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Motor neuron symptoms are linked to pathology burden in the medulla oblongata of FTLD-TDP brain donors

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State of art: motor neuron symptoms (MNS) are the hallmark of amyotrophic lateral sclerosis (ALS), which is clinically and pathologically related to frontotemporal dementia (FTD). Up to 20% of FTD patients are diagnosed with ALS. However, MNS are observed in a larger proportion of FTD patients (FTD-MNS). The burden of TDP-43 pathology in the hypoglossal nucleus (HN) is higher in ALS-FTD compared to pure FTD brain donors, but little is known about the distribution of TDP-43 pathology in the medulla oblongata of FTD-MNS.
Methodology: MNS occurring at any disease stage were assessed from the records of FTLD-TDP brain donors from the Netherlands Brain Bank (n = 38). The total amount of neurons and the percentage of neurons showing TDP-43 pathology were counted in the HN, nucleus ambiguus (NA) and inferior olivary nucleus (ION). ANOVA was performed between pure FTD, FTD-ALS, and FTD-MNS.
Results: a pure FTD syndrome was recorded in 12/38 FTLD-TDP brain donors. A concomitant FTD-ALS diagnosis had been established during life for 4/38 brain donors, while 16/38 had FTD-MNS. The most common MNS were dysphagia (63%), spasticity (29%) and wasting (24%). A higher TDP-43 burden in the HN and NA was observed in FTD-ALS compared to FTD ($p < 0.05$). TDP-43 burden in the HN and in the ION was significantly higher in FTD-MNS compared to FTD brain donors ($p < 0.05$).
Conclusion: MNS are common in FTD, even in the absence of ALS. MNS in FTD are linked to TDP-43 burden in the HN, NA and ION.

Conflicts of interest

N/A