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Picture of the month

Muscle hypertrophy in amyloid myopathy

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A 67-year-old woman was referred because of easy fatigue and proximal lower limb muscle weakness started 3 years before. She also complained of myalgias in the last year. She had been diagnosed with a IgG lambda monoclonal gammopathy of uncertain significance (MGUS) at age 50. Family history was negative for neuromuscular disorders. Physical examination showed diffuse muscle hypertrophy and macroglossia (Fig. 1A). She could walk with slightly waddling and wide-based gait because of increased size of the internal compartment of the thigh. She had mild weakness of hip flexors and extensors (Medical Research Council, MRC 4). CK level was 3x normal, electromyography was myopathic with sporadic fibrillation potentials; nerve conduction studies were normal. Laboratory examinations confirmed the presence of multiple monoclonal IgG lambda components and free lambda light chains in serum and urine.

Muscle imaging showed diffuse muscle hypertrophy without major abnormalities on T1-weighted images and mild, mostly reticular signal increase on STIR sequences in muscles and intermuscular spaces (Fig. 1B). Muscle pathology confirmed the clinical suspicion of amyloid deposition (Fig. 1C and D). The patient was finally diagnosed with systemic amyloidosis associated with IgG lambda multiple myeloma. Amyloid myopathy is a rare entity, with very few muscle MRI scans reported so far [1]. Consistently with other published evidences, in our patient muscle imaging did not disclose significant signal abnormalities on T1-weighted images but a mild intramuscular and perimuscular hypersignal on fat suppressed T2-weighted images with increased reticularity in subcutaneous fat [1–3]. A peculiar finding was muscle hypertrophy, preferential for some groups such as the adductor magnus and periscapular muscles, likely corresponding to the increased perimysial and endomysial spaces due to amyloid deposition found on muscle biopsy. Conventional MRI sequences do not appear to be sensitive for the detection of amyloid deposition, which was massive in our case.

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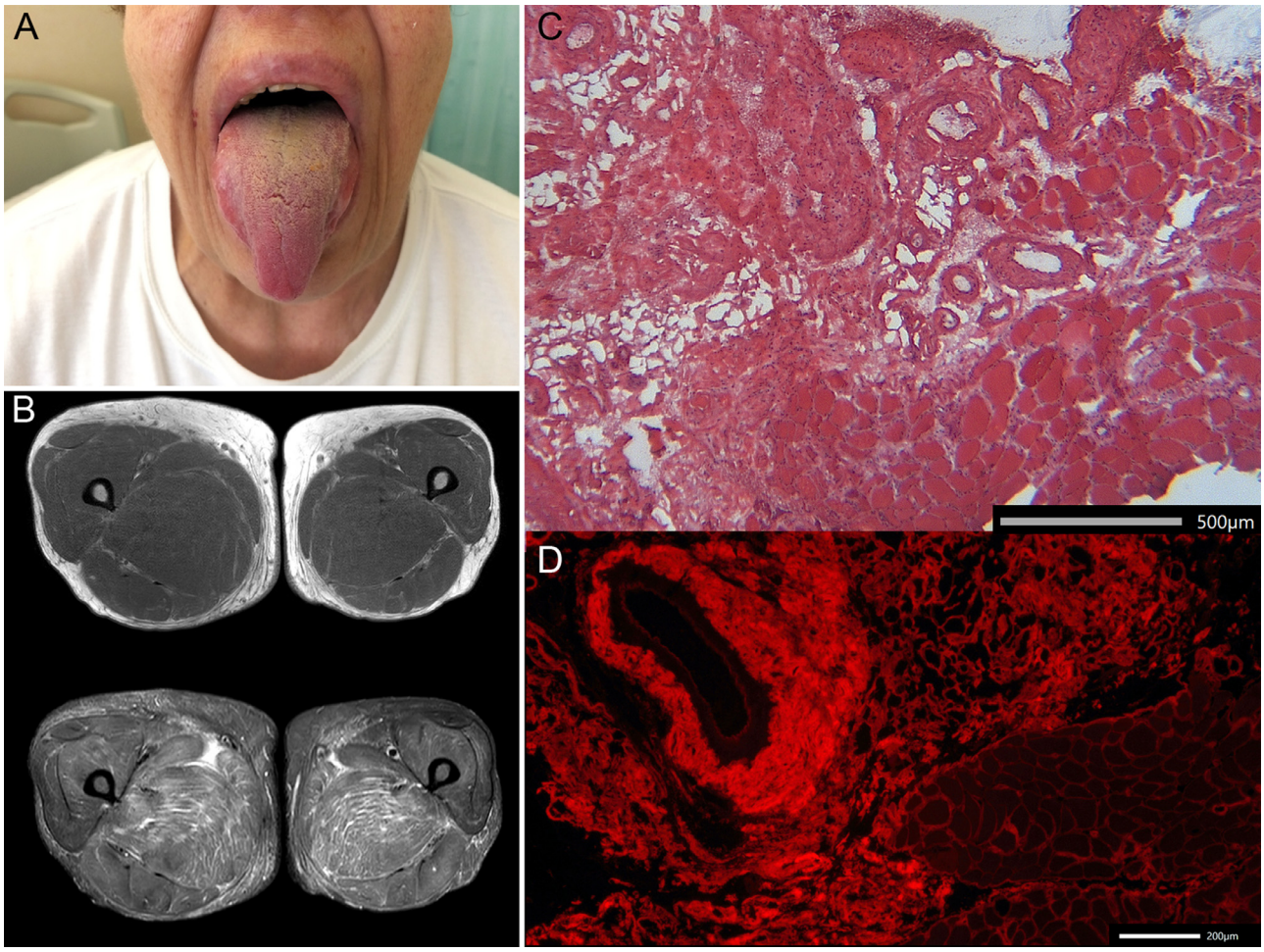

Fig. 1. (A) Hypertrophy of the tongue. (B) Muscle MRI showing striking bilateral hypertrophy of adductor magnus muscles, STIR sequences showing mild "striated" hypersignal in adductor magnus and subcutaneous fat. Muscle biopsy from the left vastus lateralis [hematoxylin and eosin (C), and Congo red staining visualized by fluorescence microscopy using Texas red filter (D)], showing massive amyloid deposition in perivascular, perimysial and endomysial spaces.

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