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STRUCTURAL EFFECTS OF NEURODEGENERATION IN SPORADIC CREUTZFELDT-JAKOB DISEASE

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This study investigated structural brain changes associated with sporadic Creutzfeldt-Jakob Disease (sCJD) in 11 autopsy-confirmed cases assessed at Barnes-Jewish Hospital. The processes that promote the spread of pathology in patients with sCJD remain unclear with important implications for diagnosis and treatment. Accordingly, we identified regions most affected by sCJD by measuring cortical thickness for various brain regions, parsed using Freesurfer. We compared these values to 22 age- and sex-matched, cognitively normal controls. Significant decreases in cortical thickness were noted in the superior and inferior temporal gyri, fusiform gyrus, precentral and postcentral gyri, precuneus, caudal middle frontal gyrus, superior frontal gyrus, superior and inferior parietal lobules, lingual gyrus, supramarginal gyrus, transverse temporal gyrus, paracentral lobule, entorhinal cortex, insula and pars opercularis ($p < 0.01$) compared to controls. Our results confirm that cortical thickness decreases with sCJD. Certain brain regions appear to be disproportionately affected, identifying areas of apparent selective vulnerability to sCJD-mediated brain changes. We compared the cortical thickness of the 17 ROIs for slow and fast disease progression using a t-test. Cortical thickness tended to be highest in controls and smallest in long-duration sCJD subtypes (MV2, VV2, MM2), while thicknesses of shorter duration subtypes (MM1, MV1, VV2) fell in between those of controls and long duration subtypes. This was a general trend for the ROIs but not always the case. More research is needed before a definitive conclusion can be made regarding the relationship between neurodegeneration and disease duration. Additionally, we found no correlation between the thickness of significant regions and CSF Tau, confirming CSF Tau is not a specific biomarker for regional patterns of atrophy in patients with sCJD. We conclude that the temporal and posterior areas of the brain are selectively vulnerable to neurodegeneration causing cortical thickness decreases in patients with sCJD. This knowledge can improve CJD diagnosis and aid in treatment options in the future. In subsequent projects, volume measurements may be the best way to see early changes associated with CJD but more longitudinal studies are needed.