Chapter 15
Amyotrophic Lateral Sclerosis: A Predominant Form of Degenerative Disease of the Motor Neuron System

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ABSTRACT

Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disorder (ND) that primarily comprises the neurons responsible for controlling voluntary muscle movement. The unique neuropathologic findings include anterior horn cell degeneration producing muscle atrophy or amyotrophy, degeneration, and sclerosis of the corticospinal tracts. It is a common neuromuscular disease worldwide and has been identified in people of all races. There seems to be neither identified risk factors nor family history associated with most of the documented ALS cases. There exists no treatment for ALS that can prevent neither its progression nor reverse its development. However, there are treatments available that can help control symptoms, prevent unnecessary complications, and make living with the disease easier. This chapter extensively discusses this neurodegenerative disorder based on the currently available knowledge on the condition.

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INTRODUCTION

ALS, also known as Charcot's or Lou Gehrig's disease, is a progressive and deadly neurodegenerative disorder (ND) which greatly impacts on patient's quality of life and results in death 2-5 years after diagnosis (Pasinelli & Brown, 2006). From its underlying pathophysiology, the disease was named, that is "amyotrophic" referring to atrophy or death of muscle fibers and "lateral sclerosis" referring to stiffness or hardness of the lateral column of the spinal cord as fibrous astrocytes replace degenerated axons of the upper motor neuron (Armon & Lorenzo, 2017). The disease is associated with the loss of motor neurons that regulate voluntary muscle activities which include walking, breathing, chewing, talking etc. Most importantly, neurons of the spinal cord, brainstem and those present in the motor cortex (Stamenkovic et al, 2017). The effect on the motor neurons accounts for it being also called motor neuron disease (MND).

Clinically, the hallmark of ALS is the presence of upper motor neuron (UMN) and lower motor neuron (LMN) features involving the brainstem and multiple spinal cord regions of innervation. Patients with ALS can present either with bulbar-onset disease (about 25%), limb-onset disease (about 70%), or initial trunk or respiratory involvement (5%), which subsequently spreads to involve other regions (Vucic et al, 2007). Atypical modes of presentation may include weight loss, which is indicative of poor prognosis, cramps and fasciculations in the absence of muscle weakness, emotional lability, and frontal lobe-type cognitive dysfunction (Ferguson & Elman, 2007).

Currently, there is no defined diagnostic test or biomarker for ALS hence experts only rely on its clinical presentation to diagnose. Henceforth, research to establish a novel biomarker that accurately assesses the progression of the condition is of paramount interest in improving therapeutic trial design while also decreasing cost of clinical trials. Lately, it is being progressively identified that population registries are important additions to improved clinical assessment techniques. These collaborative activities will certainly lead to a better understanding of ALS and its often random development, and will lead to the establishment of guidelines for better care of patients (Kiernan et al, 2011).

It is believed that ALS may differ based on clinical phenotype with longer survival reported in ALS without cognitive impairment (Montuschi et al, 2015). The chapter therefore discusses ALS as informed by currently available knowledge on this ND.

BACKGROUND

ALS is a clinically and genetically heterogeneous, devastating, rapidly progressive neurodegenerative motor disorder with cognitive and behavioral impairments as core features (Schmidt et al, 2016). Alongside behavioral features, executive dysfunction is present in up to 50% of ALS patients (Beeldman et al, 2016). Anatomical connectivity studies have revealed clear white matter impairments, mostly affecting tracts directly linked to the motor cortex (Schmidt et al, 2014).

Improvements in our knowledge of the glutamate neurotransmitter system coupled with the detection of causal genes associated with the progression of familial ALS (fALS) have inspired research interest (Kiernan et al, 2011). Whereas these research findings allow for vital comprehensions regarding the ultimate consequences of ALS on the brain, the causal pathogenic mechanism of ALS remains largely unknown (Schmidt et al, 2016).