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Sclerosis Lateral Amyotrophic, A Vision From Palliative Care

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Abstract

This article is a case study of symptomatic management of Amyotrophic Lateral Sclerosis (ALS) in Spain by a home care and support teams program in the Public Health Service. The goals during the evolution of this disease were thoroughly treated by careers, as well as on the institutional and systemic framework. Family involvement and intervention were essential to define the quality of the care. Abilities and attitudes for caring were extraordinary trained to walk with this patient during the last years of his life. Palliative care was guided to understand his suffering and all his needs and to help the caregivers. The purpose of this article is to identify some factors that might contribute to his autonomy and safety.

Keywords: Sclerosis lateral amyotrophic; Management; Needs; Caring

Introduction

Amyotrophic lateral sclerosis (ALS) is a neurodegenerative and progressive disease affecting the motor neuron and causing muscle weakness in the extremities and respiratory and bulbar (oropharyngeal) muscle weakness. It exhibits atrophy, spasticity, weight loss and, ultimately, respiratory failure. It represents the third most frequent degenerative disease, after Alzheimer's and Parkinson's diseases, with an incidence of only 2 cases per 100.000 inhabitants and a prevalence of 6 to 8 per 100.000.

ALS clinical manifestations are a combination of asymmetric weakness and muscular atrophy of the extremities, accompanied by signs of affectation of the first motor neuron (exaggerated reflexes, increased tone, cutaneous extensor plantar reflex). Usually, ALS initially appears either in a single member (a weak hand or a dropped foot) or as a combination of this and bulbar symptoms (dysphagia, dysarthria).

Despite its apparently focal beginning, there is a diffuse involvement of other muscles in most patients. By the time ALS becomes clinically apparent (up to 40% of the anterior horn motor neurons may be lost before motor anomalies can be detected), it generally progresses quickly. Prognosis for patients with a motor neuron disease can be variable, although it usually has a gradual evolution with a fatal end. Around 50% of the patients do not live longer than 2 or 3 years after the diagnosis; only 5 to 30% survive the 5 years following the diagnosis, and 5% of them survive the 10 years following the diagnosis.

Respiratory failure caused by a diaphragmatic paralysis is the leading cause of death. Advanced ages and dyspnea beginning or bulbar symptoms are variables associated with a worse prognosis. Being female and the lack of a stable partner also have a negative impact. The worse prognosis appears in those cases with a fast progression rate in the first 6 months after the diagnosis. It has been shown that psychosocial well-being implies an increased survival [1].

It could be said that the advanced stage of ALS can come defined by the presence of one of the following criterion:

- Need of invasive or non-invasive mechanical respiratory support.
- Need of nasogastric tube or PEG enteral nutrition support.
- Refusal of the abovementioned support measures.

The appropriateness of the Palliative Care welfare approach in ALS can depend on either the prognosis, in an advanced or terminal stage of ALS, or the needs detected in the comprehensive valuation of the patient and their family and the complexity of those needs.

Attention to patients in ALS palliative care is an uncommon aspect in the activity of Palliative Care specific programs.

The key aptitudes for a quality care of these patients [2-4] are: intervention of experienced neurology specialists, treatment with Riluzole, proper monitoring of symptoms under the charge of a multidisciplinary team, improvement of both nutrition and quality of life through the use of percutaneous endoscopic gastrostomy, improvement of survival by using non-invasive mechanical ventilation, and quality of life improvement of patients and their families through the use of PC in the final stages --- are some of the most relevant conclusions and recommendations of the abovementioned revisions.

PC intervention helps understanding the progression of the disease, controlling the symptoms, considering the treatment expectations, addressing disease-related, end of life, and legal issues (mental capacity, wishes), and coordinating with specialists for interventions (e.g.: ventilation). It also helps dealing with the needs for equipment, and provides orientation, support and well-being advice [5].

Clinical Case

50 year old male. No toxic habits, no known medical family history. Traumatic rib fractures. Worked as a butcher.

The patient showed, in 2008, a clinical profile of progressing gait disturbance, needing help to walk up and down the stairs with a feeling of stiffness. Admitted in the neurology service and diagnosed with Amyotrophic lateral sclerosis (ALS). From that moment on, the patient was taken care of by a multidisciplinary team (Neurology, Pneumology, Physiotherapy, Nutrition with support from an NGO Foundation that provides technical aids at home, Social Worker and Psychology) in a hospital's Outpatient Clinic.

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The clinical profile was progressive, and in March 2010, at the age of 46, PADES team (home care and support teams program) started monitoring the case.

He was diagnosed on the first visit with advanced ALS. He lived in his own home with his mother, who moved in with him to become his main carer. Several aspects were examined: high complexity patient, no cognitive impairment (Pfeiffer 0 mistakes), moderate risk of pressure ulcer development (Braden 14). On the first physical examination he showed spastic paraplegia, dysarthria (he communicated through alphabet boards), and dysphagia to solids and occasionally to liquids. At that point, the patient maintained a sitting position most of the day, and even went out in a wheelchair, assisted by a carer from 2 to 3 hours per day. No skin lesions.

