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A Case Report of Rapidly Progressing Amyotrophic Lateral Sclerosis in 31 Year Old Patient with Sympathovagal Imbalance

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Abstract

ALS is a fatal neurodegenerative condition characterized by degeneration of upper and lower motor neuron in cerebral cortex, cranial nerve nuclei and anterior horn cells of spinal cord. Three drugs are FDA approved, Riluzole, Edaravone and AMX0035 (recently approved), which can only prolong survival time by 2-3 months. Hence, the basic management is supportive and palliative. Here we discuss case report of 31 year old Afro-American male who noted subtle weakness and reduced grip strength in July 2022. He had difficulty focusing his eyes specially when changing depth. Electromyography revealed intraspinous sub acute denervation involving motor neurons at the cervical, thoracic or lumbosacral levels. He became non ambulatory after April 2023.Cardiac issues faced may be secondary to Sympathovagal imbalance. Currently he is on ventilator due to respiratory failure. A very rare sight is presentation of ALS at the age of 31 years.

Keywords: Riluzole; Sympathovagal imbalance; amyotrophic lateral sclerosis

Introduction

Amyotrophic Lateral Sclerosis also known as Motor neuron is characterized by muscle weakness, muscle function decline and eventually paralysis. This happens due to irreversible degeneration of upper and lower motor neurons in the cerebral cortex, spinal cord and cranial nerve nuclei. The disease may have bulbar onset or limb Onset and can be familial or sporadic. Common presenting symptoms are limb weakness, muscle twitching, stiffness and cramps, dysphagia, dysarthria and impaired balance [1]. A number of criteria are known to diagnose ALS , Gold Coast, revised El Escorial and Awaji , with Gold Coast having 92% diagnostic sensitivity as compared to 88.6% of revised El Escorial criteria [2]. In regards of prognosis, For determining the rate of progression, ALSFRS-R scoring system is used widely [3]. Average life expectancy is 2-5 years. With respect to drug treatment 3 drugs are FDA approved, Riluzole, Edaravone and AMX0035 (recently approved) [4], which only can prolong life survival time by 2-3 months. Thus, the basic management is supportive and palliative. Neurology team remains the primary team for ALS patients when diagnosing, with other departments working as a unit alongside. Pulmonology Department is crucial because mostly the cause of death in the patients is respiratory failure. In this specific case, patient was on ventilator due to respiratory failure and had pulmonary embolism too thus needing a pulmonology consult. Lastly the palliative care department is pivotal for these patients with fatal diseases. Here we describe a case of ALS with sympathy vagal imbalance who deteriorated progressively within months.

Case Presentation

This is a case report of 31 year old Afro-American male who first noted subtle weakness in July 2022 along with reduced grip strength bilaterally. Within a couple of months he noted increasing generalized weakness. The weakness was initially confined to upper extremities however later involved lower as well. The weakness is symmetrical with no clear proximal or distal pattern. In oct 2022 he had multiple falls due to weakness and balance problems. Weakness persists throughout the day and is not associated with activities, Changes in speech like slowed and slurred speech were noted in sept 2022. He had difficulty focusing his eyes specially when changing depth. Dysphagia more for liquids than solids, occasional drooling, trouble clearing his throat and intermittent dyspnea with activity but no double vision. He also reported having muscle twitches in arm and chest muscle and right lower extremity in May 2022.

Before all the symptoms appeared he gave no history of infections, vaccinations or illnesses. He denies any sort of unexplained fever, chills, rashes, joint pain swelling, rashes, ulcers and Raynaud phenomenon. Approximately lost 30 lbs. of weight in year 2022.

He had MRI Brain and Cervical spine with and without contrast in November 2022 which was unrevealing. Next he had Electromyography done on 23rd November 2022 which showed intraspinous subacute denervation involving motor neurons or axons at the cervical, thoracic or lumbosacral levels. All the muscles tested showed evidence of abnormal insertional activity in the form of positive waves. His Lab workup revealed abnormal values of

Creatinine kinase 632 Units/L, Aspartate aminotransferase 62 Units/L, Alanine transaminase 56 Units/L

White Blood Cell 2.76 x 109 /L, Absolute Neutrophil Count 1.0 x109 /L

While normal or negative status for HIV, ANA, RF, SSA, SSB, SPEP, B12, UDs, TSH, and COVID

HBA1c 5.7

He again presented to Hospital on 3rd February 2023 due to increased falls, episodes of choking on thin liquids, shortness of breath when in supine position and inappropriate smiling and laughing. The patient was alert and awake with spastic dysarthria and weak cough. He had atrophy of upper back muscles, fasciculations on arms bilaterally and on tongue, lateral tongue atrophy. Atrophy of first dorsal interosseous, fingers held in flexion position bilaterally and unable to extend even with gravity eliminated, but was able to hold head against gravity.

