

# Recurrent Pneumonia and Progressive Respiratory Failure as Initial Manifestations of Amyotrophic Lateral Sclerosis: A Case Report

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## Abstract

Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disease that presents diagnostic challenges, particularly in patients with overlapping chronic conditions. Presented is the case of a 71-year-old male with severe COPD who initially presented with respiratory distress but was later found to have progressive asymmetric motor weakness, muscle atrophy, and upper motor neuron signs suggestive of ALS. After an abnormal EMG, the clinical picture supported a motor neuron disease diagnosis. The patient elected to forgo further interventions in favor of comfort-focused care, ultimately requiring palliative intubation for symptom relief. He passed away peacefully after an 18-day hospital course and contributed to organ donation through donation after circulatory death (DCD). This case underscores the importance of early recognition of ALS, the role of multidisciplinary care, and the value of shared decision-making in managing terminal neurodegenerative disease.

**Keywords:** Amyotrophic Lateral Sclerosis (ALS), Chronic Obstructive Pulmonary Disease (COPD), Recurrent Pneumonia, Progressive Respiratory Failure.

## Introduction

Amyotrophic Lateral Sclerosis is a fatal neurodegenerative disease characterized by progressive upper motor neuron and lower motor neuron damage. It has a silent manifestation that usually begins asymmetric weakness in the extremities, and as the disease progresses most patients develop dysphagia and respiratory system failure. The definitive cause of ALS is idiopathic, but studies have suggested there is an interaction between genetic predisposition and environmental factors. Specifically, mutations in Superoxide Dismutase 1 account for 15-20% of all familial ALS cases. Environmental exposures to pesticides, lead, and cyanobacteria have also been associated with many neuromuscular degenerative diseases including ALS, parkinsonism, and dementia. Potential hypothesized pathophysiological cascades involve abnormal RNA processing and protein aggregation, mitochondrial dysfunction, and defective neurofilaments. There are a few FDA approved medications, including Riluzole and Edaravone, which prolong life expectancy and slow disease progression. However, the mainstay of treatment involved supportive care, and prevention of complications.

## Case Presentation

### Patient History and Presentation

A 71-year-old male patient with a significant past medical history of severe COPD, chronic respiratory failure, hypertension, sleep apnea, urinary retention, constipation, and nerve damage presented to the ER for evaluation of respiratory distress and shortness of breath that started a few hours prior to arrival. He was discharged home from another local hospital earlier the same day, and taken to a long-term rehab facility where he developed pulmonary symptoms. He denies any fever, chills, but reports wheezing, cough, mild non radiating chest tightness, without any aggravating or alleviating factors.

### Vital Signs on Presentation

- BP: 153/70
- HR: 117
- Temp: 98.8°F (37.1 °C) Oral
- Respiratory Rate: 30
- SpO<sub>2</sub>: 99% on BIPAP 15/5 100% FiO<sub>2</sub>

## Physical Examination

**Constitutional:** He is in moderate respiratory distress requiring bipap

**HENT:** Normocephalic and atraumatic, mucus membranes moist.

**Cardiovascular:** Normal rate and rhythm, normal heart sounds, no murmur/gallop.

**Pulmonary:** Moderate respiratory distress present with extra-accessory muscle usage, requiring bipap. No stridor. Decreased breath sounds bilaterally and wheezing bilaterally. No rhonchi or rales. RR 30s.

**Abdomen:** Soft, with no tenderness, guarding, or rebound tenderness.

**Musculoskeletal:** Extremities nontender with no decreased range of motion, joint swelling, or obvious deformity

**Neurological:** Alert and oriented to person, place and time. Sensation and motor function is intact. No weakness. The patient is able to follow commands.

**Skin:** warm and dry, no rashes

## Laboratory Results

Hemoglobin: 11.4

Hematocrit: 35.9

WBC: 26.1

BNP: 269.6

Lactate: 2.2

Hemoglobin A1C: 5.9

Vitamin B12: 724

Folate: 5.1

TSH: 0.96

Sed Rate: 3

CRP: 0.979

Arterial Blood Gas:

