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ORIGINAL ARTICLE

Relevance of the pyramidal syndrome in amyotrophic lateral sclerosis☆

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Hypertonia

Abstract
Introduction: Pyramidal signs (hyperreflexia, spasticity, Babinski sign) are essential for the diagnosis of amyotrophic lateral sclerosis (ALS). However, these signs are not always present at onset and may vary over time, besides which their role in disease evolution is controversial. Our goal was to describe which pyramidal signs were present and how they evolved in a cohort of patients with ALS, as well as their role in prognosis.
Methods: Retrospective analysis of prospectively collected patients diagnosed with ALS in our centre from 1990 to 2015.
Results: Of a total of 130 patients with ALS, 34 (26.1%) patients showed no pyramidal signs at the first visit while 15 (11.5%) had a complete pyramidal syndrome. Of those patients without initial pyramidal signs, mean time of appearance of the first signs was 4.5 months. Babinski sign was positive in 64 (49.2%) patients, hyperreflexia in 90 (69.2%) and 22 (16.9%) patients had spasticity. Pyramidal signs tended to remain unchanged over time, although they seem to appear at later stages or even disappear with time in some patients.
We found no association between survival and the presence of changes to pyramidal signs, although decreased spasticity was associated with greater clinical deterioration (ALSFR scale) ($P < .001$).
Conclusion: A quarter of patients with ALS initially showed no pyramidal signs and in some cases they even disappear over time. These data support the need for tools that assess the pyramidal tract.

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PALABRAS CLAVE
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Hipertonía

Relevancia del síndrome piramidal en la esclerosis lateral amiotrófica

Resumen
Introducción: Los signos piramidales (hiperreflexia, espasticidad, signo de Babinski) son fundamentales para el diagnóstico de esclerosis lateral amiotrófica (ELA). Sin embargo, no siempre están presentes al comienzo, pueden variar con el tiempo y es controvertido su papel en la evolución. El objetivo del estudio es describir qué signos piramidales están presentes inicialmente y cómo evolucionan en una cohorte de pacientes con ELA, así como su papel pronóstico.
Métodos: Análisis retrospectivo de pacientes recogidos de manera prospectiva, diagnosticados de ELA en nuestro centro, desde 1990 hasta 2015.
Resultados: Del total de 130 pacientes con ELA, 34 (26,1%) no presentaron inicialmente ningún signo piramidal, mientras que 15 (11,5%) presentaban un síndrome piramidal completo. De aquellos pacientes sin piramidalismo inicial, la mediana de aparición de los primeros signos fue de 4,5 meses. El signo de Babinski estaba presente en 64 (49,2%), la hiperreflexia en 90 (69,2%) y en 22 (16,9%) pacientes existía espasticidad. Los signos piramidales tendían a mantenerse inalterados en el tiempo, aunque existe un porcentaje de pacientes en el que aparecen tardíamente o desaparecen con el tiempo.

No se encontró asociación entre supervivencia y la presencia o modificación de signos piramidales, aunque la disminución de la espasticidad se asociaba a mayor deterioro clínico (escala ALSFR) ($p < 0,001$).
Conclusión: Una cuarta parte de pacientes con ELA no presentaron inicialmente ningún signo piramidal y, en algunos casos, estos desaparecen con el tiempo. Esto resalta la necesidad de la inclusión de herramientas para la valoración de la vía piramidal.

Introduction

Combined degeneration of the upper and lower motor neurons constitutes the pathological hallmark of amyotrophic lateral sclerosis (ALS).[1] The diagnosis of ALS requires evidence of lower motor neuron damage from clinical or neurophysiological studies,[2] combined with evidence of upper motor neuron damage, which is determined by clinical examination. Pyramidal signs are not always present at disease onset, however, and may change over the course of the disease. Assessment of these signs may be subject to inter- and intraexaminer variations. Furthermore, their role in disease progression and prognosis is yet to be determined.[3,4]

The purpose of this study is to conduct a descriptive analysis of the presence of pyramidal signs in a cohort of patients with ALS, both at baseline and throughout the course of the disease, in addition to studying the association between pyramidal signs and prognosis.

Methods

We conducted a retrospective analysis of patients diagnosed with motor neuron disease at our centre between 1991 and 2015, using a prospective database. The analysis included those patients meeting the revised El Escorial diagnostic criteria[2] for probable or definite ALS and gathered data on clinical, epidemiological, and functional (ALSFRS scores at 6 and 12 months) variables and survival data (from symptom onset to death). We excluded those patients with pure upper (primary lateral sclerosis) or lower motor neuron disease (progressive muscle atrophy) and those for whom no data on disease progression were available. The following variables related to pyramidal signs were recorded: muscle tone (increased, decreased, normal), deep tendon reflexes (hyperactive, present, diminished/absent), and plantar reflex (extensor, flexor, indifferent); pyramidal signs were evaluated at baseline and at every follow-up consultation (mean of 3 months between visits) until the last consultation (before the patient died or the study ended). Pyramidal signs were considered to be present when they affected at least 2 limbs, in the case of hypertonia and hyperreflexia, or at least one limb, in the case of the Babinski sign. Changes in a pyramidal sign were recorded as such when they were observed in at least 2 consecutive visits. Neurological examinations were performed by 2 expert neurologists. The study was approved by the ethics committee at our centre, Hospital del Mar, in Barcelona.

Statistical analysis

Comparisons between 3 groups were conducted using the chi-square test for categorical variables and the Kruskal-Wallis test for quantitative variables. Comparisons between 2 groups were conducted using the Mann-Whitney U test for quantitative variables and the Fisher exact test for categorical variables. Survival times were analysed using the Kaplan-Meier method; Kaplan-Meier curves were compared with the log-rank test. The Cox regression model was used