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Facial and bulbar muscle atrophy in acetylcholine receptor antibody-positive myasthenia gravis

Atrofia de musculatura facial e bulbar em miastenia gravis com presença do anticorpo Anti-AChR

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A 62-year-old man presented with seven years of progressive dysphagia, dysphonia and difficulty in closing both eyes. His examination showed weakness and atrophy of facial and bulbar muscles without ocular involvement (Figure A, B and C). Single-fiber electromyography revealed increased jitter (Figure D). To evaluate for concurrent myopathy, a muscle biopsy was performed and showed angulated atrophic type II fibers, a particular finding described in patients with myasthenia gravis[1,2] (Figure E). Acetylcholine receptor antibody was positive (2.2 nmol/L). Pronounced facial and tongue atrophy is uncommon in myasthenia gravis and usually associated with the muscle-specific receptor tyrosine kinase antibody, which was negative in this patient[3,4].

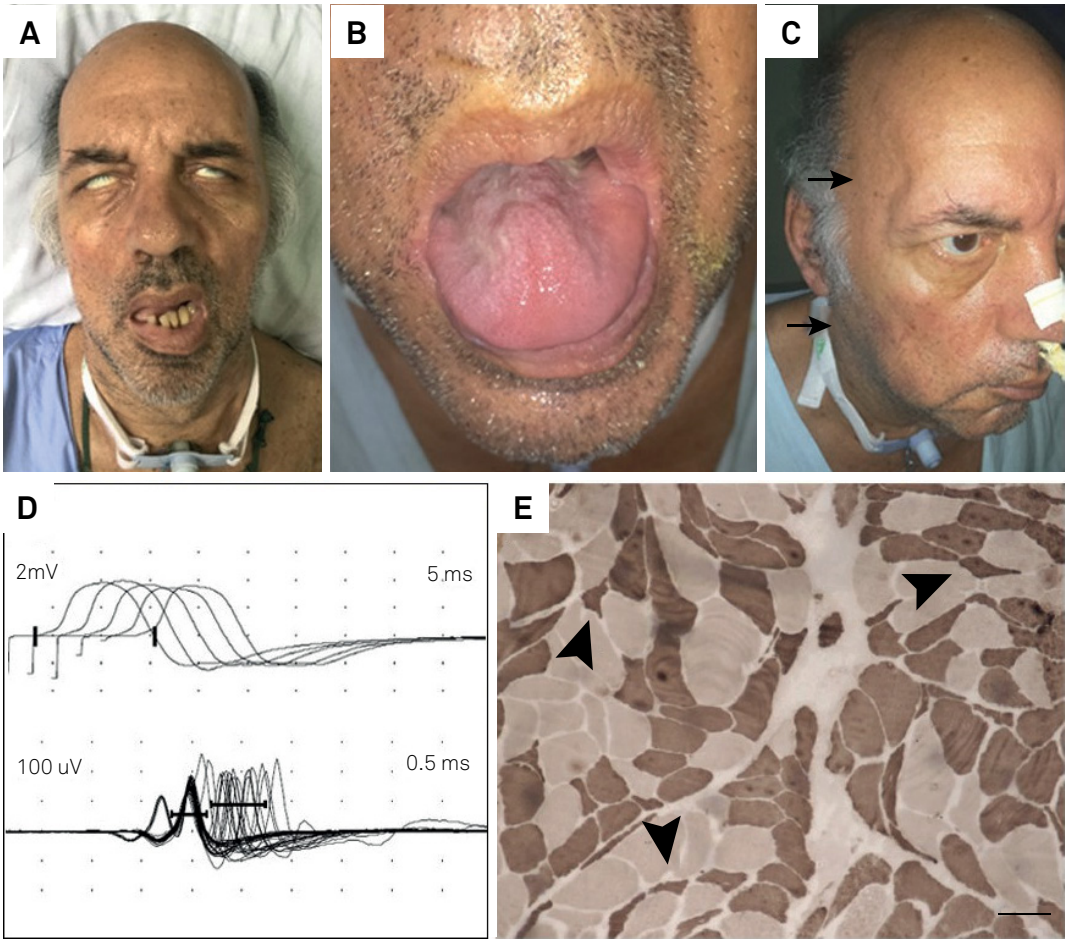

Figure. Clinical, electrophysiological and left biceps brachii muscle biopsy findings. Bilateral facial weakness, Bell's sign (A) and atrophy of the tongue (B), temporal and masseter muscles (arrows) (C). Repetitive stimulation was unremarkable (upper image) and the single-fiber electromyography demonstrated increased jitter (bottom image) (D). Muscle ATPase (pH 9.4) stain revealed angulated fiber II atrophy (arrowhead) (E). Bar = 100µm.

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