ATYPICAL CLINICAL PRESENTATION OF MYASTHENIA GRAVIS (MG)

E.P. Verrengia, S. Jann, V. Oppo, E.C. Agostoni

ASST Grande Ospedale Metropolitano Niguarda – Dipartimento di Neuroscienze

Objective: MG in patients 70 years or older is not infrequent and predominantly in males with a lower number of cases with high-rate of AchR antibodies. Clinical evaluation in elderly MG cases often shows fatigue, diffuse limb weakness or ocular symptoms, but MG can be a very heterogeneous disease.

Results: A 77 year-old man presented acute upper limb and neck weakness from January 2016. He was unable to keep his head up and showed no variability along the day. Despite the negativity of neurophysiology exam (single fiber electromyography- EMG), patient was treated with Pyridostigmine 60 mg four times/day with only slight and temporary improvement. Two months later, he developed progressive worsening of weakness and huge weight loss (8 kg). On April 2016 he was unable to raise his neck and to perform any activity using upper limbs, preserving the ability to walk. Neurological examination revealed a severe hypotrophy and weakness of bilateral cervical-brachial muscles with no neurologic bulbar involvement and normal deep tendon reflexes. The patient reported fatigue, in absence of symptoms fluctuations. In acute phase the patient underwent steroid treatment (Prednisone 50 mg daily) with no further benefit. During the hospitalization, he presented a urinary tract infection with worsening of general state and neurological bulbar involvement (dyspnea, dysphagia). Five days after low-dose steroid treatment, despite a negativity of antibodies against muscle-specific tyrosine kinase (MuSK), high-rate of Acetylcholine Receptor (AchR) were detected (111,9 nMol/L). Intravenous immunoglobulin (IVIG) was chosen as rescue therapy with excellent clinical results, followed by prednisone 50 mg/day as maintenance dose. To improve motor function, a rehabilitation period was prescribed. Two months later, the patient presented a normal muscular strength and tropism with no exhaustion signs.

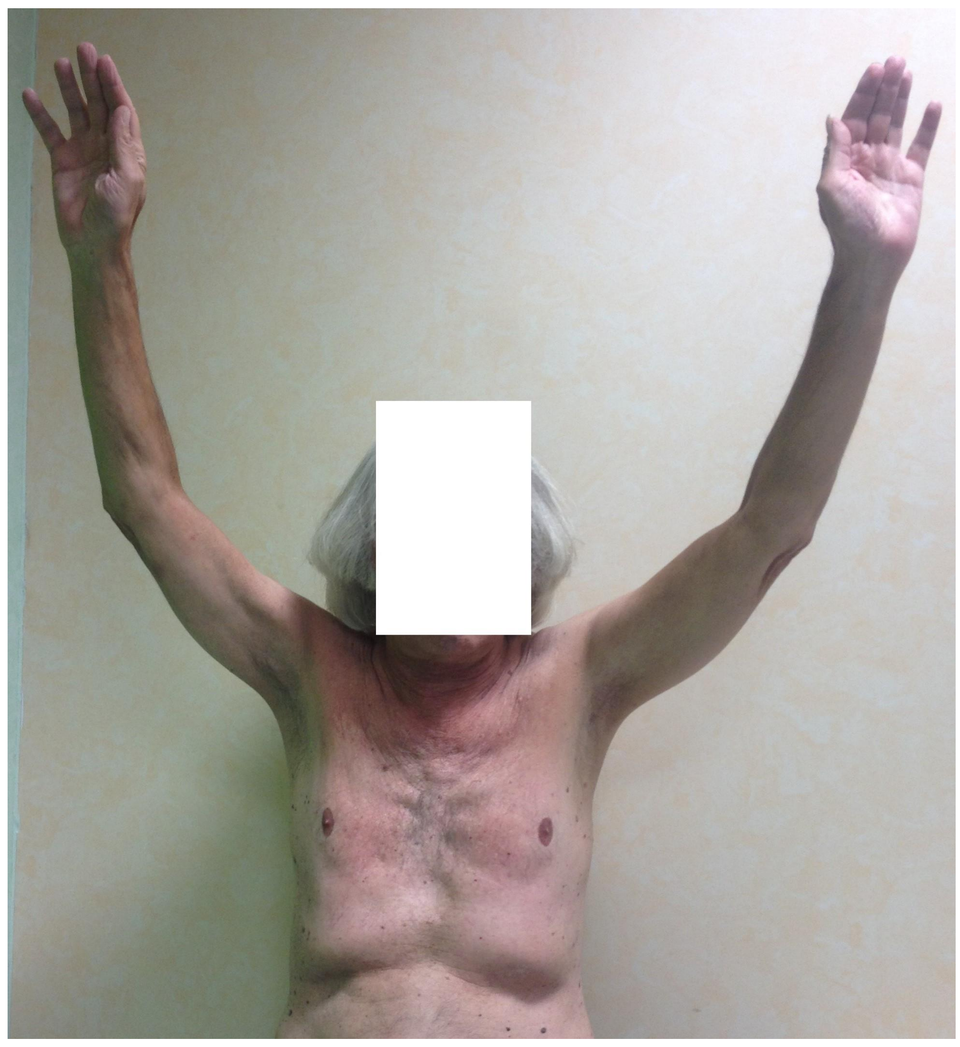

Fig.1 *Clinical follow-up 2 months after admission*

Discussion: The presence of proximal neck and upper limb weakness was at first related with other neurological conditions: cervico-brachial inflammatory, dysimmune/paraneoplastic myopathy or neurodegenerative disease. The diagnosis was difficult to establish for two main reasons: atypical clinical presentation at onset, negative neurophysiology and poor response to Pyridostigmine treatment.

Conclusion: MG is a very heterogeneous disorder with different age of onset and distinct clinical entity: from typical spinal/ocular or bulbar involvement to proximal limb weakness with myopathy signs. The dosage of antibodies involved in the neuro-muscular transmission is still important to support MG diagnosis and treatment.

Bibliography. 1.S. Binks, A. Vincent, J. Palace**. Myasthenia gravis: a clinical-immunological update.** J Neurol (2016) 263:826–834. **2**. A.V. Nikolic´, G.G. Bacˇic´, M.Zˇ, Dakovic´, S. Đ., Lavrnic´,V. M. Rakocˇevic´ Stojanovic, I.Z. Basta, D.V. Lavrnic´ Myopathy, muscle atrophy and tongue lipid composition in MuSK myasthenia gravis. Acta Neurol Belg (2015) 115:361–365. **3**.M.A. Hellmann, R. Mosberg-Galili, I. Steiner. Myasthenia gravis in the elderly. Journal of the Neurological Sciences 325 (2013) 1–5