Not only bright tongue sign in Pompe disease

Figure 1 Lingual tumor

Tumor at the right side of an atrophic tongue.

Figure 2 Cerebral MRI

Areas of signal fat infiltrating tongue intrinsic muscles and the floor of the mouth, predominantly on the right side, where they form a pseudonodular image. (A) Sagittal T1-weighted, (B) axial T1-weighted, and (C) axial T1-weighted with fat suppression.

A 66-year-old woman with the diagnosis of late-onset Pompe disease, receiving enzyme replacement therapy for the past 3 years, presented with a tongue tumor (figure 1) of uncertain evolution over time. It was soft at palpation and in the MRI study, the tongue looked bright in T1 but also an intense localized fatty degeneration generating a pseudohypertrophy of the tongue could be seen (figure 2). In late-onset Pompe disease, skeletal muscle is the most affected tissue and bright tongue sign is well known in patients with Pompe disease,^{1,2} but a benign tumor composed of muscle lingual pseudohypertrophy is an unusual finding.

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