CORTICAL-BASAL GANGLIONIC DEGENERATION

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Neurology 1990;40:1203-1212

We report our experience with 15 patients believed to have cortical-basal ganglionic degeneration. The clinical picture is distinctive, comprising features referable to both cortical and basal ganglionic dysfunction. Characteristic manifestations include cortical sensory loss, focal reflex myoclonus, "alien limb" phenomena, apraxia, rigidity and akinesia, a postural-action tremor, limb dystonia, hyperreflexia, and postural instability. The asymmetry of symptoms and signs is often striking. Brain imaging may demonstrate greater abnormalities contralateral to the more affected side. Postmortem studies in 2 patients revealed the characteristic pathologic features of swollen, poorly staining (achromatic) neurons and degeneration of cerebral cortex and substantia nigra. Biochemical analysis of 1 brain showed a severe, diffuse loss of dopamine in the striatum. This condition is more frequent than previously believed, and the diagnosis can be predicted during life on the basis of clinical findings. However, as with other "degenerative" diseases of the nervous system, a definitive diagnosis of cortical-basal ganglionic degeneration requires confirmation by autopsy.

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Comment from Ryan J. Uitti, MD, FAAN, Associate Editor: This manuscript provided a major contribution to the description of a newly recognized clinical entity: cortical basal ganglionic degeneration.

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Neurology 2011;76;1634

DOI 10.1212/01.wnl.0000398198.48275.f4

This information is current as of May 9, 2011

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