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Case Report on Amyotrophic Lateral Sclerosis

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ABSTRACT:

Introduction: Lower and higher motor neurons in the brain and spinal cord deteriorate as a result of the neurodegenerative disease amyotrophic lateral sclerosis (ALS), which progresses. Early-onset and present treatments, such as respiratory support, increase survival in this illness, yet its origin is still unknown.

Symptoms And Important Clinical Findings: The patient was a 68-year-old male with a chief complaint of difficulty speaking, laryngological dysfunction, Inappropriate crying, difficulty in walking, and falling for 1 month.

The Main Diagnosis, Therapeutic Intervention, And Outcome: Due to the decline of the upper motor and motor neurons, the condition produces muscle weakness, difficulty in speaking, and spasms all over the body. Individuals with the disorder may eventually get rid of capable of All voluntary movement must be initiated and controlled. Still, bladder and bowel function, as well as Muscles outside of the (ocular muscles that allow you to move your eyes) are typically spared until the later stages of the illness.

Therapeutic intervention: Breathing Exercise: To improve the patient breathing pattern and help the Patient breathe easier.Physical therapy: The physiotherapist suggests the patient low-impact exercises which will help to patient cardiovascular fitness. This therapy helps the patient in walking. The doctor recommends the use of a walker or wheelchair.

Outcome: Amyotrophic lateral sclerosis is a neurological illness that worsens with time. Due to the failure of the ventilator muscles, motor neuron loss produces weakness, disability, and eventually death in ALS. The average survival time is 3-5 years, with an average beginning age of 55years.

Conclusion:A patient with a rapidly progressing, unexplained neuropathic condition should be evaluated for amyotrophic lateral sclerosis. This should be the case even if there are clinical and electrodiagnosticfindings.The development of upper motor neuron symptoms and a lack of response to therapy should confirm the presence of amyotrophic lateral sclerosis.

Keywords: difficulty walking, muscle wasting, legs, shoulders, or tongue, contraction of muscle, tripping or falling, weakness in the hands, or clumsiness.

INTRODUCTION:

With the progressive increase in the brain and spinal cord, motor neurons are damaged as a result of amyotrophic lateral sclerosis. The pathobiology of this neurodegenerative condition is comparable to front temporal dementia, and many people experience symptoms similar to birth abnormalities. The disease is caused by a wide range of genes and pathophysiological pathways, and understanding this heterogeneity will be necessary for developing successful treatments.Scientific advancements in the fields of genetics, disease modeling, biomarkers, and therapeutic alternatives, as well as clinical and diagnostic approaches[1-2]Amyotrophic lateral sclerosis (ALS), is a rapidly progressive nervous system illness in which the upper and lower neurons of the brain and spinal cord deteriorate. Although the causes are unknown, early discovery of the condition and modern treatments, such as ventilation, improve the Patient's longevity. Nerve factor insufficiency, glutamate shortage, industrial pollution, and occupational or chemical exposure are all potential causes.[3]

Amyotrophic lateral sclerosis [ALS] is a neurological disease in which the brain and spinal cord's lower and higher motor neurons deteriorate. Even though the cause of this condition is unknown, idiopathic and contemporary treatments, such as asses ventilation, have been shown to improve survival.[4]

Nerve growth factor deficiency, glutamate deficiency autoimmunity, and mutations in the superoxide dismutase 1 gene are all possibilities. Occupational exposure to chemicals and industrial pollution are linked. Nerve growth factor deficiency, autoimmunity due to glutamate deficiency, and mutations in the superoxide dismutase gene are all possible causes. Chemical exposure from welding and soldering could be issued. The proposed ALS diagnosis criteria, as well as a response, are presented by the World Federation of Neurology. look at the genes most closely associated with amyotrophic lateral sclerosis in this chapter great detail regarding the illness's clinical symptoms other than to say that it's a paralytic illness that starts with localized motor weakness in the distal extremities but can also affect the bulbar musculature and escalates to total body paralysiswill die of ventilatory failure in around five years if you do not obtain aid with your breathing. These characteristics have been connected to frontotemporal dementia (FTD), which is characterized by behavioral and verbal difficulties at first. The important microscopic discovery is motor neuron death, frequently accompanied by proteinaceous aggregate accumulation in neurons and no neuronal cells. Motor neuron death, typically accompanied by proteinaceous aggregates and, in certain circumstances, intranuclear RNA deposits in neurons and no neuronal cells, is the most common observation at the microscopic level. In the majority of instances, autopsy reveals atrocity and microgliosis, as well as motor neuron loss. Both lower motor neurons (LMNs) and upper motor neurons (UMNs) are affected.

