BULBAR AMYOTROPHIC LATERAL SCLEROSIS: Beyond the motor system

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Bulbar-onset or development of bulbar signs over disease course has been associated with an increased burden of cognitive-linguistic impairments in ALS. Limitations of neuropsychological testing indicate a need for direct investigation of pathological changes in the brain using neuroimaging and neuropathology. The purpose of the work was to further our understanding of bulbar ALS and its association with extramotor deficits by examining neuroanatomical changes using a multimodal approach: Study 1 used MRI techniques to examine gray and white matter in 16 ALS patients with bulbar signs; Study 2 examined neuropathology in brain tissue in 3 bulbar-onset (bALS), 3 spinal-onset with antemortem bulbar impairments (sALSwB), and 3 spinal-onset without bulbar impairment (sALSnoB) cases. MRI findings revealed associations between bulbar motor dysfunction (articulatory rate) and gray matter changes in bilateral inferior frontal (IF) and transverse temporal (TT) gyri. Neuropathological findings revealed increased pathology in IF, posterior superior temporal gyrus, and TT for patients with bALS, followed by sALSwB, and sALSnoB. The work suggests a link between the bulbar motor involvement and neuroanatomical changes in the brain beyond the motor system.