



A 48-year-old man with myasthenia gravis complaining of muscular stiffness

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A 48-year-old man developed bilateral arm flexion weakness, dysphagia, and nasal regurgitation of liquids which progressed over 1 year. Serum CK ranged from 230 to 440 mg/dl (normal, 200 mg/dl). Left deltoid muscle biopsy showed epimysial and endomysial inflammatory cell infiltrates. He was diagnosed with an inflammatory myopathy and treated with prednisone; within 2 weeks, he noted substantial improvement. He tapered his prednisone over 12 months, and relapsed with proximal arm weakness, diplopia, dysphagia, and head drop. Further evaluation showed a 25% decrement to low-frequency repetitive nerve stimulation of the spinal accessory nerve. The acetylcholine receptor antibody level was 65.2 (normal, 0.08), and chest CT showed anterior mediastinal mass. He was treated with 60 mg/day of prednisone for 6 weeks without benefit, underwent plasmapheresis with improvement, underwent thymectomy, with pathology revealing a thymoma. He gradually improved and by the age of 52 was without symptoms on low-dose prednisone for 6 months when he started to note rippling waves of muscle contractions across his chest, back, and limbs that were precipitated by percussion. Rapid extension of his arms became painful. Serum CK increased from 440 to 788 mg/dl. The acetylcholine receptor antibody level was 5.7. Needle EMG studies did not show fibrillation potentials or myotonic or neuromyotonic discharges. Electrical silence was present during episodes of muscle rippling. He continued to taper his prednisone and went on pyridostigmine alone for 2 years without symptoms of myasthenia gravis but continued to have persistent symptoms of muscle rippling and stiffness with rapid limb movement.