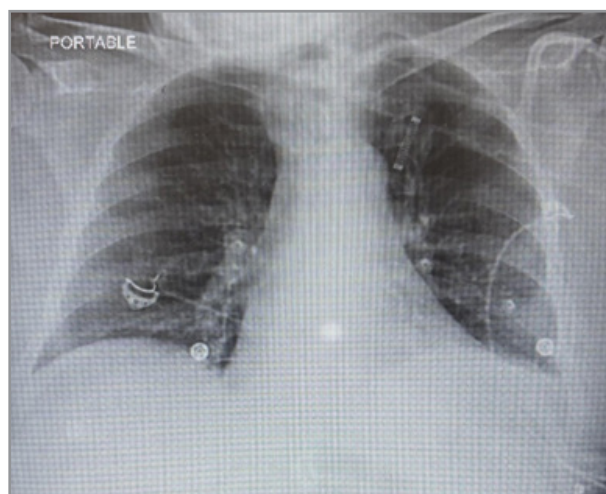
pH: 7.43

PCO<sub>2</sub>: 44

PO<sub>2</sub>: 477

HCO<sub>3</sub>: 29.2

BE 4.3



**Figure 1:** Chest X-Ray (ED): No pneumothorax or pleural effusion. No consolidation. No acute chest process

## Management and Outcome

This was a 71-year-old male patient with a history of severe chronic obstructive pulmonary disease (COPD) on baseline BIPAP presented initially to an outside hospital (OSH) with acute COPD exacerbation. He had recently been discharged earlier the same day from another local hospital to a rehabilitation facility but experienced rapid onset of shortness of breath and was subsequently admitted to our hospital, where he was found to have methicillin-resistant *Staphylococcus aureus* (MRSA) pneumonia. He was treated with vancomycin and maintained on BIPAP, but due to ongoing diffuse weakness and inability to titrate down his oxygen demands, he was transferred to an academic hospital for further workup and neurological evaluation. The patient had a six-year history of progressive dyspnea, previously attributed to worsening COPD, however over the preceding six months he had developed asymmetric motor deficits, including right lower extremity weakness consistent with foot drop, left hand "curling," and left upper extremity weakness. He also reported severe constipation during this time. On examination, there was marked diffuse muscle atrophy involving the thenar and hypothenar muscles, thighs, and calves, with hyperreflexia and clonus noted in the left foot, and inconsistent but concerning findings for upper motor neuron signs. Cranial nerves were intact, and no acute structural abnormalities were observed on MRI of the

brain or spine.

The clinical presentation—characterized by progressive, asymmetric upper and lower motor neuron involvement in the absence of structural lesions—was most consistent with a neurodegenerative process such as amyotrophic lateral sclerosis (ALS) or another neuromuscular disorder (NMD). His EMG results suggested either polyradiculopathy vs motor neuron disease, and all MRI results were unremarkable, pointing to a final diagnosis of ALS. The patient declined further diagnostic workup including, imaging, or laboratory testing, preferring to focus on comfort after a thorough discussion regarding prognosis and lack of curative or disease-modifying treatments for suspected ALS. His management included continuous BIPAP for respiratory support, prophylactic doxycycline for infection prevention, enoxaparin for DVT prophylaxis, and anxiolytics (Xanax and Valium) for anxiety related to respiratory distress. Due to discomfort from prolonged BIPAP use and significant respiratory fatigue, the patient was palliatively intubated to facilitate comfort and to allow time for family to arrive from out of town. He was maintained on peripheral parenteral nutrition (PPN), vasopressors, fentanyl, and Valium for sedation and symptom control.

The patient remained hospitalized for 18 days and ultimately

passed away peacefully after transitioning to comfort-focused care. Prior to death, he had elected to donate his organs after circulatory death (DCD), and organ procurement was successfully performed. This case highlights the clinical complexity of differentiating neurodegenerative motor neuron disease in the setting of chronic respiratory illness and demonstrates the importance of patient-centered care planning and end-of-life decision-making in advanced, incurable neurological disease.

## Discussion

An important distinction in this case was the difficulty in distinguishing this patient with a history of chronic respiratory disease from the emerging features of ALS, considering longstanding COPD, baseline respiratory failure, and limited physiologic reserve. Dyspnea, fatigue, and weight loss, which are all common symptoms in advanced COPD, can mask or mimic the early systemic effects of ALS, potentially delaying diagnosis and appropriate care planning. Moreover, existing co-morbidities may impinge and prevent patients from aggressive diagnostic or therapeutic interventions, especially when symptoms evolve insidiously or overlap.

Once ALS became the working diagnosis, the patient made an informed decision to prioritize comfort over further testing or disease-modifying attempts. This decision reflected the patient's understanding of ALS prognosis and the lack of curative therapies, as well as an awareness of the burdens associated with ongoing care in terminal disease. He declined invasive testing, non-essential medications, procedures, and transitioned to palliative-focused management. Despite the use of continuous BIPAP, his symptoms progressed, and the discomfort from noninvasive ventilation became unbearable. Towards the end, he decided to be palliatively intubated to relieve suffering and allow his family to travel and be present during his final days.

During his hospital course, symptom control was managed with vasopressors, peripheral parenteral nutrition (PPN), fentanyl, and benzodiazepines to ensure comfort and dignity at the end of life. After 18 days of hospitalization, he passed away peacefully, having made prior arrangements for donation after circulatory death (DCD). His organ procurement continued to contribute to the lives of others after his death.

This case emphasizes the critical importance of timely recog-

nition of neurodegenerative diseases in patients with complex medical histories and highlights the need for continual collaboration between pulmonology, neurology, and palliative care. Although therapeutic options for ALS remain limited, supportive care remains the cornerstone of management and can help patients in a different way. Nutritional support, respiratory assistance, physical therapy, and psychological support can significantly improve quality of life, even if not altering the disease course.

Clearly, this case highlights the ethical and clinical importance of shared decision-making in the context of a progressive, incurable illness. By focusing care on the patient's values, preferences, and goals, clinicians can provide a framework for meaningful engagement, even in the absence of curative options. The patient's decision to forego burdensome interventions in favor of comfort and organ donation reflects the profound influence individuals can maintain on their decisions, even in the face of terminal illness.

## Conclusion

This case highlights the diagnostic and management challenges of amyotrophic lateral sclerosis in the context of complex comorbidities such as advanced COPD. The patient's clinical course demonstrated the subtle progression and diagnostic ambiguity of ALS, particularly when overlapping with chronic respiratory disease. Ultimately, his decision to pursue comfort-focused care reflects the importance of patient-centered approaches, emphasizing autonomy, symptom relief, and dignity at the end of life. Though therapeutic options for ALS remain limited, timely recognition, interdisciplinary coordination, and compassionate communication remain essential in guiding care and supporting patients and families through this terminal illness.

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