International Journal for Multidisciplinary Research (IJFMR)
E-ISSN: 2582-2160 ● Website: www.ijfmr.com ● Email: editor@ijfmr.com

Amyotrophic Lateral Sclerosis with Frontal Lobe Syndrome: A Case Report

Rubina Mariam Oommen[1], Amrutha Gopal[2]

[1,2]Pharm D Intern, The Dale View College Of Pharmacy And Research Centre, Punalal Po, Trivandrum, Kerala, India

ABSTRACT

Amyotrophic lateral sclerosis, also known as ALS, is a neurological illness that gradually damages brain and spinal cord nerve cells. Muscle function will eventually be lost in those with ALS. Although the exact cause is unknown, genetic, and environmental factors might be at play. Early signs and symptoms frequently include slurred speech, unusual limb weariness, cramps and muscle twitches, and clumsiness. As the illness worsens, a person suffers symptoms all over their body. Since there is currently no known cure for ALS, the only available treatments are symptom management and halting the disease's development. FDA-approved drugs like Edaravone and Riluzole have some advantages in terms of prolonging survival and maintaining function.

We report a 62-year-old male patient with atrophy, twitching, and muscle weakness in a case consistent with ALS symptoms.

INTRODUCTION

ALS, previously known as Lou Gehrig's disease, is a neurological condition impacting motor neurons, the nerve cells controlling voluntary muscle movement and breathing in the brain and spinal cord. When motor neurons degenerate and are destroyed, they cease transmitting messages to the muscles, leading to muscle weakness, twitching (fasciculations), and atrophy. Consequently, individuals with ALS gradually lose the brain's ability to initiate and regulate voluntary movements like walking, talking, chewing, and other functions, as well as breathing as the disease progresses. [1]

As the motor neurons (nerve cells) deteriorate, they lose the ability to transmit signals to the muscles. ALS typically impacts both upper and lower motor neurons. You may have different symptoms if upper or mostly lower neurons are affected.

Breathing can become difficult or ineffective due to ALS, as it impacts the muscles responsible for chest and lung movement. With ALS, you may have pulmonary (breathing) issues such as dyspnoea, even when not moving, weak cough, breathing, and throat cleaning is difficult, surplus saliva, inability to lie completely flat in the bed, recurring pneumonia, chest infection, and respiratory malfunction.

Two categories are used to classify ALS based on its origin:

1. Sporadic ALS: Between 90% and 95% of ALS cases are incidental. This indicates that the illness occurs at random. There is no known risk factor or family history of the associated disease.
2. Familial ALS: Also referred to as genetic ALS, it affects between 5% to 10% of ALS patients. It occurs when you receive the illness from one or both of your parents. Genetic alterations or mutations cause the condition, which is inherited in families.[2]

The primary symptom of ALS which occurs in 70% of patients is limb weakness in one arm or both arms or legs. Less than 5% of patients initially experience respiratory muscular weakness, while about 25% initially experience impairment in their speaking, chewing, and swallowing muscles. Growing weakness in the arms and legs, twitching of the muscles, cramping or stiffness in the limbs, delayed, slurred, or stopped speech, trouble swallowing or chewing, a weak cough, and shortness of breath are common signs of advanced ALS. Dementia affects 5% to 15% of ALS patients, and many may have mild personality changes or a reduction in their capacity to manage their emotions.

Electromyography and nerve conduction studies, procedures that assess muscle and nerve function, and the patient's symptoms and physical examination results, are used to diagnose ALS. To rule out other illnesses, people with suspected ALS typically get brain and spine magnetic resonance imaging (MRI). Some patients may also have lumbar punctures, specific blood tests, or muscle biopsies.

