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.Frontotemporal lobar degeneration
with ubiquitin-positive
intracytoplasmic inclusions.
Frontotemporal lobar degeneration
(FTLD) with ubiquitin-positive
inclusions (FTLD-U) represents a
clinically heterogeneous group of
disorders. A neuropathological

subgroup of FTLD-U has previously been identified, with ubiquitin-immunoreactive intracytoplasmic inclusions in frontal or temporal lobes. To evaluate further the specific neuropathological features of FTLD-U, we examined 546 cases and found that all the cases examined had inclusions composed of ubiquitinated fibrillar material, but the appearance of the inclusions was not uniform. A subset of the cases had large numbers of inclusions with a characteristic granular appearance,

resembling a distribution of optically small discs with a circumscribed, typically perikaryal rim. Similar inclusions, visualized by the Gallyas technique for amyloid, were also present, but there was no co-localization of either tau or ubiquitin immunohistochemistry and the tau-positive inclusions did not have a granular appearance. These cases were not distinct from other FTLD-U subgroups (compact/cortical FTLD-U, FTLD with motor neuron disease type C, and progressive supranuclear

palsy), but they had no definitive clinical features to suggest that they might belong to any of these groups. The granular inclusions were present in both elderly and younger cases, and some were restricted to motor areas. Inclusions with a granular appearance were present in some cases with no evidence of FTLD-U, in parietal or occipital lob 6d1f23a050

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