

Distal myasthenia gravis

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Abstract

Myasthenia gravis (MG) characteristically involves ocular, bulbar, and proximal limb muscles. MG with predominant distal limb involvement is uncommon. We present a case of MG with distal, asymmetrical, upper limb involvement, seven years after disease onset.

Key words : Myasthenia gravis ; electromyography.

A 24-year-old woman with a history of MG presented with progressive weakness in both hands. At age 17, bilateral ptosis, diplopia, dysphagia, and dysphonia, in presence of acetylcholine receptor binding antibodies and decrement on repetitive nerve stimulation (RS) led to the diagnosis of MG. Complete clinical remission was seen after thymectomy. The last six years, the patient was asymptomatic in absence of medical treatment. On admission, according to the Medical Research Council (MRC) scale, third to fifth digit extension was graded 2, extension of the second digit 3, flexion of the fifth digit was graded 4, first to fourth digit flexion 3, finger abduction 3, wrist extension 2 on the right and 3 on the left. Lower limb and neck muscles were normal. Deep tendon reflexes were symmetrically present and there were no sensory abnormalities. Nerve conduction studies of the median, ulnar, and radial nerves were normal as was needle electromyography. RS of both ulnar and median nerves revealed a slight amplitude decrement of less than 10%, whereas RS of the radial nerve showed an amplitude decrement of 23%. Intravenous immunoglobulin therapy 2 g/kg was given over 5 days and prednisone 50mg per os once daily was started. Clinical improvement occurred two weeks later although a right-sided finger extension weakness of 4 persisted.

MG is an autoimmune disorder characterized by a postsynaptic defect in neuromuscular transmission (1). In addition to ocular and bulbar involvement, weakness of the trunk and extremities is a common feature. However, distal extremity muscles are typically spared or less prominently involved than proximal ones. Only a few cases of MG with predominant distal limb involvement have been



FIG. 1. — Clinical examination shows, in attempt to wrist and finger extension, a third to fifth digit extension weakness graded 2, extension of the second digit 3, wrist extension 2 on the right and 3 on the left.

reported (2-6). A retrospective chart review of MG patients reported 3% of distal extremity weakness exceeding proximal weakness by at least one MRC grade during the course of their illness (2). Hand muscles and particularly finger extensors (as in our patient) were the most frequently affected distal limb muscles. Diagnostic sensitivity of RS of hand muscles seems to be higher in patients with clinical distal weakness than in patients with generalized MG. Another, prospective, series found in 7% of MG patients a predominance of muscle weakness in distal limb muscles (5). In both studies, distal weakness was seen as the presenting symptom of MG in one third of these patients. The majority of the remaining patients, as in our patient, developed distal weakness only years after disease onset.

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