TÜRK NÖROLOJİ DERGİSİ TURKISH JOURNAL OF NEUROLOGY

Kognitif III - Uluslararası Kognitif Nörobilim Sempozyumu /
Cognitive III - International Cognitive Neuroscience Symposium

Türk Nöroloji Dergisi 2006; Cilt:12 Sayı:4 Ek:3 Sayfa:55

15:15-16:00

The Evolution of Frontotemporal Dementia

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The diagnosis of Frontotemporal dementia (FTD/Pick complex) remains challenging despite of several recent clinicopathological and epidemiological studies. The relationship of the clinical presentations is not clear or even denied, resulting in nosological arguments and controversy. The clinical consensus criteria of Neary et al.(1998) use FTLD for the overall designation of the behavioural and aphasic manifestations, but leaves out the extrapyramidal syndromes of CBD or Progressive Supranuclear Palsy (PSP). This remains one of the most controversial aspects of the nosological considerations.

The tau-ubiquitin dichotomy in FTD pathology is becoming standard, although voices of caution in favor of clinical and even pathological unity are being raised. The purpose of this study is to follow a substantial cohort of FTD/Pick complex patients prospectively to clarify the issues in diagnosis, the relationship of the clinical presentations and the evolution of the illness. We aimed to quantitate the association of certain syndromes that appear to suggest a biologically based dichotomy and also the extent of their eventual overlap. Although the emphasis is on the longitudinal clinical study, substantial pathological material is also updated and discussed.

Patients with behavioural variety (FTD-bv) developed progressive aphasia (PA) in over half, and semantic dementia (SD) and coticobasal (CBDS/PSP) syndrome in smaller numbers. Primary progressive aphasics in turn often developed FTD-bv and CBD/PSP. Triple syndromes were relatively common. A significant association of SD with FTD-bv and CBDS/PSP with PPA was observed. The frontal behavioural inventory showed high specificity, sensitivity and predictive value in FTD-bv diagnosis. Visuospatial function was preserved except in CBDS/PSP. Survival and sex distribution was similar in all groups. Clinical diagnosis showed a sensitivity of 100% and specificity of 76.9% against autopsy.
Conclusion: Diagnostic and nosological confusion is reduced when the evolution and relationships of the syndromes of FTD/Pick complex is quantitated. The clinical associations follow the tau vs ubiquitinopathy distinction, but there is too much overlap for a definite dichotomy.