BRACHIAL AMIOTROPHIC DIPLEGIA: A RARE CAUSE OF "MAN IN THE BARREL SYNDROME"

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Objectives

To describe a patient with slowly progressive brachial diplegia giving him the clinical appearance of seeming constrained in a barrel ("man in the barrel syndrome", MIBS).

Patient and methods

We report on a 69-year-old patient having a 35-year history of slowly progressive weakness of the proximal segments of both upper limbs and shoulders which started on the left side and involved the right side 15 years later. Electromyography, cervical MRI and MRI of the cervical roots and brachial plexus were performed.

Results

Clinical evaluation showed severe weakness and atrophy of muscles of upper limb girdle while distal segments of upper limbs as well as lower limbs were preserved. Deep tendon reflexes were absent at the upper limbs and normal at the lower ones.

Electromyography showed chronic neurogenic abnormalities in the upper limb proximal muscles.

Cervical MRI and MRI of the roots, primary trunks, branches of division and cords of both brachial plexus were normal.

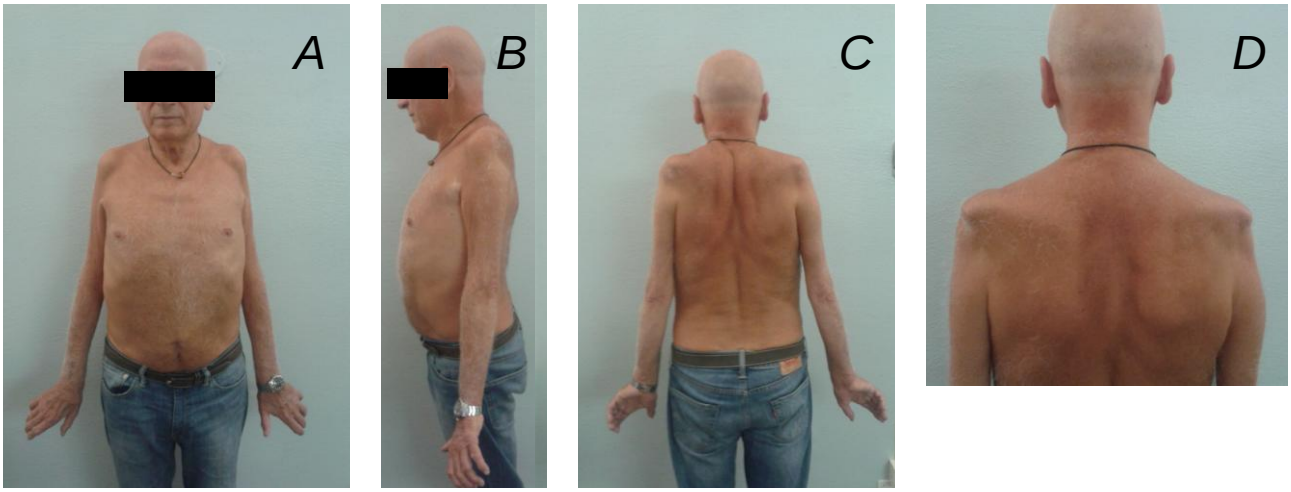

Fig. 1: *Our patient presents with severe muscle atrophy of upper limbs especially at limb girdle (A, B, C). Abduction of the upper limbs is not possible (A, B). In D, a detail of the muscle atrophy in upper limb girdle.*

Discussion

The term "Brachial Amiotrophic Diplegia" identifies one of the conditions causing the clinically heterogeneous syndrome named MIBS and is a primary sporadic motor neuron disorder which remains restricted to the upper limbs over time. Only a few cases have been described with a follow-up ranging between 2 and 11 years. It is characterized by brachial diplegia and preservation of motor function in the leg and facial muscles.

Conclusions

This patient has a 35-year history of disease with no evidence of clinical spreading to other muscle districts. This is the longest follow-up described of primary brachial amiotrophic diplegia, thus strengthening the concept that this condition entails a favorable prognosis and has to be distinguished from ALS variants such as upper limb onset ALS and flail arm syndrome.