LOWER MOTOR NEURONE SYNDROME, ANTI-MAG NEUROPATHY AND WALDENSTRÖM MACROGLOBULINEMIA: OCCAM`S RAZOR VERSUS HICKAM`S DICTUM

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BACKGROUND: Diseases selectively involving lower motor neurone (LMN) dysfunction have a limited differential diagnosis, including spinal muscular atrophies and motor neuropathies. Around 8-10% of patients who are initially diagnosed with motor neuron disease (MND) will eventually be found to have a different underlying condition. Others will turn out to have more than one.

CASE REPORT: A 73-year-old caucasian male presented with progressive right upper limb (RUL) paresis with muscle atrophy and fasciculations. He was diagnosed with monomelic MND and started on riluzole. In the following years he developed RUL numbness, gait unsteadiness, left foot drop, and severe weight loss. He presented to us in 2015 with ongoing worsening of the symptoms. His neurological examination revealed tongue fasciculations, dysphagia and dysphonia, shoulder girdle and RUL atrophy with fasciculations, areflexic quadriparesis (RUL and left foot predominance), hypoesthesia and hypopallesthesia of RUL and distal lower limbs, overall suggestive of LMN syndrome and peripheral neuropathy. EMG was compatible with MND and sensorimotor polyneuropathy. Blood tests revealed an IgM kappa gammopathy with positive anti-MAG antibodies, and the myelogram confirmed Waldenstrom`s Macroglobulinemia (WM). The patient currently awaits chemotherapy initiation.

DISCUSSION: We present an interesting and controversial clinical case comprising three rare disorders concurring in the same patient, in a paradigmatic example of a diagnostic conundrum. Following the principle of Hickam`s dictum, could it be just an unfortunate coincidence? Alternatively, based on Occam`s razor, and according to published data, both LMN syndrome and anti-MAG neuropathy may represent different manifestation of WM and thus constitute a single disease.