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Prefrontal Symptomatology in People with Amyotrophic Lateral Sclerosis: Clinical Utility and Psychometric Properties of Prefrontal Symptoms Inventory (PSI)

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Abstract

The Amyotrophic Lateral Sclerosis (ALS) is a multisystemic disorder. It is considered a neuromuscular disease but also involves cognitive (executive functions, social cognition, attention, memory and language), emotional or behavior changes in over 50% of the reported cases and to of frontotemporal diagnosis lobar degeneration of behavioral variant in up to 15% of the cases. For this reason, the presence of cognitive and motivational problems was analysed in a Spanish sample of ALS patients through the prefrontal symptoms inventory (PSI) to determine applicability in this disease STI. A sample of 31 patients with a potential ALS or definitive diagnostic criteria according to El Escorial was used. Obtained results ISP were compared with a sample of 31 healthy people in the same proportions of gender, age and education level. Obtained results showed a not significant difference between the two populations in the motivational factor problems, related to the depression symptomatology frequently associated with ALS. A significant positive correlation between age at diagnosis and the scale of the motivational problems was observed, with a not significant trend related to problems in the executive control and in social the behavior control and with the age at diagnosis, in the same sense with age at diagnosis. Therefore, it can be concluded that, despite the results obtained, emotional and behavioral deficits in ALS patients and symptoms related to frontotemporal dementia (among others, anosognosia or lack of consciousness symptoms) could have interfered in patient perception about their symptomatology.

Keywords: Amyotrophic lateral sclerosis; Frontotemporal dementia; Cognitive dysfunction

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Summary

Amyotrophic lateral sclerosis (ALS) is a multisystem disorder, which besides being considered as neuromuscular disease, presents cognitive changes (executive functions, social cognition, attention, memory and language), emotional or behavioral in more than 50% of the cases described and up to 15% degeneration diagnosed frontotemporal lobar (DFT) behavioral variant. For this reason, the presence of cognitive and motivational problems analyzed in a Spanish sample of people with ALS by Prefrontal Symptoms Inventory (PSI) to determine the applicability of it in this disease. The sample consisted of 31 patients with probable or definitive diagnosis of ALS was used as the criteria of the Dump. The results of the PSI with a group of 31 healthy people with the same proportions of gender, age, and educational level were compared. As for the results, no significant difference between the two populations, factor motivational problems, often related to depressive symptoms in ALS is observed. Significant positive correlation between age was also obtained for diagnosis and motivational problems scale was received, observing a nonsignificant tendency to correlate problems in the Executive and Problem Control in the Control Social Behavior, equally and in the same direction with the age at which the disease was diagnosed. Therefore it can be concluded that, despite the results obtained.

Introduction

Amyotrophic lateral sclerosis (ALS) is a multisystem disorder in which both corticospinal motor neurons motor-neurons or higher Betz cells in humans-such as spinal motor neurons degenerate progressively. This leads to the failure of the motor neuron circuits and the neuromuscular system with loss of voluntary movement [1].

In ALS patients, brain pathology may show cortical atrophy of the precentral gyrus and adjacent cortices [2,3]. Histopathology shows loss of pyramidal cells of Betz giant and surrounding motor neurons of the motor cortex, the cranial nerve nuclei and anterior horn cells of the spinal cord.

Although ALS is considered a pure neuromuscular disease found cognitive, emotional or behavioral changes more than 50% of the cases described, with important prognostic implications, and up to 15% meet diagnostic criteria for frontotemporal lobar degeneration (DFT) of behavioral [4-5] variant. Thus, alterations have been described in executive functions, in upper attentional and working memory management [5], as in cognitive flexibility [6] and concept formation [7]. Other authors suggest that 35-40% of patients have impaired speech [8], verbal fluency [9], with anomie and syntactic involvement [10]. They have also described alterations of memory [11],

With regard to social cognition, described that 30% of people with ALS without DFT have trouble detecting social grievances in recognition tasks bloopers (Faux Pas) [12,13] and have alterations taking decisions on task Iowa gambling [14]. Furthermore, 40-80% of patients present apathy, some authors being the most frequent alteration [15-16] along with the reduction of empathy, egocentrismo increase the ability to predict and mental states the other to act on [17,18].

This paper analyzes the presence of cognitive, motivational problems of emotional control in a Spanish sample with ALS by Prefrontals Symptom Inventory (PSI) [19]. It is a self-report questionnaire with excellent psychometric properties has been successfully used in different clinical and non-clinical populations, from subjective memory complaints [20], chronic [21] pain, personality disorders [22], addictions [23] or acquired brain injury and degenerative dementias [24]. It also presents optimal indicators convergent validity with neuropsychological tests [25] so it is a tool of choice when describing the prefrontal symptoms in daily life.

