International Journal of Advanced Multidisciplinary Research
ISSN: 2393-8870
www.ijarm.com
(A Peer Reviewed, Referred, Indexed and Open Access Journal)
DOI: 10.22192/ijamr Volume 8, Issue 1 -2021

Case Report

DOI: http://dx.doi.org/10.22192/ijamr.2021.08.01.004

Amyotrophic lateral sclerosis: Case report

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Keywords
Amyotrophic lateral sclerosis,
Lou Gerigh's disease,
upper motor neuron lesion

Abstract

Amyotrophic lateral sclerosis is a rare disease, prevalence of 6 / 100,000 people, with insidious onset and early diagnosis is often difficult. We report the case of a 51-year-old patient with starting symptons with weakness in the lower limbs and subsequent loss of ambulation, upper and lower motor neuron syndrome was integrated with subsequent diagnosis of Amniotrophic Lateral Sclerosis.

Introduction

ALS is a disease that affects adults of any race and ethnicity, and the risk of developing it increases with age, with a peak incidence between 40-60 years. In 90 to 95% of cases it occurs randomly, without any associated risk factor. It is unknown why the motor neurons degenerate in patients with ALS, although genetic, environmental, and age-related factors have been considered. Patients usually do not have a family history of the disease (only 5-10% of all cases are inherited). The familial form of ALS generally results from a hereditary pattern, which requires one of the parents to carry the responsible gene[1].

The prognosis is worse in the forms that present rapidly with breathing and / or swallowing problems, being worse in the familiar forms and in those that begin in advanced ages. Median survival from diagnosis is approximately 12 to 19 months[2].

Case Report

We present the case of a 51-year-old female patient with no known chronic degenerative diseases. It began 8 months ago, with weakness in the left lower limb, it conditioned frequent falls, contralateral lower limb weakness was added, limiting walking, 3 months later, she presented bilateral upper limb weakness, with fasciculations in the left shoulder and forearm, and at level lingual, shows changes in voice tone and slow language, 1 month later shows greater difficulty in performing activities such as eating, dressing and walking. 1 month later, he was unable to walk, due to worsening weakness in the lower limbs, and had lost 5 kg of weight in a period of 6 months.

It is evaluated in the External consultation of our unit for Internal Medicine and Neurology: with the following findings; Awake, comprehensive mental functions, without cognitive alterations, with bradylalia, hypophonia, lingual atrophy and lingual fasciculations, dysphagia. At the motor level: distal hypotrophy of 4 limbs, bilateral thenar and hypothenar hypotrophy, interosseous atrophy in hands, presence of spontaneous fasciculations in 4 limbs, predominantly in the arms and forearms; Strength: 3/5 in 4 limbs, hyperreflexia; patellar, achilea, bicipital and tricipital, presence of exhaustible clonus in lower extremities, bilateral babinski. Unable to walk due to quadriparesis. (Figure 1-4)

Complementary studies
Hypochromic Microcytic Anemia, this secondary to metrorrhagia.
Pelvic USG: with uterine myomatosis.
Cervical tomography: no evidence of structural lesions.
MRI of the brain and cervical region: No evidence of structural lesions at the supra or infratentorial level or at the spinal level.
Electromyography: Shows demyelination with axonal degeneration and presence of nerve conduction block in both tibial nerves, suggesting an acute demyelinating component of a lesion in a lower and upper motor neuron. Compatible with ALS disease.

The patient was treated with physical therapy and ISRS due to depression.

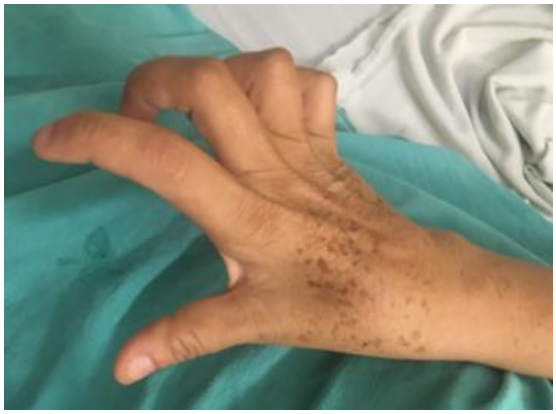

Figure 1. Amyotrophy of the first dorsal interosseous muscle (double anatomical snuffbox sign)

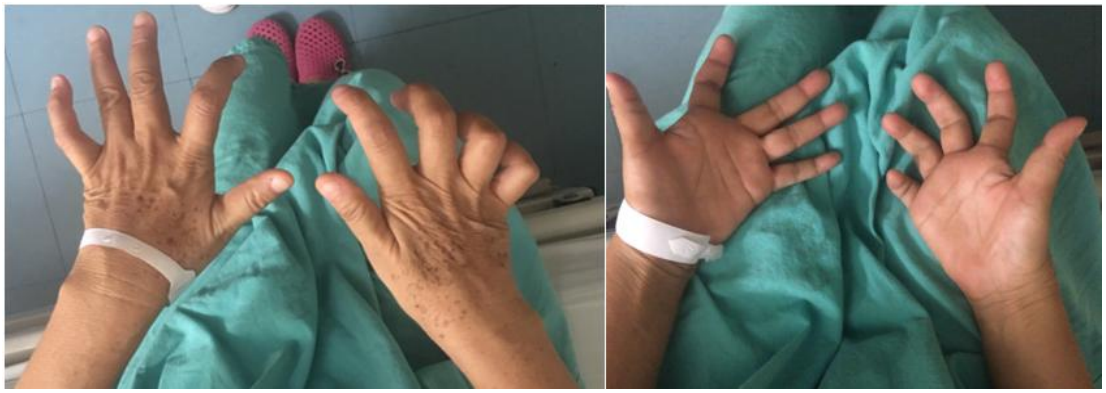

Figure 2. Palmar and dolsar image with significant wasting of the Abductor pollicis brevis and first dorsal interosseous with split hand sign with sparing of the abductor digitiminimi