Dominant features are inherited in about 10% of cases (familial ALS; ALS). In the early to mid-1980s, scientists began earnestly seeking ALS genes. It was believed that researchingpathways implicated by mutant ALS genes would provide insight into the disease's etiology if the basic pathology of ALS was unknown. In around 10% of cases, dominant characteristics are inherited (familial ALS; ALS). Researchers began looking for ALEarly to mid-1980s saw the beginning of the study of ALS genes. genes in earnest in the early to mid-1980s. If the primary pathology of ALS was unknown, it was hoped that investigating pathways implicated by mutant ALS genes might give information about the disease's aetiology.[5]It's unexpected that gene-finding technology has advanced considerably in the last 30 years. Genetic linkage approaches were used in the first and most fruitful period of ALS gene identification. common species co-migrating variations with the condition could be used in pedigrees to infer the genes' broad chromosomal addresses. When linkage was established, this strategy necessitated both a comprehensive multigenerational family structure with discrete inheritance patterns and a significant amount of work to find the causative mutation within the linked locus; often, this necessitated painstaking hand sequencing of dozens of candidate genes. The second phase focused on genome-wide association studies (GWASs), which provided at least two significant benefits: the ability to search for genetic variants linked to both sporadic and familial ALS, as well as the utilization of hundreds of thousands of single nucleotide polymorphisms across the genome. Although this strategy has been used to identify multiple candidate genes, it is less reliable than linkage studies. Next-generation high-throughput In the third and current phase of ALS genetic research, sequencing has been combined with genomic capture to significantly improve the finding of ALS-causing gene variations. ALS genetics will define critical molecular processes in ALS pathogenesis was extensively established thirty years ago, even though no viable ALS remedy exists.[6]

Patient-Specific Information:-

The patient was a 68-year-old male with chief complaints of difficulty speaking, laryngological dysfunction, inappropriate crying, and difficulty walking and falling.

Medical History:-

The patient has difficulty speaking, laryngological dysfunction, difficulty in walking, No any history of diabetes, Mellitus, tuberculosis, and asthma

Family history:-

He belonged to a joined family with five family members.

Psycho-social History: -He was mentally healthy, conscious, and oriented to date, time and place. He had maintained a good relationship with doctors, nurses, and other patients.

Relevant past intervention with outcomes:-

The present case had no history of a similar attack and no history of hypertension, DM, heart disease, tuberculosis, or asthma.

Clinical Findings :

The Patient complained of dysarthria gradually worsening; a month later, he experienced generalized fasciculations, and his speech became slurred. He was admitted to the ENT department after laryngological dysfunction was ruled out.

Physical Examination:-

The patient's complaint is difficulty in speaking, laryngological dysfunction, and difficulty in walking.

Temperature:-97°/

Pulse:-82b/m
Blood pressure:120/80mmhg
Respiration:22

Neurological examination:

In Neurological Examination, the Patient has a low psychological function. Emotional incontinence and a smooth, glossy tongue, as well as a speech fault, are all symptoms of emotional incontinence. Muscle degeneration or weakening was detected, as well as generalized fasciculations. It is normal to have an impact, feeling, and coordination.

Important Clinical Findings :

DIAGNOSTIC ASSESSMENT:

The patient biochemistry test is negative. Serum, proteins, normal immunoglobulin levels, and creatinine antibodies are negative. Due to the Patient's unwillingness, cerebrospinal fluid analysis was not done.

Electromyography was done in the right bicep branchi. Computerised tomography was also done, and MRI report showed a Bone abnormality in the interface between the skull and cervical spine.

PROGNOSIS: Amyotrophic lateral sclerosis is a disease that progresses slowly.50% of Patients survive for fewer than three years after being diagnosed. And only 20%patient can survive for 5 to 10 years. In Amyotrophic lateral sclerosis, Patients develop respiratory weakness, and most Patients can die from pulmonary complications.

