Neuropsychological Profiles in the Diagnosis of Dementia

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Thirty years ago, neurodegenerative dementia was considered a neuroanatomically and functionally diffuse class of diseases, and, therefore, of no importance to understanding cognitive localization in the brain. Public awareness of neurodegenerative dementia has increased dramatically over the past 30 years, leading to much earlier diagnosis. In early stages of dementia, it has been shown that neurodegenerative changes are highly selective, targeting very specific cortical and subcortical regions and producing highly circumscribed cognitive deficits before they evolve into more generalized impairment. Four distinctive neuropsychological profiles of dementia, each associated with a different large-scale neuroanatomical network, will be contrasted with respect to their salient clinical features and associated neuroanatomical and neuropathologic signatures. An amnestic profile has been associated with medial temporolimbic dysfunction and most of these cases are diagnosed with pathologic Alzheimer’s disease at post mortem autopsy. The profile of primary progressive aphasia has been associated with structural and functional disruption in left perisylvian “language regions” and is neuropathologically heterogeneous with most cases due to one of the several forms of frontotemporal lobar degeneration (FTLD). An early profile of progressive visuospatial dysfunction has been associated with posterior cortical atrophy and hypometabolism in visual processing regions of the brain. At post mortem, this clinical profile is primarily linked with Alzheimer neuropathology in a distribution that favors visual association cortex. Finally, the syndrome marked by early progressive comportmental and executive function deficits is marked by frontotemporal atrophy and dysfunction and, at post mortem, with one of the many entities under the rubric of FTLD. The neurocognitive profile approach to dementia has led to important discoveries about how cognition is organized in the brain as well as to more effective diagnosis and management of these illnesses.