Material and Methods

Participants

The clinical sample consisted of 31 people, 19 men and 12 women, all with probable or definite diagnosis of ALS according to the El Escorial criteria [26] of a clinic in Valencia. The mean age at diagnosis received was 52.6 months (SD=11.1). The average time that the sample had diagnosed was 36.2 months (SD=35.5) with a range between 12 and 203 months.

The general population sample was obtained from the ISP database obtained for validation. This sample was recruited from psychologists from different entities involved in the project, which administered to people around them. The criteria for obtaining were: (1) that people were older and completed the test voluntarily; (2) that lacked a history of neurological diseases or disorders diagnosed psychopathological; (3) that recruited people present the greatest possible variability in terms of age, sex and educational level achieved. Based on this data subjects have the same sex, age and educational level was sought that participants in the clinical sample. When they appeared several subjects who met these conditions, It was selected randomly to one of them. Table 1 descriptive of both samples, as shown, are identical in both cases is.

Instruments

1=Rarely; 2=Sometimes yes and sometimes no, 3 the Symptom Inventory Prefrontals (ISP) [19], of 46 items, which responds on a scale of Likert (0=never or almost never administered=often, 4=always or almost always). The factorial study found a solution of 3 factors were named: (i) implementation problems, (ii) Problems in emotional control and (iii) Problems in social behavior. The first factor to be unfolded into three sub-factors, namely Attentional Problems, Problems of Control Executive and motivational problems. Validation in Spanish population reported adequate internal consistency values (.87>α>.81), suitable indicators for adjustment factor solution and concurrent validity with similar tests.

Table 1. Descriptions of the clinical sample and the general population.

	Clinical Sample	General Population
Age		
Mean	55.45	55.45
DT	10.1	10.1
Rank	37-79	37-79
Sex		
Males	19	19
Women	12	12
Level studies		
Primary or less	5	5
Compulsory secondary	7	7
Secondary postcompulsory	9	9
University	10	10

Process

After filling a basic demographic data (age, sex and educational level) the ISP according to criteria described in the original publication [19] was administered. The inventory was applied in a session of 10 to 15 minutes Neuropsychologists properly trained in similar conditions identical material.

To ensure the protection of personal information and medical documentation of patients this study was approved by the Ethics Committee on Human Research Ethics Commission on Experimental Research of the University of Valencia. All participants signed an informed consent and legally authorizing different interventions and study procedures performed.

Analysis of data

One mancova was used to estimate differences between the scores obtained in each sample scales ISP. Wilks' Lambda (λ) was used to estimate the interaction effect of demographic variables. Spearman's rho (ρ) for calculating correlations applied.

Results

Scores on the scales and subscales ISP between the two samples were compared. First, it was found that there was no interaction effect in any of the demographic variables, which was fulfilled in all cases: sex (λ_6=0.81; p=0.08), age (λ_6=0.80; p=0.06) and educational level (λ_6=0.88; p=0.31). In Table 2 the differences between the two samples are shown. As can be seen, none of the differences reached statistical significance.

He was studied in clinical shows the possible correlation between the age at diagnosis is received and the time elapsed and the score on scales ISP (Table 3). It was observed that given positive significant correlation between age at diagnosis and motivational problems scale, so that the greater the older perception in this item was received. You can also see a nonsignificant by correlating problems in the Executive and Control Problems in the Control of Social Behavior, Social equally and in the same direction with the age at which the disease was

Table 2. Differences between the clinical sample and the general population on the scales and the total score of the ISP.

	Clinical Sample		General Population			
	Half	DT	Half	DT	F1	S.I.G.
Motivational problems	7.03	5,771	9.39	4,153	3.4	0.07
Issues in the Executive Control	10.26	8,446	11.58	7,754	0.4	0,52
Attentional problems	6.42	5,137	7.00	3,821	0.3	0,62
Control problems of Social Behavior	6.68	7,525	7.03	4,813	0.0	0,83
Emotional problems Control	7.58	4,945	9.00	3,152	1.8	0.18
Total Symptoms Prefrontal	37.97	26.989	44,00	18.907	1.0	0,31

Table 3. Correlations between age at diagnosis and time since then and the score on the scales of the ISP received.

	Age at diagnosis was received	Months from diagnosis
motivational problems	0.02*	0.23
Issues in the Executive Control	0.07	0,31
Attentional problems	0.13	0.23
Control problems of Social Behavior	0.07	0.20
Emotional problems Control	-0.14	0.28
Total Symptoms Prefrontal	0.10	0.33

*Statistically significant $p<0.05$ differences.

diagnosed trend.