As for the emotional aspect, he showed sadness due to loss of autonomy; he went through several progressive grieves, and lost his job as a butcher in the family business. He was handsome and attractive to women, but his intense social life changed gradually. He was separated, with a son, and a second partner with whom he broke up after the diagnosis. He needed his mother's implication and dedication in order to progressively cover more life necessities. Both patient and family were aware of the diagnosis and prognosis. Family relationships were good during the process, although the mother showed failure in her own moral duty at the final stages (bad sleeping, no leisure activities), even though she counted on the help of an external carer 8 hours a day.

On treatment with vitamin E, Escitalopram 15 mg/day, Diazepam 2,5 mg/12hrs, Baclofen 25 mg/8hrs, Tizanidine hydrochloride 2 mg/12hs, Riluzole 50 mg/12hs, cream of magnesia 1/day.

Clinical Course

Nutrition

The patient shows dysphagia to liquids in 2010, so he starts with percutaneous gastrostomy feeding. As a complication, he develops peritonitis, and is discharged with jejunostomy tube. Initial mixed feeding (oral and enteral) until total loss of oral feeding in August 2011. Monitored by a nutrition team and receiving vitamin supplements through a tube. Frequent need of tube replacement (both at home and in the hospital) due to obstruction. Significant constipation managed with lubricants, combined osmotic laxatives and enemas.

Emotional aspect

Active listening and emotional support was performed in every visit, both for the patient and his family and carers. The patient showed sadness and anxiety due to the progressive worsening of the base disease. Treatment with Escitalopram 15 mg/day was initiated, but changed to Mirtazapine 15 mg/day, with partial response, due to ineffectiveness. The patient rejected the treatment and psychological attention that were offered.

During the course of the disease, the patient recounted that he found significant support in part time private careers hired by his family. He used guttural sounds in order to express something was happening to him or that he wanted to say something. Communication with careers and team was always carried out with an alphabet written for these patients: a board with 5 groups of letters (one for the vowels and four for the consonants). They figured out the order of the letters by asking him; if the patient closed his eyes, it meant that was the letter he wanted to point out. As the disease progressed, "communication" slowed down.

Ethical aspect

The patient is always aware of the diagnosis and prognosis of the disease. Several interviews with the patient are performed, where he establishes his advance directives, the first one being, in November 2012, that "he wants everything done", meaning every necessary therapeutic diagnostic test, independently of the disease. He expresses that he is afraid to die. During the second interview, in February 2013, the patient recounts that he wishes to be looked after at home so he does not have to be taken to the hospital or practiced any complementary examination".

He recounted "fear of dying by drowning" in several occasions, so all the guidelines for action in the case of a dyspneic crisis (extra doses of Morphine by PEG, and how it affects the organism) were explained to him as many times as necessary. An empathic response was the dialogue we established in order to deal with this issue. We frequently gave the patient back his own situation, with the aim of walking together towards reality. Ángel also communicated with us through non-verbal language, showing gratitude, peace of mind and a feeling of safety. A less frequent verbalization of the fear of dying by drowning and a better nightly rest were two basic criteria proof of the work carried out.

As a result, both the patient and his family calmed down and felt relieved. All therapeutic decisions were addressed in partnership with the patient and his family, and a document of advance directives was prepared. We agreed with the patient on palliative sedation in the case of refractory dyspnea.

Neurological aspect

Progressive worsening of neurological symptomatology until paraplegia and anarthria, only maintaining ocular motility. Initially, he practiced rehabilitation at home, but it was called off due to a poor tolerance. Spasticity treated with Baclofen with little improvement.

Sialorrhea

Initially treated with Amitriptyline 50 mg/day but increased to 75. Botulinum toxin was injected in both parotid glands (Neuroblock 2000U), but was ineffective. Subcutaneous hyoscine butylbromide was administered without response. Finally, treatment with amitriptyline 75 mg/day taken in three doses was reinitiated, with a better control of secretions as a result.

Respiratory aspect

Progressive dyspnea, non-invasive ventilation (BIPAP) is started in January 2010, 1-2 hours/day, with poor tolerance attributed to bulbar affectation. Cough assist is prescribed in 2012 for a better control of secretions, which he did not tolerate either.

Dyspnea worsens progressively in 2013, with nocturnal flare-up episodes. We thought there was an emotional component, so Morphine through jejunostomy was started in the case of crisis, and Midalozam 7,5mg at night, which was later set as fixed treatment. The patient refused to use NIMV, so domiciliary oxygen was used to control dyspnea. The patient needed frequent extra doses of oral morphine. We tried to proceed to transdermal Fentalyn (subcutaneous), which showed no improvement. Thus, oral morphine in increasing doses (20 mg/4h) and Midazolam 7,5 mg at night (with extra dose depending on symptomatology) were prescribed.

