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RECURRENT SPONTANEOUS PNEUMOTHORAX IN THE SETTING OF AMYOTROPHIC LATERAL SCLEROSIS

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ABSTRACT

Spontaneous pneumothorax (SP) occurrence in amyotrophic lateral sclerosis (ALS) patients is relatively rare and may thus be under-recognised. This latter is a progressive neurodegenerative disorder, leading to muscle weakness and such respiratory complications. This article reports a case manifesting such a rare association.

KEYWORDS

Spontaneous pneumothorax, amyotrophic lateral sclerosis

LEARNING POINTS

- The presence of spontaneous pneumothorax in amyotrophic lateral sclerosis patients can exacerbate respiratory insufficiency, leading to acute respiratory failure.
- Given the under-recognition of this latter complication, clinicians should maintain a high index of suspicion, especially in patients with amyotrophic lateral sclerosis presenting with sudden onset of dyspnoea or chest pain.
- Early detection and appropriate management are crucial to prevent further respiratory compromise.

INTRODUCTION

Spontaneous pneumothorax (SP), characterised by the accumulation of air in the pleural space without preceding trauma, can be classified into primary (PSP) and secondary (SSP) types. PSP occurs in individuals without known lung disease, while SSP arises in patients with underlying pulmonary conditions. Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disorder marked by motor neuron degeneration, leading to muscle weakness and respiratory complications. The occurrence of SP in ALS patients is relatively uncommon and may be under-recognised[1,2]; we report a case manifesting such a rare association.

CASE DESCRIPTION

The patient, a 66-year-old male non-smoker who is non-allergic, presented to emergency room (ER) with an altered level of consciousness. Past medical history was positive for ALS chronically maintained on riluzole, complicated by recurrent hospital admissions for aspiration pneumonia.

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No past surgical history was noted. On arrival, vitals were pertinent for desaturation reaching 60%, tachycardia (115 beats per minute) and blood pressure of 80/50. Initial workup in the ER was pertinent for increased inflammatory markers and right lower lobe consolidation on chest CT along with type 3 respiratory failure, thus requiring immediate endotracheal intubation. The patient was admitted to the intensive care unit with the working diagnosis of respiratory failure due to aspiration pneumonia and was started on antibiotics (piperacillin/tazobactam).

On day four of admission, results of deep tracheal aspirate culture showed positive for *Candida albicans*, and blood culture turned negative. Inflammatory markers started to decrease along with the patient's oxygen requirements; the process of weaning started. The patient's level of consciousness off sedation was appropriate but he still failed weaning from the respirator due to low tidal volume and high respiratory rate. This process was repeated once daily for three consecutive days without success. During this time the patient received extra doses of diuretics and methylprednisolone without benefit; therefore, the decision was taken to keep him awake, on assist control mode of ventilation to discuss the decision to go for a tracheostomy with the family. The patient was fully synchronised and calm with the respirator, without the need for any sedation agents.

On day eight, patient developed sudden onset of hypotension, tachycardia and tachypnoea without significant desaturation. A chest X-ray was inconclusive; an urgent chest CT showed right significant pneumothorax and new left lower lobe consolidation *(Fig. 1)*. A chest tube was inserted and the patient started on meropenem and tigecycline for ventilator-associated pneumonia.

On day 14, patient's clinical and haemodynamic status started to improve; he was scheduled to have a tracheostomy insertion to prepare for home discharge, and a tracheostomy was performed without complications. Two hours post procedure, patient started to deteriorate with desaturation and hypotension. Bedside chest X-ray showed left upper pneumothorax *(Fig. 2)*. A chest tube was inserted on the left side. During the rest of his hospitalisation, the patient completed the course of antibiotics, both chest tubes were removed, and he was discharged home on bilevel positive airway pressure alternating with collar mask oxygen without any further complications.

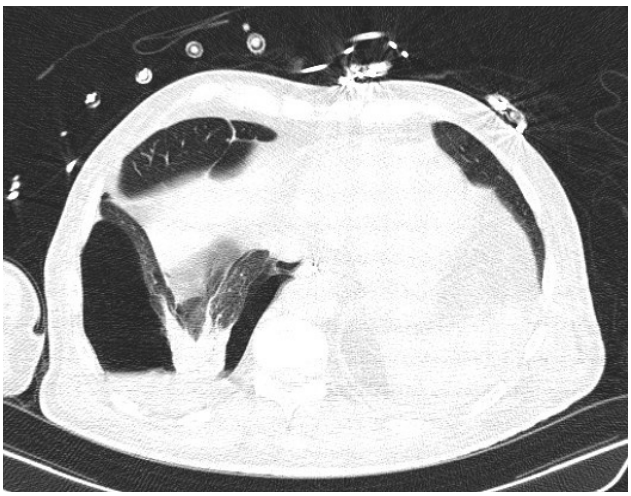


Figure 1. CT scan of the chest showing a moderate size right basal pneumothorax.

