Posterior primary progressive prosopagnosia

Structural and molecular imaging

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Figure ^{18}F -FDG, ^{18}F -florbetapir, and ^{11}C -PBR28 PETs from a patient with amyloid-negative posterior primary progressive prosopagnosia

Greatest cortical thinning (top, yellow) corresponded to lowest metabolism and highest tau and inflammation uptake (arrows). standardized uptake value ratios (SUVR) were obtained by referencing to the cerebellar gray matter. ^{11}C -PBR28 parametric images of total distribution volume (V_T) were calculated using the Logan plot and a metabolite-corrected arterial input function.

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Amyloid-negative pathology underlying primary progressive prosopagnosia usually affects the tip of the right temporal lobe first.^{1,2} However, a 69-year-old right-handed man, with amyloid-negative PET imaging, had severe progressive prosopagnosia associated with changes in the right posterior temporo-occipital cortex, including the fusiform face area, but sparing the temporal lobe tip. Brain regions affected were documented by cortical thinning on MRI and by abnormal metabolism, tau, and inflammation on PET (figure). By age 71, he had developed signs of the clinical corticobasal syndrome (CBD), as manifested by apraxia, motor impersistence, rigidity, and a supranuclear eye movement disorder. CBD was heralded by right premotor involvement in his earlier cortical thickness study (figure).

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Disclosure

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