Emotions and Amyotrophic Lateral Sclerosis
What the Neurologist and Psychiatrist Should Know

Mini Review

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Abstract

Amyotrophic lateral sclerosis is a progressive and fatal neurodegenerative disease affecting motor neurons in the anterior horn of the spinal cord, and brain connections of motor cortex and subcortical areas. Few studies demonstrate the cognitive aspects, and behavioral in Amyotrophic lateral sclerosis, as well as in specific treatment in this area, which can often have a significant influence on the quality of life, as well as to demonstrate clinical parameters of emotional impairment. Understanding the emotional behavioral in Amyotrophic lateral sclerosis may in the future guide the proper management of the patient, evaluation, establishment of treatment and development of innovative interventions that can provide a quality of life for the patient even with the unfavorable prognosis.

Keywords: Amyotrophic Lateral Sclerosis; Emotional Behavioral; Cognition; Theory of Mind

Amyotrophic lateral sclerosis (ALS) belongs to a wider group of neurological disorders known as motor neuron diseases, which are caused by gradual degeneration of anterior horn and pyramidal tract neurons, however, deficits in cognition and behavior are also commonly reported [1,2]. Most patients die from respiratory failure, usually within 3 to 8 years from when the symptoms first appear. However, about 10 percent of people with ALS survive for 10 or more years [2]. Cognitive impairment is correlated with pathologic based on neural circuit’s changes in cerebral cortex, beyond the motor areas and frontotemporal dementia. After clinical evaluation, evidence of impairment can be detected in up to 50 percent of patients through neuropsychological testing [3]. When cognition is impaired

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in ALS, the cognitive and emotional profile is modulated, with difficulties in executive function, including judgment, planning, inhibition, and emotions [4]. Given that social and emotional cognitive difficulties are a core feature of ALS and cause significant functional impairments, researchers have begun to investigate emotional and social cognition in ALS [5]. Higher-level social-cognitive difficulties, such as difficulty predicting others’ thoughts and feelings (i.e. theory of mind, emotional evaluation), have been most commonly investigated in ALS [1,6-8].

Unfortunately, few studies demonstrate the cognitive aspects, behavioral, and emotional symptoms in ALS, as well as in specific treatment in this area, which can often have a significant influence on the quality of life, family members and caregivers [9]. Assessment of the emotional processing in ALS patients should improve comprehension of neurologists\psychiatrists of their adaptive functioning [8,10]. Moderate depressive or anxious symptoms are often observed, but less frequently than in other neurological diseases. “Many neurologists prefer not to face such problems, usually informing their families about the ruthless and overwhelming rapidity of disease progression. We cannot disregard the fears, wants, expectations, and individual needs of our patients. Real needs submerged in inert and quiet bodies. Evaluating the cognitive and behavioral features in these patients is important for therapy and specific care”. In the context, to minimize these aspects of ALS patients, experimental studies about reduced memory capacity (i.e. emotional material), domains of emotional processing and evaluation emotional with social situations should be encouraged. ALS patients have neurodegeneration of cortical, limbic structures such as the amygdala, and nucleus accumbens might affect emotional processing abilities especially for aversive emotional information but environmental factors may also contribute to these changes. Furthermore, reduced afferent peripheral inflow to subcortical and cortical networks such as the limbic system may explain variance in emotional processing and “dampening” of negative feelings [11].

Spirituality/religious belief are recognized internationally as a domain within end-of-life care and it is important in patients’ and carers’ quality-of-life. We should listen more to our patients, questioning about beliefs, spirituality/religions, expectations, and especially their emotional needs in the face of ALS. “More important than prescribing a particular class of anti-depressant medications, is seeking a better compression of the present and past emotions”. O’Brien MR, Clark D [12], studied and reported unsolicited (internet and print) narratives written by patients in advanced stages of ALS. Narratives of 161 diagnosed with ALS during a period of 37 years (from 1968 to 2005) were analyzed thematically. The findings revealed that religious faith sustains and helps people to avoid despair, and personal spirituality helps them make sense of what is happening to them. “Assessment of religious or spiritual needs should become a routine part of practice and is the responsibility of all members of the multidisciplinary team”.