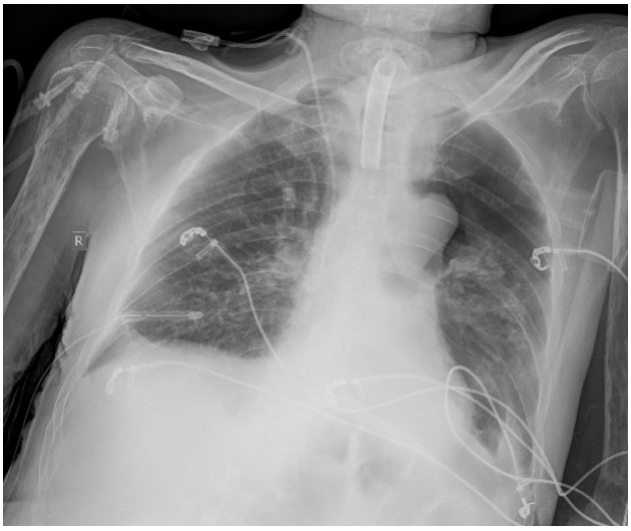


Figure 2. Chest X-ray showing left-sided pneumothorax.

DISCUSSION

The occurrence of SP in ALS patients is relatively uncommon and may be under-recognised. A multicentre retrospective study involving 86 ALS patients found that 7% (six patients) experienced PSP. Notably, low body mass index (BMI) and a history of smoking were significant risk factors associated with PSP in these patients. The study reported that ALS patients who smoked and had a low BMI were at a higher risk for developing PSP, with a combined odds ratio of 19.25[1].

The exact mechanism linking ALS to SP is not fully understood. However, several factors may contribute to this:

- *Respiratory muscle weakness:* ALS leads to progressive weakness of respiratory muscles, including the diaphragm and intercostal muscles, resulting in reduced lung volumes and atelectasis. This can increase the risk of alveolar rupture and subsequent pneumothorax[3].
- *Mechanical ventilation:* ALS patients with respiratory failure often require mechanical ventilation. Positive pressure ventilation can predispose to barotrauma, leading to pneumothorax. However, the aforementioned study excluded patients with a history of central venous catheter use and those with insufficient medical records regarding ventilator use, to eliminate iatrogenic causes[1-4].
- *Nutritional status:* Malnutrition and low BMI are prevalent in ALS due to dysphagia and increased metabolic demands. Low BMI has been identified as a risk factor for PSP, possibly due to decreased subcutaneous fat and muscle mass, leading to less support for the pulmonary structures[5].

The presence of SP in ALS patients can exacerbate respiratory insufficiency, leading to acute respiratory failure. Given the under-recognition of this complication, clinicians should maintain a high index of suspicion, especially in ALS patients

presenting with sudden onset of dyspnoea or chest pain[6,7]. Early detection and appropriate management are crucial to prevent further respiratory compromise. The management of SP in ALS patients should be individualised, considering the patient's respiratory status, disease progression and overall prognosis. Treatment options include:

- *Observation:* Small, asymptomatic pneumothoraces may be managed conservatively with close monitoring[8].
- *Oxygen therapy:* Supplemental oxygen can facilitate the resorption of intrapleural air[9].
- *Interventional procedures:* Needle aspiration or chest tube placement may be necessary for large or symptomatic pneumothoraces[8].
- *Surgical intervention:* In cases of recurrent pneumothorax or persistent air leaks, surgical options such as video-assisted thoracoscopic surgery may be considered[10].

CONCLUSION

The occurrence of SP in amyotrophic lateral sclerosis patients can exacerbate respiratory insufficiency, leading to acute respiratory failure. Given the under-recognition of this latter complication, clinicians should maintain a high index of suspicion, especially in ALS patients presenting with sudden onset of dyspnoea or chest pain. Early detection and appropriate management are crucial to prevent further respiratory compromise.

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