Abstract

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A case of myasthenia gravis

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Background

An 86-year-old man came to our with reported weakness in the right upper limb and lower limbs, along with rhinolalia, dysarthria, and dysphagia for solids and liquids that had developed a few months prior. He had smoking history and a past medical history of chronic ischemic heart disease, atrial fibrillation with CRT-D implantation, two previous TIAs, arterial hypertension, and dyslipidaemia. Upon admission, he exhibited mild dyspnea with mild hypoxemia and hypercapnia on arterial blood gas analysis. Consultations were scheduled with a speech therapist, who identified coordination issues with swallowing without aspiration, and a neurologist, who observed a predominant right-sided pyramidal-extrapyramidal syndrome without Disautonomia. Brain CT revealed chronic cerebral vascular disease without focal abnormalities attributable to acute events. The involvement of the bulbar region, along with signs of both upper and lower motor neuron involvement, raised differential diagnostic concerns between amyotrophic lateral sclerosis (ALS) and myasthenia gravis. Definitive diagnosis relied on motor evoked potential studies, demonstrating a detrimental response in action potential amplitude following repetitive nerve stimulation, suggestive of a junctional pathology like myasthenia gravis. This justified the involvement of respiratory muscles and prompted serum testing for anti-acetylcholine receptor (AChR) antibodies, which were positive, as well as treatment with pyridostigmine and corticosteroids. Chest CT excluded thymoma, while thyroid ultrasound revealed a multinodular goiter underlying subclinical hypothyroidism with positive anti-thyroid peroxidase (TPO) antibodies.

Discussion

The term "myasthenia gravis" derives from Greek and means "severe weakness." It is an autoimmune disorder affecting the neuromuscular junction, characterized by autoantibodies directed against post-synaptic acetylcholine receptors. A small percentage of cases present with antibodies against muscle-specific kinase. Muscle fatigue ability is a characteristic feature, involving progressive reduction in strength during physical exercise, with diurnal fluctuations, worsened by exertion, improved after sleep or rest. It affects the muscles of the limbs and facial region, along with or pharyngeal bulbar involvement (mastication, dysarthria, dysphagia, rhinolalia), ocular symptoms (ptosis and diplopia), and occasionally respiratory muscle involvement, without autonomic involvement or normal osteotendinous reflexes. Only 15% of patients with myasthenia gravis have thymoma, while the remainder have thymic hyperplasia. The thymus appears to play a pathogenic role in the autoimmune response [1]. Diagnosis is based on neurophysiological tests such as motor evoked potentials (decremental response of action potential to low-frequency repetitive nerve stimulation) and single-fiber electromyography (increased jitter), antibody assays and, although seldom used, "Tensilon test," consisting of administration of anticholinesterase drugs followed by transient improvement of clinical symptoms. Currently, there is no definitive cure for myasthenia gravis, but there are treatment options available for symptom management. This includes anticholinesterase drugs like pyridostigmine; if no improvement is observed, corticosteroids and immunosuppressive drugs (azathioprine as the first choice) are initiated to reduce the autoimmune response. Plasmapheresis or immunoglobulin's are used in case of myasthenia crisis, and physical therapy is employed to maintain muscle strength. Thymectomy is indicated in the presence of thymoma and generalized myasthenia [2].

Despite myasthenia gravis usually occurs in young women and adult men, it can develop at any age and should be suspected in the presence of muscle weakness (Figure 1)

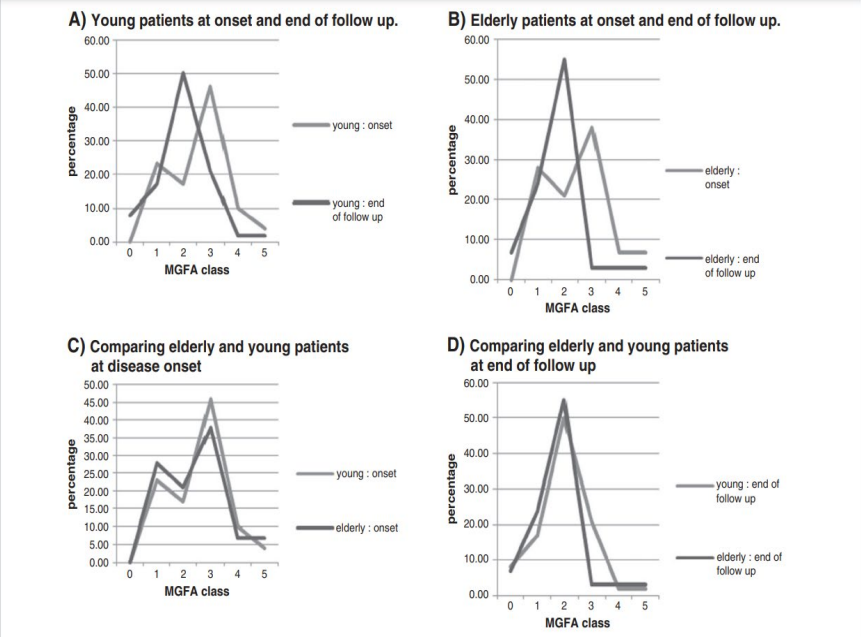

Figure 1 MGFA (Myasthenia Gravis Foundation of America) class at onset and end of follow up. A. Young patients at onset and end of follow up. B. Elderly patients at onset and end of follow up. C Comparing elderly and young patients at disease onset. D. Comparing elderly and young patients at end of follow up.

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