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## Case report

# Unilateral progressive muscular atrophy with fast symptoms progression



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## ABSTRACT

Progressive muscular atrophy (PMA), or the lower motor neuron disease, is a sporadic disorder characterized by onset in adulthood, pure lower motor neuron involvement and relatively benign course. Muscle atrophy and weakness may be symmetrical or asymmetrical, but they are always bilateral. We present a male patient with exclusively left-side flaccid paresis due to lower motor neuron disease without electromyographic evidence of neurogenic lesion of contralateral muscles and with no signs of corticospinal tracts involvement. The rapid disease progression was typical of the generalized phenotype of PMA and it suggested the relation to the aggressive course of classical ALS.

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## 1. Introduction

Progressive muscular atrophy (PMA), or lower motor neuron disease, may be defined as a sporadic disease with onset in adulthood, characterized by progressive muscles weakness and atrophy due to degeneration of anterior horns cells and lack of upper motor neuron involvement signs [1]. Clinical signs of lower motor neuron lesion in PMA may be symmetrical or asymmetrical, but they are always bilateral. We report a patient with exclusively unilateral lower motor neuron involvement.

## 2. Case report

A 60-year-old male patient was admitted with a 2-year history of progressive weakness and muscle atrophy, which developed in both left limbs at the same time. His familial and past medical history was not indicative of any neurological disorders.

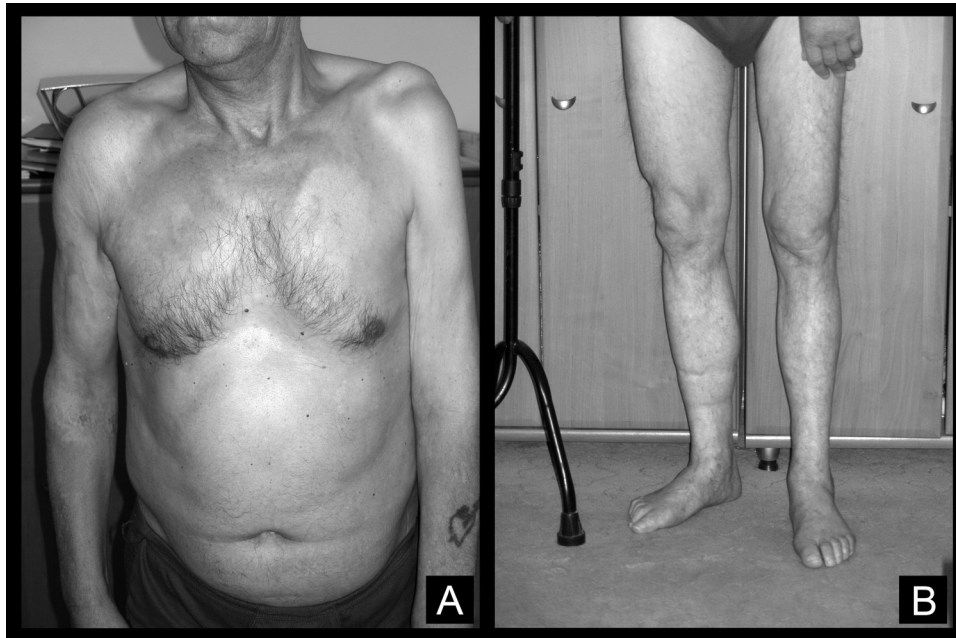
Physical examination showed cranial nerves intact. There was severe atrophy of shoulder girdle and arm muscles and moderate atrophy of the forearm and hand muscles on the left side (Fig. 1, Video 1). No fasciculations were observed in

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**Fig. 1 – The atrophy involved both proximal and distal muscles of the left upper (A) and lower (B) limb.**

affected muscles. Strength was 1/5 in all muscles of the left upper extremity, and deep tendon reflexes were diminished. Sensation was normal in all modalities. The right upper limb appeared normal on examination. The Hoffmann's sign and palmomental reflex was absent bilaterally. Abdominal reflexes were symmetrically diminished. Muscles of the left lower extremity were moderately atrophied, fasciculations were not observed. There was weakness more severe distally (0/5) than proximally (3/5). No sensory deficit was noted. Tendon reflexes were absent at the left knee and ankle. The right lower limb was normal. Plantar responses were flexor. The patient walked with a left foot drop. There was no sphincter dysfunction or dysautonomia.

Needle EMG examination showed severe pathological activity at rest (fibrillations, positive sharp waves) in tibialis anterior, rectus femoris, abductor pollicis brevis, deltoid and interosseus I muscles on the left side; MUAPs (enlarged) could be recorded only from the last of the above mentioned.

The EMG of the tongue and of the right rectus femoris, interosseus I and deltoid muscles were normal, mild rest activity and normal MUAPs were recorded from the right tibialis anterior muscle.

MNC studies of the right median nerve, both ulnar nerves and the right tibial nerve were normal. Low amplitude on stimulation of the left median and left tibial nerves were found, conduction blocks were not detected. SNC studies were normal.

