NeuroImages

Tongue involvement in amyloidoses
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Light chain amyloidosis and transthyretin (TTR) amyloidosis both have peripheral neuropathy as a prominent neurologic feature. We report two patients with amyloidotic peripheral neuropathy and remarkably different abnormal appearance of the tongue. One showed macroglossia (figure, A) secondary to deposition of amyloid between the muscle fibers, whereas the other showed tongue atrophy (figure, B) presumably caused by TTR amyloid deposit in the hypoglossal nerves.


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Figure. (A) A 61-year-old woman had an enlarged tongue. At age 58 she had restrictive cardiomyopathy and 2 years later proximal muscular weakness developed. In addition, EMG demonstrated a prominent motor axonal neuropathy. A monoclonal serum protein was identified leading to a diagnosis of amyloidosis, which was confirmed by endomyocardial biopsy. (B) A 53-year-old woman reported paresthesiae and dysesthesia in the feet and showed a severely atrophic tongue with continuous fasciculations. At age 37 she presented with loss of vision caused by amyloid infiltration of the vitreous body. Neurologic examination showed a mild sensory neuropathy and EMG confirmed axonal polyneuropathy. Molecular studies revealed the Pro36 TTR mutation.
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