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Data-driven atrophy progression model of sporadic Frontotemporal dementia

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State of the art: Frontotemporal dementia (FTD) encompasses a wide variety of syndromes accompanied by heterogeneity in the observed atrophy patterns. While longitudinal studies help in better understanding the progression of genetic FTD, progression of sporadic FTD (sFTD) is sparsely understood. In this study, we estimated subtypes of atrophy progression timelines, jointly for various clinical phenotypes of sFTD using disease progression modelling.
Methodology: FTD patients in the Amsterdam Dementia Cohort were categorized as sporadic based on their Goldman score and genetic testing. Cross-sectional Freesurfer volumes of 176 sFTD patients comprising 116 behavioural FTD (bvFTD), 40 right-temporal variant FTD (rtvFTD), and 20 semantic-variant primary progressive aphasia (svPPA) were used in our analysis. Homogeneous subtypes and their atrophy progression timelines were jointly estimated for each clinical phenotype, using a novel extension of discriminative event-based model (Venkatraghavan et al. NeuroImage 2019; 186 518-532). For estimating these timelines, 680 subjective cognitive decliners were used as reference group.
Results: rtvFTD and svPPA were homogeneous with right and left temporal-lobe asymmetry, respectively. Three distinct timelines were established for bvFTD: early frontal-lobe atrophy subtype, early Insula and Cingulate atrophy subtype, and early right temporal-lobe atrophy subtype. The difference between rtvFTD and the right temporal-lobe subtype of bvFTD is the early bilateral involvement of Hippocampus and Amygdala in the latter.
Conclusion: The temporal variants, rtvFTD and svPPA, exhibited a homogeneous asymmetrical atrophy pattern. bvFTD was found to be heterogeneous with three distinct atrophy-based subtypes. Our novel approach might be useful for clinical phenotyping and monitoring progression of sFTD.

Conflicts of interest

Frederik Barkhof is a consultant to Combinostics.