Evolution

During all these years, coordination between healthcare services was always established in a very direct way, over the telephone, and monitoring the case through meetings between the different professionals that looked after him. In these meetings, therapeutic targets were set based on the moment and his needs, from the knowledge of every professional. Thanks to the shared story, a monitoring of the evolution was generated in order to give the most appropriate response.

In November 2013, dyspnea becomes refractory with significant episodes of dyspnea, and the patient, with a diagnosis of refractory dyspnea, agrees to be referred to the hospital, in coordination with the multidisciplinary team that took care of him at the hospital. During admission, the patient needs palliative sedation due to refractory symptom.

One week after exitus of the patient, a follow-up visit with the family (mother and sister) is scheduled in order to start with mourning support.

Pain

He expressed pain and general malaise during the final stages of the disease, which improved with oral morphine.

Discussion

The focal point of addressing neurodegenerative diseases lies in the complexity of psychological and biomedical problems. According to a study conducted by the Irish multidisciplinary units, patients with ALS in these units died later than those who were treated in the general neurology service, and multidisciplinary care benefited especially those patients whose symptoms appeared at bulbar level. In the case shown in this article, six years elapsed from the diagnosis to the death, which gives the patient a higher survival than the average [6].

In this case, we would like to stand out the importance of the multidisciplinary unit, with the help of the Foundation, that contributed to multiple aspects (technical assistance, psychologist, social worker), and the healthcare specialists like the pneumologist, neurologist and dietician. The different assistive devices provided constant and comprehensive support, which was especially helpful when functional dependency increased. PADES (home care and support teams program), a specialized team from the chronic patient support unit, the support from infirmary, the case management and the social value contributed to perform an individualized therapeutic intervention plan [7,8].

The PADES team is formed by a doctor specialized in palliative care, together with 2 nurses. Visits were weekly during moments of stability, and were carried out jointly or alternating doctor and nurse, and increased frequency depending on the need of the patient and his family.

The case manager played an important role as a connecting element between the different health care levels. Processing different orthopaedic, rehabilitative and adjustment aids helped managing the emerging needs during the course of the disease within the framework of the interdisciplinary care [9].

The high economic and emotional cost of ALS were in this case mitigated both by the work of the professionals and the personal and family resources used. The benefit from the Dependency Law complemented the contribution from the family. A 24-hour attention was possible. The lack of information about possible assistance and what social services can offer might produce great emotional stress. Financing and a quick contribution will help to a better confrontation of the disease.

The successful approach to the main problems concerning the care of the patient was possible thanks to the skills and abilities of the careers. Specialists also play an important educational and communication role with the family [10].

The care of the patient served as an example to demonstrate this constructive and inspiring work, a clear example of how you can build a dignified life until the end, making the patient take part in the decision making process concerning his health and giving him full control over the disease [11]. We must look at every need as a combination of different emotional, social, physical and spiritual factors [12]. Every treatment plan has to be thoroughly individualized, providing information in advance for the making of decisions, simultaneously dealing with concepts like hope and keeping in mind personal resilience factors, cognitive barriers, behavioral changes and difficulties in the process of understanding and accepting.

Communication issues and the fear of dying by drowning were two constant causes of suffering, which were thoroughly handled for a long period of time with patience and talent.

Empathy, defined as the attitude in which a person makes a cognitive, affective and behavioral effort in order to understand alien experience, needs, meaning of things, feelings, values, dynamics, expectations, wishes, and resources, implies that our patient had to feel that he was being understood. That and the therapeutic care were the key elements to an efficient support, facilitating his wishes of staying at home as much time as possible, an environment where he felt safe.

During the final days, refractory dyspnea was the decisive cause of wanting to die in the hospital, due to quality and humanization of care reasons, promoting the patient's self-determination and supporting a carer who was emotionally disturbed due to the situation. Both palliative sedation and admission in a Palliative Care Unit are considered for this refractory symptom and its associated fear, which is frequent among these patients [13,14].

Conclusions

Excellence in care depends on the competence of the careers, as well as on the institutional and systemic framework. Interaction between the specialist, the main career and the patient are essential for an appropriate control of symptoms, anticipating the problems or palliating them with our professional behavior, but not enough. The patient needs to know what palliative careers can offer with a proper management and the expected results [15].

Family involvement and intervention are essential to define the quality of the care. But, more especially, the academic exercise, which helps building the meaning of the patient's life. Since there is no such thing as a quality of life scale, I could try answering questions about the interventions with the patient, in order to credit the positive feedback [16].

The healthcare institution belongs to the healthcare system, a macrocosmic superior sphere that influences the distribution of health resources in such complex and incapacitating incurable diseases. Given the importance of the fact that healthcare institutions provide these patients with health resources, the act of caring does not take place in a neutral space, but within an institution that guarantees the

realization of a philosophy, with a series of criteria and guidelines based on palliative care.

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