

Does corticobasal degeneration exist as a clinicopathological entity?

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Corticobasal degeneration has been described in the late 1960's as "corticodentatonigral degeneration with neuronal achromasia" in three patients presenting parkinsonism and involuntary motor activity. All patients showed asymmetric degeneration of the perirhinal and olfactory cortices, basal ganglia and substantia nigra. In most cases, the affected cortices featured gliosis and ballooned neurons. From these reports, others followed with alternative nomenclature, but similar clinicopathological features. By the late 1980's, when the term corticobasal degeneration (CBD) received almost universal acceptance, CBD was considered a unique clinicopathological entity, meaning an almost perfect correlation between the clinical features with a particular neuropathological entity. However, with the advent of immunohistochemistry for tau protein, it became evident that different pathological entities including Alzheimer's disease, frontotemporal lobar degeneration with tau inclusions of the Pick's or PSP-type could underlie a "CBD" clinical presentation. Moving forward, the term corticobasal syndrome (CBS) replaced CBD as a clinical diagnosis and the term CBD was reserved to describe a distinctive 4-repeat tauopathy with an involvement of the gray and white matter. Recent clinicopathological series report that about 1/3 of patients developing CBS show CBD. The remaining present other entities, especially Alzheimer's diseases. Ongoing studies are focusing in refining CBS clinical classification to enable better antemortem prediction of the underlying pathology in these patients.