著者| 麻田千原
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その他のタイトル| 進行した筋萎縮性側索硬化症の舌肥大
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Background: Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease that affects upper and lower motor neurons. Degenerative changes associated with ALS include the hypoglossal neurons, resulting in atrophy, weakness, and tongue fasciculations. Tongue enlargement (macroglossia) was reported in 2 patients with advanced ALS who were supported by tracheostomy-invasive ventilation (TIV), and tongue biopsy revealed marked degeneration of muscle fascicles accompanied by fatty replacement. We speculated that macroglossia in ALS was associated with an energy imbalance and abnormal fat accumulation due to disease progression and excessive energy intake during TIV use.

Object: In the present study, we examined the prevalence of macroglossia and analyzed clinical correlations in 65 ALS patients (36 men and 29 women) on TIV.

Methods: We investigated absence or presence of macroglossia, and its relationship with various clinical parameters including duration of TIV use, body mass index (BMI), energy intake, communication impairment, and oral functions with the Japanese version of the Norris Bulbar Scale. Logistic multivariate analysis was used to examine which factors affected the appearance of macroglossia.

Results: Macroglossia was identified in 22 patients (33.8%). A comparison of patients with and without macroglossia revealed that the former had a significantly younger age of onset (57.9 vs. 64.1 years (ys) in average) (p=0.012), longer total disease duration from onset to the time of the examination (139.6 vs. 93.4 months (ms)) (p=0.002), younger age at
the time of TIV initiation (60.4 vs. 67.0 ys) (p=0.002), and longer duration of TIV use (108.8 vs. 61.8 ms) (p< 0.0001), lower ALS functional rating scales revised (0.3 vs. 0.7) (p=0.036), higher BMI (20.5 vs. 18.1 kg/m²) (p=0.001), lower energy intake (920 vs. 990 kcal/day) (p=0.041) than the latter. In addition, the former showed more advanced communication impairment, and more severe oral dysfunction including disabilities of lip and perioral movements, cheek blowing-out movement, tongue and jaw movements, compared with the latter. In logistic multivariate analysis, BMI (p=0.007) and communication impairment stages (p=0.029) had significant effects on the appearance of macroglossia.

**Conclusions:** Macroglossia may be the result of overfeeding and replacement by fat during longterm TIV use in patients with advanced ALS.