

Reversible tongue atrophy in Lambert-Eaton myasthenic syndrome

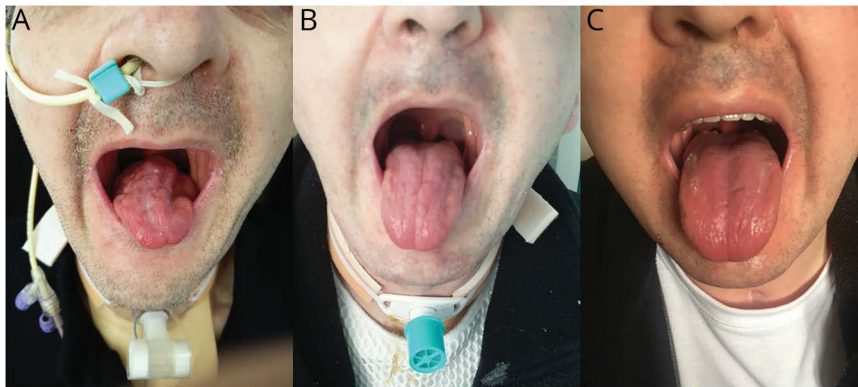
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Figure Patient photographs



(A) Two months after presentation with Lambert-Eaton myasthenic syndrome, inability to fully protrude tongue and marked bilateral tongue atrophy is evident. (B) Three months later, 2 months after radiotherapy to the patient's small cell lung tumor, only mild atrophy was apparent with a triple furrowed appearance, and (C) normal tongue 10 months after radiotherapy.

A 48-year-old man presented with severe limb, bulbar, and respiratory weakness requiring intubation and ventilation. Decrement was seen on repetitive nerve stimulation, but tested muscles were too weak for voluntary activation and tetanic stimulation was not tolerated. Positive voltage-gated calcium channel and SOX1 antibodies confirmed Lambert-Eaton myasthenic syndrome (LEMS). Despite corticosteroids, 3,4-diaminopyridine, and plasma exchange, severe bulbar weakness persisted, the patient remained tracheostomy-dependent, and he developed marked tongue atrophy (figure, A). Small cell lung cancer was subsequently confirmed. Radiotherapy 3 months after presentation resulted in recovery of bulbar function, improvement in tongue atrophy, and tracheostomy decannulation with improvement sustained 10 months later (figure, B and C). Tongue atrophy uncommonly occurs in generalized myasthenia gravis¹ but has not been previously reported in LEMS. The underlying mechanism may be neurogenic atrophy due to severe neuromuscular junction dysfunction.²

Author contributions

L.A. Cammaert: drafting/revising the manuscript, data acquisition, accepts responsibility for conduct of research and final approval, acquisition of data. A.D. Macleod: drafting/revising the manuscript, data acquisition, accepts responsibility for conduct of research and final approval,

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study supervision. G.A. Mackay: drafting/revising the manuscript, accepts responsibility for conduct of research and final approval, study supervision, review of relevant literature on potential mechanism of muscle atrophy in LEMS. C.W. Duncan: drafting/revising the manuscript, accepts responsibility for conduct of research and final approval, consultant in charge of patient.

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Disclosure

The authors report no disclosures relevant to the manuscript. Go to Neurology.org/N for full disclosures.

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