A case of flail arm syndrome associated with monoclonal gammopathy

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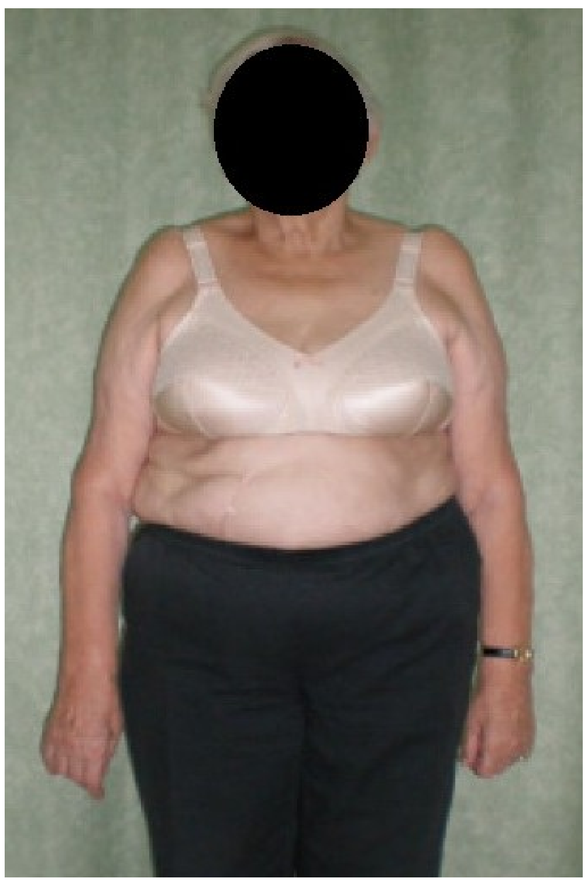
Introduction: Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disorder affecting motor neurons. Flail arm syndrome (FAS) is a rare form of ALS characterized by progressive, predominantly proximal, symmetric weakness and wasting of the upper limbs, with no significant lower limb or bulbar muscle involvement[1]. The literature suggests that patients with motor neuron disease (MND) may have a higher incidence of lymphoproliferative disorders (LPD)[2]. Monoclonal gammopathy of undetermined significance (MGUS) is the most common of a spectrum of diseases called plasma cell dyscrasias.

Case report: We describe a patient (80 years old), who at the age of sixty-five, had been diagnosed with ALS. She had complained of slowly progressive weakness that began in the left arm and has spread to the contralateral arm in a few months, associated with muscle spasms. The physical examination showed normal results in the lower limbs and bulbar region. She didn't complained of sensory symptoms and denied head or neck trauma or family history of similar diseases. Neurological examination showed complete paralysis of all muscles in both arms and absence of tendon reflexes in the arms; neurological examination was otherwise unremarkable. Electromyography (EMG) showed fibrillation potentials in the distal muscles in both arms and normal results in lower limbs. MRI of the cervical spinal cord showed only mild arthritic degeneration. Somatosensory evoked potentials (SSEPs) were normal, while the motor evoked potentials (MEP) showed slowed efferent conduction time in both upper and lower limbs. Blood tests were remarkable for the monoclonal IgG k component (2.03 g / l); the same component has been found in the cerebrospinal fluid. The follow-up with EMG and clinical examination, which showed worsening of abnormal activity in the upper limbs, without evidence of involvement of the lower limbs or the bulbar region, has led us to question the diagnosis of ALS, instead we have concluded for a diagnosis of Flail arm syndrome. The patient undergoes regular checks and, so far 20 years after symptoms onset, she presents upper limbs paralysis and only mild weakness in lower limbs.

Discussion and conclusions: The literature suggests an association between MND and LPD. The concomitant presence of MND and LPD could be coincidental, but LPD seems to be more frequent in patients with MND compared to the population in general. For this reason patients with neuromuscular diseases should be routinely screened with serum immunoelectrophoresis.
The presence of gammopathy appears to relate with the absence of upper motor neuron involvement and slowly progressive muscular atrophy.
In a patient that presents progressive weakness in the upper limbs, differentiating ALS with limb onset to FAS variant is of great importance for prognostic purpose with the latter bearing a significant better prognosis. Mean survival time from diagnosis for FAS is reported to be almost 6 years.

In our case the absence of upper motor neuron involvement and the spearing of other body areas after many years from symptoms onset allowed us to diagnose FAS.
To our knowledge it is the first reported case of FAS associated with MGUS with this unusual good prognosis.



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