Discussion and Comments

In diseases such as ALS, depending on the patient's profile, it is sometimes difficult to carry out neuropsychological and radiological exams, so it may be desirable to have other tests to obtain valid information in patients with ALS. In this regard, the use of ISP was initially proposed as a valid reason for collecting information cognitive and emotional profile in these situations alternative. The ISP [19] has good psychometric properties and has been used in various clinical and nonclinical populations, including various diseases with involvement prefrontal both degenerative (dementias [24]), as degenerative (addictions [23]). The questionnaire also shows optimum indicators of convergent validity with neuropsychological tests [25],

Given the neuropathological and radiological evidence of pathology of the frontal and temporal lobe in ALS, it is expected that both a neuropsychological evaluation and patient responses on a self completed questionnaire reflected neuropsychological deficits appear. However, ALS is a neuromuscular disease that has not only cognitive, but also emotional and behavioral up to 50% of documented cases. In addition to the expected level cognitive deficits (executive functions, social cognition, memory and language), changes may occur in behavior that sometimes fail to meet criteria for frontotemporal dementia. ALS patient may present different profiles.

These changes relate to aspects such as apathy or characteristics of frontotemporal variant described by Rascovsky [26-29] as disinhibition, loss of sympathy and empathy, persistent, stereotyped or compulsive behavior, hyperorality, loss of perception or psychotic symptoms (somatic delusions, hallucinations, irrational beliefs).

Insight loss is a key criterion for the diagnosis of frontotemporal dementia [30], which is already known that is associated with motor neuron diseases. However, it has rarely been studied in ELA [31]. Related to the loss of insight is the inability to recognize the neurological symptoms or anosognosia [32,33]. Although the term originally was used to refer to the fact that a patient was not aware of the disease that had or hemiplegia that had, in the last decade is used to describe a lack of recognition of any symptom acquired specific to the disease.

It has been suggested that anosognosia is an early sign of development of dementia in ALS patients [34]. In addition, clinical and neuroradiological level [35] has shown that anosognosia is associated with alterations of the right frontal lobe and frontal executive function, so you would expect that increasing the frontal involvement, greater executive dysfunction and therefore greater the probability that the patient has anosognosia.

Although the results we have obtained in the tested sample are not significant in any of the scales that form the ISP, next to statistical significance on the scale of motivational problems results and a trend to significance on the scale seen Emotional control problems. The scale of motivational problems evaluates issues related to the momentum and interest in initiating behavior, whose deficit is associated with ventromedial dysfunction [36,37], whereas the scale of problems in emotional control includes heterogeneous aspects, such as aggression or lability, which frequently occur in patients with orbitofrontal [20] injuries.

Regarding the results obtained for the correlations of the scales with the patient's age and the time that has passed since the diagnosis of ALS, again statistically significant relationship was observed in the Scale of motivational problems and in this case, having the patient age when diagnosed; finding that the greater the age, the greater perception and self-reference of motivational problems. Also, there is a trend towards significance between the results obtained on the scales of Problems in Executive Control and control problems of social behavior with time since diagnosis, so that a longer period of time from the obtaining diagnosis, the patient reported increased deficit executive and biggest problem in controlling social behavior. Motivation first, followed by the executive functions and the adequacy of social behavior, seem to be aspects of ALS patient is able to perceive as the most affected, with increasing time since diagnosis. ALS patients tend to show depressive symptoms and often underestimate their physical function, so it would have been expected that scores on the ISP will be more significant. However, it is possible that so it would have been expected that scores on the ISP will be more significant. However, it is possible that so it would have been expected that scores on the ISP will be more significant. However, it is possible thatthe inability to recognize or anosognosia neurological symptoms, have negatively interfered in completing the questionnaires. However, this could partly explain the most significant differences between the clinical sample of this study and in the general population, they are obtained in the motivational factor problems. Also noteworthy is that the time range of diagnosing patients in the sample is very large (12 to 203 months), so that the evolution of cognitive, emotional and behavioral of each deficit may be quite different, which you may have interfered significantly on the results obtained in this regard.

Moreover, the alterations that the decision [14] can be a confounding factor in the responses to the questionnaire completed by patients.

To reduce interference Anosognosia on the results, it would be questionnaires were completed, not only for ALS patients, but also for a reliable external observer as a family; and analyze the possible existence of differences between some answers and others. Another strategy that arises, is to use an assessment of that component, which allows to

validate the responses of people with ALS in the ISP. Scale Anosognosia of Deckel and Morrison (1996) [38], is a 5-point scale self-assessed by the patient and physician/therapeutic staff, evaluating their activities of daily living (walking, using accurately and rapidly hands and fingers, speak clearly, memory, concentration and attention, sitting still and silent, say the words you're thinking, emotional control). Finally, make a neuropsychological evaluation of the functions that appear most often affected in ALS, patients would confirm whether the sample have attentional executive involvement and objectified [25,39,40].

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