system and cerebral cortex. **DLB and PD are discussed in Chaps. 434 and 435, respectively.**

■ CORTICOBASAL SYNDROME

CBS is a slowly progressive dementia-movement disorder associated with severe degeneration in the perirolandic cortex and basal ganglia (substantia nigra and striatopallidum). Patients typically present with asymmetric rigidity, dystonia, myoclonus, and apraxia that render a progressively incapacitated limb, at times associated with *alien limb* phenomena in which the limb exhibits unintended motor actions such as grasping, groping, drifting, or undoing. Eventually CBS becomes bilateral and leads to dysarthria, slow gait, action tremor, and a frontal-predominant dementia. Whereas CBS refers to the clinical syndrome, CBD refers to a specific histopathological FTLD-tau entity (Fig. 432-2). Although CBS was once thought to be pathognomonic for CBD, increasingly it has been recognized that CBS can be due to CBD, PSP, FTLD-TDP, and AD, the latter accounting for up to 30% of CBS in some series. In CBD, the microscopic features include ballooned, achromatic, tau-positive neurons; astrocytic plaques (Fig. 432-3); and other dystrophic glial tau pathomorphologies that overlap with those seen in PSP. Most specifically, CBD features a severe tauopathy burden in the subcortical white matter, consisting of axonal threads and oligodendroglial coiled bodies. As shown in Fig. 432-2, patients with bvFTD, nonfluent/agrammatic PPA, and PSP-RS may also show CBD at autopsy, emphasizing the importance of distinguishing clinical and pathologic constructs and terminology. Treatment of CBS remains symptomatic; no disease-modifying therapies are available.

■ FURTHER READING

IRWIN DJ et al: Frontotemporal lobar degeneration: Defining phenotypic diversity through personalized medicine. Acta Neuropathol 129:469, 2015.

MACKENZIE I et al: Nomenclature and nosology for neuropathologic subtypes of frontotemporal lobar degeneration: An update. Acta Neuropathol 119:1, 2010.