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Clinical Image

CLINICAL IMAGE

Dropped head and man-in-barrel syndrome in amyotrophic lateral sclerosis

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Abstract

We report a case of progressive symmetric brachial weakness followed by cervical muscle weakness. The electromyogram confirmed the diagnosis of amyotrophic lateral sclerosis. After 3 years the patient remained able to walk unassisted and without significant bulbar manifestations or upper neuron signs. The concomitant presence of dropped head syndrome and man-in-barrel syndrome in an amyotrophic lateral sclerosis patient makes our case unique.

INTRODUCTION

Amyotrophic lateral sclerosis (ALS) is a relatively rare disease with a significant phenotypic variation often leading to misdiagnosis [1]. Bilateral flail arm syndrome also known as 'man-in-barrel syndrome' (MIBS) is one of the rarest and atypical presentation of regional ALS. Progressive weakness and atrophy of upper limbs and absent of bulbar signs is characteristic of the motor neuron MIBS [2].

CASE REPORT

A 78-year-old male presented with 11-month history of progressive bilateral symmetric brachial weakness. Neurological examination disclosed the presence of symmetric and predominant distal slight atrophy with decreased motor strength (4/5 grade on Medical Research Council) in the upper limbs. The reflexes were diminished in the left upper limbs. The rest of the general, neurological examination including cognition was unremarkable. Needle electromyography was not supportive of ALS. The complementary work up including head and cervical magnetic imaging, creatine kinase level, GM1 Ab titers yielded negative results. He slowly progressed to brachial hypotonic hyporeflexive atrophic diplegia with associated marked atrophy of the shoulder girdles and decrease of neck extension strength culminating with drop neck after ~9 months of initial consultation (Supplementary Video). At this time the electromyogram was completely conclusive of ALS with ongoing denervation on the limbs and bulbar muscles. He started riluzol treatment and after 3 years of disease he is still able to walk unassisted and without significant bulbar manifestations or upper neuron signs.

DISCUSSION

The appearance of being 'locked in a barrel' because of bilateral absence of upper limbs movements gave name to the syndrome [3]. Watershed strokes, acute central medullar or cervical spinal cord disorders and ALS among the possible etiologies for MIBS [2]. Although being a well-known manifestation of ALS, drop neck rarely occurs in the absence of generalized disease [4]. Long-term survival in ALS patients with MIBS

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is somehow expected [5] but the concomitant presence of dropped head syndrome and MIBS in an ALS patient makes our case unique. This case highlights the clinical variability and the unpredictability of ALS.

SUPPLEMENTARY MATERIAL

Supplementary material is available at *Oxford Medical Case Reports* online.

CONFLICT OF INTEREST STATEMENT

None declared.

ETHICAL APPROVAL

None required.

INFORMED CONSENT

The patient's representative signed the written consent for publication including the video.

GUARANTOR

Hipólito Nzwalo, MD, MSc.

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