Persistent nasal speech in a patient with myasthenia gravis

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A 49-year-old man with 24-year history of acetylcholine receptor antibody positive (AChR) myasthenia gravis (MG) presented with worsening dyspnea, orthopnea, diplopia, dysphagia, and muscle weakness. For 10 years after diagnosis, he received only pyridostigmine. Subsequently, after an exacerbation, he started prednisone 80 mg daily, pyridostigmine extended release 180 mg twice a day, and mycophenolate (stopped after 10 years because of infections). Prednisone was tapered and he was taking 40 mg every other day at the time of presentation (same dose for 7 years). He received 5 sessions of plasma-pheresis for MG exacerbation. After treatment his symptoms improved but nasal speech persisted. On examination, he could not elevate his eyebrows and tongue protrusion was weak and had central furrowing. Palate did not elevate, even with gag reflex testing. MRI showed T1 hyperintensity in intrinsic tongue and palate muscles (figure).

Tongue muscle MRI T1 hyperintensity has been described in muscle-specific tyrosine kinase MG and less commonly in AChR MG and is thought to represent fatty replacement.¹ As exemplified by this case, palatal muscles can also develop fatty signal changes. Furthermore, it is important to remember that persistent dysarthria and palatal weakness in patients with MG can be secondary to fatty atrophy and are not necessarily an indication for further escalation of therapy because they are likely to be permanent. Persistent ophthalmoparesis and

Figure MRI in patient with myasthenia gravis with persistent nasal speech and palatal weakness

(A) Sagittal T1 sequence of a normal tongue shows 2 thin hyperintense bands that run parallel to its surface (arrows). (B) Sagittal T1 sequence of patient with myasthenia gravis shows hyperintensity of intrinsic tongue and soft palate muscles (arrows) with loss of normal architecture and of the 2 thin hyperintense bands. (C) Coronal T1 sequence shows central furrowing indicative of tongue atrophy (arrow).

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extraocular muscle atrophy have also been described in MG.² Long duration of disease and high-dose steroids are related to the development of tongue muscle atrophy.¹ Suboptimal therapy with only pyridostigmine for many years after diagnosis also likely played a role in our case.

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