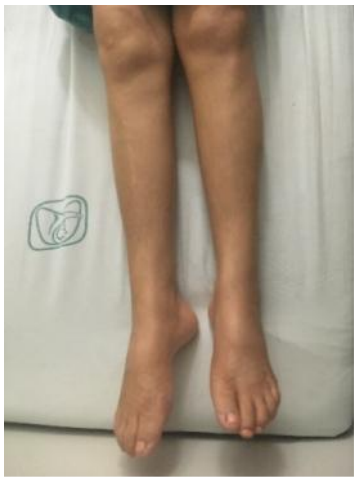

Figure 3. Wasting in lower limbs

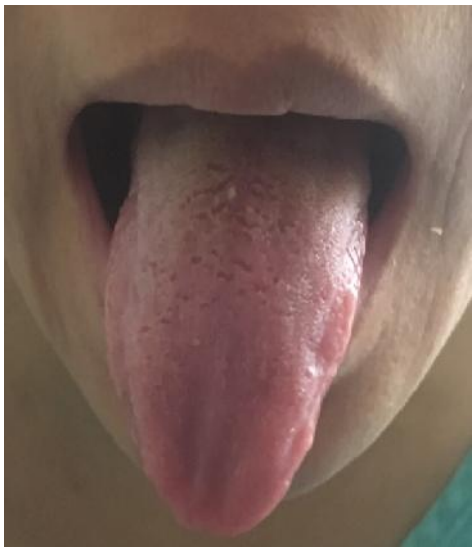

Figure 4. Wasting in tongue

Discussion

In relation to what is described in the specialized literature, in the present case, ALS debuted with the combination of signs and symptoms of degeneration of the first or second motorneuron, accompanied by focal or multifocal involvement with subsequent progression to other regions.

In this case, no information is reported to suggest the presence of the genetic factor. Although the patient in question has not reached the most critical stage of the disease, the evolution of the disease is irreversible, leading the affected individual to almost total immobility, with severe swallowing disorders and restrictive respiratory failure that, in the end, are decisive for death to occur.

The diagnosis of ALS can be delayed between 13-18 months between the onset of symptoms and confirmation of the diagnosis; Since there is no definitive test, the diagnosis is clinical and based on the exclusion of other causes. At the end of the 90's the diagnostic criteria "El Escorial" and "Airlie House Criteria" were created, which were essentially clinical; they are currently used as inclusion criteria for clinical trials and research[3].

The criteria of Awaji are the most used at present since it is based on the findings of electromyography (EMG) together with clinical findings to establish the diagnosis.[4] To establish the definitive diagnosis, it must be demonstrated: evidence of neuronal degeneration upper and lower motor, extension of neurological signs and / or symptoms to the same or another anatomical region (such as the bulbar, cervical, thoracic or lumbar region); plus the exclusion of other possible pathologies through laboratory, EMG and neuroimaging studies. However, the sensitivity of these criteria is challenging, especially in the early stages of the disease, limiting and leading to delays in diagnosis. (Table 1)

Table 1. Awaji criteria for the diagnosis of ALS

Presence of the following characteristics	Absence of the following characteristics
Evidence of second motor neuron degeneration due to clinical, electrophysiological or neuropathological data	Electrophysiological or neuropathological alterations of other diseases that could explain the involvement of the first and / or second motor neuron
Evidence of first motor neuron degeneration by clinic	2. Alterations in neuroimaging suggestive of disease that could explain clinical and electrophysiological aletrations
Progression of symptoms and signs by clinical history, examination and electrophysiological signs	

The standard diagnostic and stratification parameters of ALS do not include the cognitive or behavioral status of the patient. However, detection of cognitive and behavioral changes is important as executive impairment is associated with a faster disease trajectory and behavioral changes are associated with a greater burden on the caregiver. Several screening tools have been designed to identify ALS patients who present with cognitive-behavioral changes in the clinic, such as the Edinburgh Cognitive and Behavioral ALS Screen (ECAS), which has high sensitivity, but a lower degree of specificity[5]*

Differential diagnostics:

Patients with early presentations of the disease, with slow progression or with disorders of the peripheral nervous system can be misdiagnosed in up to 7% -8% .15 Some of these pathologies present purely bulbar clinical symptoms, involving only the NMS or the IMN, which can be a variant of ALS, a syndrome that mimics ALS or a neurological disorder with a better prognosis[6].

The most common pathologies to rule out are: cervical myelopathy, multifocal motor neuropathy, hereditary spastic paraplegia, adrenomyeloneuropathy, myasthenia gravis, Lambert-Eaton myasthenic syndrome, inclusion body myositis, plexopathy, peripheral neuropathy, among others[7,8].

Conclusion: Amyotrophic lateral sclerosis is a rare disease of the central nervous system, characterized by progressive degeneration with a serious prognosis for those who suffer from it. An early diagnosis is important in these patients. In this case the patient presented with a typical presentation with a survival superior to the reported in literature.

The work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

Funding:

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

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Website: www.ijarm.com

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Quick Response Code

DOI:10.22192/ijamr.2021.08.01.004

How to cite this article:
Andres Aguilar Silva, Marco Antonio Santos Diaz, Luis Del Carpio Orantes,Liz Ariadna Gómez Rascón. (2021). Amyotrophic lateral sclerosis: Case report. Int. J. Adv. Multidiscip. Res. 8(1): 25-30.
DOI: http://dx.doi.org/10.22192/ijamr.2021.08.01.004