Therapeutic interventions:

There is no cure for motor neuron injury or medication that can reverse it. However, treatment helps control symptoms, prevent unnecessary complications, and make living with the sickness easier.

Nursing perspectives:- Administration of glutamate blocker and muscle relaxant monitor vital signs per hourly, palliative care.

Follow-up and outcomes:

The Amyotrophic lateral sclerosis patient required one year of follow-up after diagnosis of Amyotrophic lateral sclerosis. For 1 -2 years, he was advised totake medication as per doctor's order and physical therapy.

OUTCOMES: Amyotrophic lateral sclerosis [ALS] is a neurodegenerative illness that progresses over time. Because the ventilatory muscles fail in Amyotrophic lateral sclerosis, it produces weakness, incapacity, and death. The usual survival time is 3-5 years, and the average onset age is 55 years.

DISCUSSION:

A patient with bulbar-onset Amyotrophic Lateral Sclerosisand Klippel –Fiel syndrome, as well as asymptomatic cervical development of fluid-filled cyst is presented. We ruled out cervical syringomyelia as the cause of our Patient's symptoms no sensory deficits or muscle weaknesswhich was followed by clinical signs of lower motor neuron impairment in the cervical areas. Amyotrophic lateral sclerosis (ALS) is a rapidly progressive neurological disease in which motor neurons in the brain and spinal cord are destroyed, eventually leading to respiratory failure and death.[7-14] Adenine nucleotide translocation protein 1 (ANT1), the most abundant protein in the inner mitochondrial membrane, is expressed primarily in the heart, brain, and skeletal muscles and may play a protective role in mitochondrial dysfunction. Adenine nucleotide translocator protein 1 (ANT1), the most important protein in the inner mitochondrial membrane may play a protective role in mitochondrial dysfunction.Amyotrophic lateral sclerosis-like disease since the cases were occasionally atypical, and this is part of different neuromuscular failures. The condition insidiously makes proving causality with a treatment challenge. The clinical course of the two concurrent neurological illnesses in our example recalls previous studies by Dynes et al. and Li et al.. They established the neuropathological coexistence and simultaneous clinical progression of ALS and multiple sclerosis respectively. Degeneration of pyramidal tracts and anterior horns cells in both the cervical and lumbar cords, as well as many demyelinating plaques in prototypic sites, have all been found as pathogenic features (i.e., cortex, periventricular region, corpus callosum, brainstem, and spinal cord). clinical data support the overlap of neurological illnesses in our Patients, the absence of autoptic confirmation of ALS and MS coexistence in our Patients is a severe drawback of our report. [15-24]

Lower motor neuron dysfunction has been reported in MS patients on occasion, notably in patients with difficulty with hand motor activities, and has been linked to MRI detection. According to our findings, there were no signs of demyelinating plaques in the spinal cord. Our longitudinal MRI investigation was limited in duration due to the onset of respiratory failure, making follow-up MRI scans difficult. We believe there are some common pathogenic pathways of inflammation and degradation in both ALS and MS. However, we do not think there is a relationship between the two diseases.[25-31]

CONCLUSION:

This study covers various facts about ALS, including its molecular causes, epidemiology, and treatments. Unfortunately, ALS is thought to be an incurable condition with a life expectancy of three to five years following the onset of symptoms. Although numerous antioxidants and supplements have been suggested as an alternative to traditional ALS therapies, the majority of these have either not been validated in research studies,

or the studies that have been conducted lack validity or considerable proof in their technique. To better care for ALS patients, it is essential to continue nutritional research since, according to some evidence, it may assist to lessen the disease's effects on patients' everyday lives. As an illustration, a thorough and cohesive study on alpha-tocopherol and creatine is.

The understanding of ALS pathogenesis has undergone significant breakthroughs. Nineteen genes and genetic loci have been found that are associated with ALS. Understanding the genetic mechanisms behind ALS will help determine the best treatment course. Antiapoptotic drugs, anti-aggregation, antioxidant, anti-excitotoxic, anti-inflammatory, neuroprotective, and neural growth factors are currently the subject of several clinical trials. Recent understandings of ALS's underlying process have assisted in slowing the disease's progression. Thus, as patients advance beyond their initial onset, future treatments should focus on preventing neuronal damage.

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