For ruling out mimic's serum autoimmune panel, CSF autoimmune panel, CSF WNV IgM, serum heavy metals and other serum MND mimics were ordered. A repeat EMG and CT brain and cervical spine was also planned. For CSF albuminocytologic dissociation empiric IVIG/Steroids were considered. The workup was high concerning for ALS meeting clinically definite El escolar criteria. The workup and labs were unrevealing except for the elevated CSF protein (96) and weakly positive ANA -1:160. A 3 day course of high dose [4] steroids was given with some subjective improvement but not objective. Planned for follow up on 20th march 2023.

He presented again on 19 February for potential IVIG, had blurred vision, symptoms unresolved, no increasing weakness than baseline, no diplopia, no dysphagia, no dysphonia, no bowel bladder incontinence or sensory disturbances.

He was diagnosed ALS in February 2023

He was seen by ALS clinic in March 2023 and was initiated on riluzole and was provided cough assist and hoyerlift, hospital bed and other resources. He was admitted again in May 2023 due to shortness of breath which was found to be due to acute pulmonary embolism in right upper lobe. He was established with hospice and palliative care and lastly visited them on June 9, 2023. In April he was still able to move but after that he was unable to ambulate. He could communicate only by mouthing yes and no and shaking his head only.

In July 2023, He presented with cardiac arrest. he was having episodes of shortness of breath and was intubated in emergency and he subsequently developed pulseless ventricular tachycardia which converted to ventricular fibrillation , defibrillation was done twice , 9 doses of epinephrine followed by ROSC .cause of arrest was thought to be secondary to succinylcholine administration in ALS patient as well as hypoxemia acute on chronic respiratory failure. He is bedbound, nonverbal, communicates through blinking eyes and shaking head.

Currently this ALS patient is in acute respiratory failure on ventilator, with left sided pneumonia, DVT, PE, urinary retention and a recent Methicillin sensitive staphylococcus aureus pneumonia.

Current medications:

Riluzole, Apixaban, Guaifenesin, Lorazepam, Losartan, Tamsulosin

Baclofen, piperacillin tazobactam

Almost 10% of people diagnosed with ALS has a family history of disease (Familial ALS) with an autosomal dominant pattern mostly , while 90-95% of patients have no family history thus Sporadic ALS. More than 50 genes have been identified that are linked with ALS while only pathogenic variants of 4 of them are usually identified commonly to cause disease SOD1, C9ORF72, -FUS, and TARDBP in victims of ALS [5]. Despite all this research the exact genetics remains unexplained.

Discussion

ALS is a fatal neurodegenerative condition characterized by degeneration of upper and lower motor neuron in cerebral cortex, cranial nerve nuclei and anterior horn cells of spinal cord. Around the world, the mean age for initiation of symptoms for sporadic ALS is 58-63 years while it is 40-60 years for Familial ALS [6]. In this case report surprisingly the patient reported ALS symptoms at just an age of 30 which is an uncommon presentation.

Another striking feature of ALS seen in some patients is the autonomic dysfunction making ALS a multisystem involvement disease [7]. In this specific case we noticed symptoms of sympathovagal imbalance (tachycardia, Blood pressure variation, sweating etc), which may also be the cause of cardiac problems arising in the patient.

Otherwise cardiac issues are underappreciated in these patients. Tachycardia, changes in blood pressure, sweating, dysphagia, urinary retention, blurred vision and balance problems all are indicative of dysautonomia. Sympathetic hyperactivity and Sympathovagal imbalance are striking features of Autonomic dysfunction in ALS [8].

Riluzole is the only drug approved for the treatment of ALS. The rate at which muscular function declines is decreased by riluzole [9]. Our patient received riluzole approximately 9 months after the symptom onset but no significant improvement was noted.

There are various ALS staging systems available worldwide simplest staging the disease into early, middle and late stages [10]. The patient is currently in late stage of ALS and had rapid progression from middle stage to this late stage approximately 3 months which is a bit fast. It took him a year to develop late stage (paralysis of voluntary muscles, respiratory complications requiring ventilator support, hospice care dependent etc) from the first appearance of symptoms.Whether psychosocial therapies enhance the quality of life and general wellbeing of persons with neuromuscular illnesses is currently unknown with confidence. Even while a number of psychological therapies have shown modest benefits for both quality of life and wellbeing, these advantages are largely transient and biased [11].

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