No abnormalities were seen on MRI scans of the cervical and lumbo-sacral spinal cord, there was mild bilateral L5 and S1 roots compression due to intervertebral disc herniation. Blood and CSF studies were unremarkable.

We have no opportunity of re-examining this patient. He died 10 months later due to respiratory insufficiency; the autopsy was not performed.

### 3. Discussion

We present a patient with unilateral flaccid paresis without any sensory abnormalities as well as clinical upper motor neuron and bulbar signs. Electrophysiological studies revealed multilevel involvement of anterior horn cells limited to the left side of the spinal cord. We suggest that the presented case can be diagnosed as PMA.

Patients with PMA may be classified according to the pattern of muscle involvement at disease onset [2]. In subjects with segmental phenotype the prognosis is relatively good and muscle weakness and atrophy may remain confined to the originally affected region of the body even after many years. In such cases dysphagia and dysarthria are rarely observed and respiratory insufficiency is not common. In patients with generalized weakness, the disease progresses more quickly and it can lead to death due to respiratory insufficiency [3]. Also predominant axial involvement was connected with earlier respiratory failure [4]. The rapid disease progression in our case is typical of the generalized phenotype of PMA and it suggests the relation to the aggressive course of classical, generalized ALS.

In different subtypes of PMA, clinical signs of lower motor neuron lesion are symmetrical or asymmetrical, but they are always present bilaterally. Sporadically reported unilateral cases of primary lateral sclerosis – the syndrome being a result of selective degeneration of corticospinal tracts – are known as Mills syndrome [5–8]. To our knowledge, no subjects with

strictly unilateral and exclusive lesion of the anterior horns of spinal cord were reported. In 2005 Rajabally et al. [9] described a patient with a very slowly progressive unilateral paresis due to lower motor neuron involvement. However, in their case Hoffmann sign was present bilaterally and reflexes were pathologically brisk, more on the affected side. Also the muscle tone was moderately increased on the affected side. All these data strongly suggested that the upper motor neurons were affected by the disease process.

The pathogenesis of sporadic cases of ALS as well as of PMA remains largely unknown. One of the crucial problems is the understanding of the mechanism of disease propagation. In ALS, the spread within the lower motor system is usually prior to contralateral limb with subsequent involvement of the ipsilateral extremity [10]. So far, however, the spread of neurodegeneration through motor neurons remains the question of hypotheses rather than facts [10-12]. At present, these hypotheses are not of help in understanding why in the presented case the degeneration of lower neurons did not cross the midline.

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### Conflict of interest

None declared.

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### Ethics

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; Uniform Requirements for manuscripts submitted to Biomedical journals.

### REFERENCES

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- [1] Visser J, de Jong JM, de Visser M. The history of progressive muscular atrophy: syndrome or disease. *Neurology* 2008;70:723-7.
- [2] van den Berg-Vos RM, Visser J, Franssen H, de Visser M, de Jong JM, Kalmijn S, et al. Sporadic lower motor neuron disease with adult onset: classification of subtypes. *Brain* 2003;126:1036-47.
- [3] Van den Berg-Vos RM, Visser J, Kalmijn S, Fischer K, de Visser M, de Jong V, et al. A long-term prospective study of the natural course of sporadic adult-onset lower motor neuron syndromes. *Arch Neurol* 2009;66:751-7.
- [4] de Carvalho M, Scotto M, Swash M. Clinical patterns in progressive muscular atrophy (PMA): a prospective study. *Amyotroph Lateral Scler* 2007;8:296-9.
- [5] Mills CK. A case of unilateral progressive ascending paralysis probably presenting a new form of degenerative disease. *J Nerv Ment Dis* 1900;27:195-200.
- [6] Gastaut JL, Bartolomei F. Mills' syndrome: ascending (or descending) progressive hemiplegia: a hemiplegic form of primary lateral sclerosis? *J Neurol Neurosurg Psychiatry* 1994;57:1280-1.
- [7] Turner MR, Gerhard A, Al-Chalabi A, Shaw CE, Hughes RA, Banati RB, et al. Mills' and other isolated upper motor neurone syndromes: in vivo study with 11C-(R)-PK11195 PET. *J Neurol Neurosurg Psychiatry* 2005;76:871-4.
- [8] Rigamonti A, Lauria G, Prone V, Agostoni E. Mills' syndrome: an Italian case and revision of the literature. *Neurol Sci* 2013;34:255-6.
- [9] Rajabally YA, Hbabih M, Abbott RJ. Hemiplegic ALS. Mills syndrome. *Neurology* 2005;64:1984-5.
- [10] Ravits JM, La Spada AR. ALS motor phenotype heterogeneity, focality, and spread. Deconstructing motor neuron degeneration. *Neurology* 2009;73:805-11.
- [11] Talbot K. Amyotrophic lateral sclerosis: cell vulnerability or system vulnerability? *J Anat* 2014;224:45-51.
- [12] Sekiguchi T, Kanouchi T, Shibuya K, Noto Y, Yagi Y, Inaba A, et al. Spreading of amyotrophic lateral sclerosis lesions—multifocal hits and local propagation? *J Neurol Neurosurg Psychiatry* 2014;85:85-91.