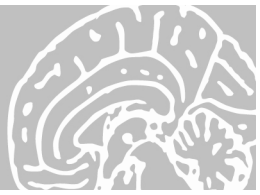
Frontotemporal Lobar Degeneration



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Frontotemporal dementia (FTD) is a heterogeneous group of clinical and pathological syndromes which begins with behaviroal or language symptoms. There are three clinical subtypes of FTD as follows: behavioral variant of frontotemporal dementia (bvFTD), semantic variant primary progressive aphasia, nonfluent/agrammatic variant primary progressive aphasia. The FTD has frontotemporal lobar degeneration (FTLD) pathology which is divided as four major histopathological subcategories: FTLD-tau, FTLD-TDP (TAR-DNA binding protein), FTLD-FUS (Fused in sarcoma), FTLD-UPS (Ubiquitin proteasome system). Of these, most patients with FTD have FTLD-tau and FTLD-TDP. The FTLD-tau consists of Pick's disease, progressive supranuclear palsy, corticobasal degeneration (CBD), and argyrophilic grains disease and the FTLD-TDP can be sub-classified as type A, B, C, and D based on the morphological apperance and cellular locaion of the abnormal TDP inclusions. Here, the first autopsy-confirmed Korean cases of bvFTD and FTD with motor neuron disease whose pathological diagnosis were CBD of FTLD-tau and FTLD-TDP type B, respectively, will be presented.