While there is presently no cure for ALS, there are several medications that may slow the disease's progression. An oral drug called Riluzole can extend survival by three months on average. When administered intravenously or orally, Edaravone has the potential to somewhat slow the advancement of ALS.[3]

Frontal lobe syndrome refers to impairment of the higher functioning parts of the brain, such as language and speech production, planning, motivation, and social conduct. The term is frequently used to describe a clinical condition that arises due to damage or malfunction in the prefrontal cortex.[4]

The characteristic feature of ALS is progressive degeneration of both lower and higher motor neurons. Additionally, up to 50% of patients experience mild cognitive and behavioral impairment due to brain degeneration involving the frontal and temporal lobes, which extends beyond the motor cortex.[5]

CASE REPORT

A 62-year-old male patient complained of slurred speech, increased laughter, walking difficulties, and muscle cramping when he was hospitalized in the neurology department one year ago. Physical examination confirmed that the tongue was atrophic and showed positive fasciculations. Additionally, there was wasting of the supraspinatus and infraspinatus muscles, contracture of the right tendon Achilles more than the left, spastic gait, upgoing plantar reflexes in both legs, accentuated deep tendon reflexes, and retention of the right drop foot and abdominal reflexes. The lower leg did not vibrate; there was only a slight pinprick sensation.

On evaluation, the brain's MRI confirmed mild widespread atrophy and slight signal alterations on both sides; the lower limb's NCV revealed decreased conduction velocity; and the hand's small muscles' EMG revealed reduced amplitude and reduced recruitment. The bulbar, corticobulbar, and corticospinal structures were affected, which is consistent with TDP-43 most likely.

It is evident from the laboratory investigations that the patient has amyotrophic lateral sclerosis (ALS) with frontal lobe characteristics and has diabetes and hypertension as comorbidities. The patient has been provided with a tab. Riluzole is the prototype drug to treat ALS, although it is not a cure for the condition, it may extend the survival of the patient. The patient had been informed of the possibility of injecting Edavarone, which may or may not help him with his condition. By the end of the course, the patient's stiffness had decreased, his gait had improved, his walking speed had increased, and his dysarthria had marginally improved. Currently, he is taking tabs Metformin, Atocor, and Ecospirin.

When the patient's blood pressure and blood sugar levels were checked, they were found to be normal. He needs routine follow-up after being discharged.

CONCLUSION

Lou Gehrig's disease, also known as amyotrophic lateral sclerosis (ALS), is a progressive neurological condition that damages motor neurons in the brain and spinal cord. This illness affects voluntary motions including breathing, walking, and speaking by causing muscle weakening, twitching, and ultimately paralysis. Over time, ALS can cause severe impairment by affecting the muscles involved in breathing and swallowing.

Electromyography (EMG), nerve conduction investigations, imaging, and clinical assessment are all used in a complete evaluation to rule out other illnesses before making the diagnosis of ALS. Although ALS has no known cure, treatment aims to control symptoms and maybe reduce the disease's development. Certain medications, such as Edaravone and Riluzole, are used to extend survival and partially relieve symptoms.

To better understand ALS, find appropriate treatments, and enhance the quality of life for those who suffer from this difficult disease, ongoing research is essential. Though there are now few alternatives for therapy, the medical community is still working to make improvements that will perhaps lead to improved outcomes and a cure in the future.

REFERENCES

1. National Institute of Neurological Disorders and Stroke. Amyotrophic lateral sclerosis (ALS). www.ninds.nih.gov. 2023.
2. Cleveland Clinic. Amyotrophic Lateral Sclerosis (ALS). Cleveland Clinic. 2021
3. Marcus R. What Is Amyotrophic Lateral Sclerosis? JAMA. 2022 Dec 27;328(24):2466.
4. Falchi D. Frontal lobe syndrome: A mini-review. Zenodo (CERN European Organization for Nuclear Research). 2022 Oct 17;
5. Usman U, Choi C, Camicioli R, Seres P, Lynch M, Sekhon R, Johnston W, Kalra S. Mesial prefrontal cortex degeneration in amyotrophic lateral sclerosis: a high-field proton MR spectroscopy study. AJNR Am J Neuroradiol. 2011 Oct;32(9):1677-80.