Theory of Mind (ToM), the ability to recognize thoughts and emotions of another, may be one of the cognitive domains affected in amyotrophic lateral sclerosis (ALS). Social and emotional cognition in ALS has recently become the focus of research despite dysfunction of these processes being an early diagnostic feature of frontotemporal dementia [13]. Patients with frontotemporal dementia display deficits in ToM, empathy and emotion recognition [14]. ToM refers to a person’s ability to infer mental states of oneself and others such as beliefs, preferences and intentions, and aids the understanding of other people’s behaviour. Studies indicate some dysfunction in ALS and deficits in ToM have now been reported using relatively complex cartoons and stories [15]. Patients with ALS and frontotemporal dementia have also shown impairment on tasks of social and emotional cognition. The results showed that the ToM processes involved in this task have been further fractionated into cognitive and affective subcomponents, measuring the recognition of thoughts (beliefs and intentions) and feelings (emotions), respectively [13]. Deficits for affective judgment has been found in patients with damage to the ventromedial prefrontal cortex.

In order to study early changes in the brains of individuals with ALS based on ToM, Trojsi., et al [16] analyzed a group of 21 patients (9 bulbar onset (ALS-B) and 12 spinal onset (ALS-S), by functional magnetic resonance imaging (MRI). The results showed a reduced activity mainly in the areas of neuropsychological performance, including cognitive and affective ToM and multi-domain cognitive functions. ALS-B subgroup exhibited a significant impairment of both affective and cognitive ToM subcomponents, whereas the ALS-S group

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showed a significant impairment of the cognitive subcomponent alone. At the end of the study, it was also raised the hypothesis that ALS-B is biologically more aggressive with respect to cognitive behavior and other modalities, including emotions.

Girardi., *et al.* [15] highlighted a ToM deficit in patients with ALS using a preference judgement task. Their findings not only revealed a ToM deficit in this executively undemanding task, but also showed that this deficit was unrelated to the presence of executive dysfunction in emotional aspects. Meier., *et al.* [17] found that patients with ALS identified significantly fewer faux-pas stories than controls did, but were just as accurate in identifying control stories. Although the faux-pas task involves not only representing the knowledge or beliefs (cognitive ToM) of others but also appreciating their emotional state (affective ToM), the authors suggested that the deficit was more likely to lie primarily with the affective ToM component of the task. Moreover, when they repeated the analyses, co-varying out performance on oral letter fluency, the findings were unchanged, indicating that the ALS group had real difficulty with the faux-pas stories with emotional context [18].

Despite recent neurocomputing programs advances, whether positive and negative emotions networks can be voluntarily modulated is still unknown. For this purpose, Li Z., *et al.* [19] used a multivariate voxel pattern analysis and real-time functional MRI neurofeedback (rtfMRI-nf) aiming to clarify this issue. During an emotion regulation task, participants’ emotional states (positive or negative) were given to them as feedback. Individuals were able to increase the percentage of positive emotional states, enhancing emotion regulation network activities. Participants showed an improvement on the positive subscale of positive and negative affect scale that came close to significance. In addition, the activation of emotion-related brain regions, including amygdala, anterior cingulate cortex, insula and dorsomedial prefrontal cortex, was also increased during rtfMRI-nf training task. Authors consider and support the hypothesis that humans are able to voluntarily modulate positive emotion networks, leading to exciting applications in the treatment of various neurological and psychiatric disorders, such as ALS and other motor neuron diseases.

Neurologists as well as psychiatrists should be able to identify, medicate (if necessary) and point out to the multidisciplinary team, the emotional problems presented by patients with ALS, in order to offer directed, updated and individualized treatment. The numerous studies presented above allow us to believe that the science of emotions is progressing. Due to this fact, we must be constantly updated on the subject.

**Bibliography**

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