IMAGES IN CLINICAL MEDICINE

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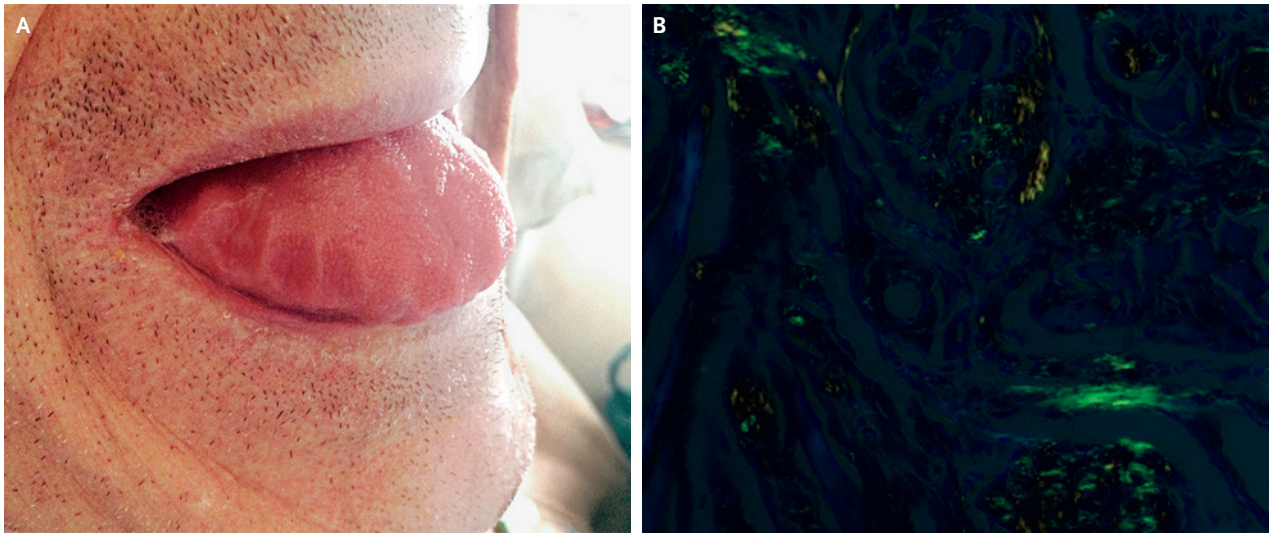
Macroglossia in Light-Chain Amyloidosis



A 78-year-old man presented to the emergency department with a 2-week history of generalized edema, weakness, and chest pain. He reported that during the past year his tongue had become larger and increasingly stiff and that it had become difficult for him to swallow. Physical examination revealed macroglossia, with indentations from the teeth (Panel A). Laboratory findings were notable for proteinuria (2+ protein on dipstick urinalysis and 505 mg of protein per 24-hour urine collection) and elevated levels of troponin I (1.99 μg per liter [reference range <0.06]) and N-terminal pro–brain natriuretic peptide (58,302 pg per milliliter [reference range, <450]). Transthoracic echocardiography and cardiac magnetic resonance imaging were performed, and the results were consistent with an infiltrative cardiomyopathy. Serum immunoelectrophoresis revealed elevated levels of free lambda light chains (2830 mg per liter; reference range, 8.3 to 27.0). A biopsy of the tongue was performed, and the findings confirmed a diagnosis of light-chain (AL) amyloidosis, which appears under polarized microscopy as an apple-green birefringence after Congo red staining (Panel B). Lambda chains were revealed on immunohistochemical analysis (not shown). In primary systemic, or AL, amyloidosis, monoclonal light chains are deposited in tissues. AL amyloidosis is a well-recognized cause of macroglossia. Early diagnosis and treatment are critical to prevent irreversible organ damage. This patient died shortly after the diagnosis was made as a result of complications from aspiration pneumonitis.

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