paris Pitié-Salpêtrière, 14 from Colmar, 10 from Angers and 5 from Marseille). Sixty (50.0%) cases were FTLD-TDP, 53 (44.2%) FTLD-Tau and 6 (5.0%) FTLD-FUS. FUS cases were significantly younger and argyrophilic grain disease (AGD), corticobasal degeneration/progressive supranuclear palsy (CBD/PSP) patients were significantly older. FUS cases had the lowest survival and AGD cases had the longest. TDP-A/B patients had a significantly higher prevalence of family history. The distribution of the symptoms at first referral was significantly different between pathologies. Language complaint was significantly more frequent in TDP-C, cognitive complaint in AGD and psychiatric symptoms in FUS. Symptoms at any point during the disease differed as well: as expected motoneuronal pathology was only observed in TDP-A/B cases. However, deep phenotyping of behavioural symptoms did not retrieve meaningful differences between pathologies, nor did assessment of executive, memory and visuospatial functions. Alzheimer's disease was the most frequent misdiagnosis, mostly in AGD cases, followed by psychiatric disorders (mostly in FUS cases).

This study brings some incremental knowledge on clinicopathological correlations in FTLD, which will become an outstanding issue when disease-modifying drugs are available.

Conflicts of interest

None in relation with this presentation