CASE REPORT

Where the Wind Meets the Willow: Navigating the Unpredictable

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Abstract

Diaphragmatic paralysis, often stemming from neurological disorders like GBS or chronic inflammatory demyelinating polyneuropathy (CIDP), poses significant challenges in respiratory management. While bilateral phrenic nerve involvement can result in acute respiratory failure necessitating mechanical ventilation has been documented, instances of alternating hemidiaphragmatic paralysis are rare. Herein, we describe a case displaying this distinctive presentation of CIDP.

Keywords: Acute respiratory distress diaphragmatic paralysis, Case report, Chronic inflammatory demyelinating polyneuropathy, Phrenic nerve palsy.

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Introduction

Chronic inflammatory demyelinating polyneuropathy (CIDP) is a condition affecting the nervous system, marked by ongoing weakness and reduced sensation in the arms and legs. It's caused by inflammation of nerves, leading to damage to the protective covering of the nerves called myelin. This damage disrupts nerve signalling, leading to symptoms such as muscle weakness, tingling, numbness, and impaired coordination.

Our patient, diagnosed with CIDP, experienced alternating diaphragmatic paralysis, a condition scarcely documented in medical literature. The presentation of alternating paralysis, where diaphragm intermittently became paralysed while the other remained functional, posed a complex diagnostic and therapeutic dilemma. The patient's condition rapidly deteriorated, necessitating mechanical ventilation to manage acute respiratory failure.

This study underscores the necessity for additional studies into the underlying mechanisms and best treatment approaches for diaphragmatic paralysis linked to CIDP. Improved understanding of these rare manifestations is crucial for enhancing clinical outcomes and refining therapeutic approaches for patients with CIDP and related neurological disorders affecting respiratory function.

Case Study

Patient Information

- Age: 73 years old.
- Presenting complaint: Breathing difficulty, orthopnoea-like symptoms, slip and fall and head injury.
- Past medical history: Systemic hypertension.

Clinical Presentation

A 73-year-old female, came with a history of breathing difficulty and orthopnoea-like symptoms for one week (Fig. 1). Additionally,

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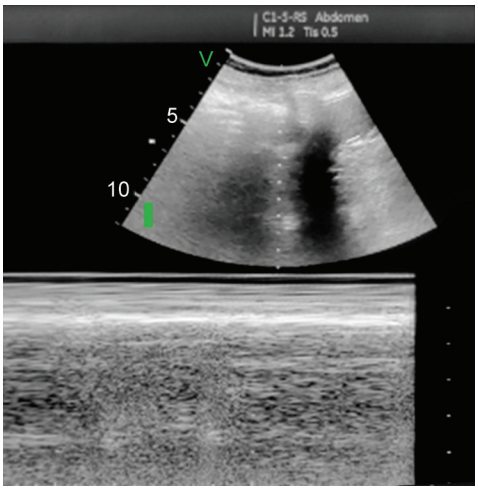


Fig. 1: Right diaphragm paralysis: There's no movement at all, showing up as a flat tracing on the ultrasound

she had sustained a head injury following a slip and fall at her home. Upon arrival at the hospital, she experienced aggravated breathing difficulty when put in supine position, symptomatically better on sitting position but still had increased work of breathing and persistent low oxygen saturation levels.[1,2]

Initial Workup

- **Cranial Nerve Examination:** All cranial nerve function were intact. Neck muscle weakness was present.
- Blood Investigation showed significant findings.
- **CT Brain and HRCT:** The patient underwent a CT scan to assess the extent of her head injury and lung status However, during the procedure, her breathing difficulty worsened, leading to respiratory arrest.
  - **CT Brain:** No acute infract and haemorrhage.
  - **HRCT lungs:** Subsegmental atelectasis of right middle lobe.

Interventions

- **Emergency intubation:** Due to the patient's worsening respiratory status that prompted respiratory arrest, she was emergently intubated to ensure adequate ventilation and oxygenation.
- **Mechanical ventilation:** Following intubation, the patient was placed on mechanical ventilatory support to assist with breathing on Volume control mode.

Clinical Course

- Despite initial stabilization with mechanical ventilation, the patient's respiratory status remained tenuous. She required ongoing ventilatory support due to the failure of weaning trials.
- Further assessment and management focused on optimizing respiratory parameters, addressing underlying pulmonary pathology, and providing supportive care to facilitate recovery.
- Neurological evaluation of nerve conduction study revealed demyelinating axonal motor sensory radiculopathy.
- Creatinine Kinase level was, 551.
- Nerve biopsy was suggestive of demyelinating neuropathy with focal secondary axonal loss.

Final Workup and Diagnosis

Subsequent failure of weaning trials led to suspicion of diaphragmatic involvement. Ultrasound sonography (USG) lung revealed impaired diaphragmatic movement, confirming the diagnosis.[3]

Management

- Patient was started on pulse steroids.
- Intravenous Immunoglobulin G (IVIg) 25 mg/day was given for 4 consecutive days.
- An empirical antibiotic cover was given to prevent infections.
- Tracheostomy was performed as patient needed to be prolonged mechanical ventilatory support.
- Diaphragm Pacing was suggested and planned as weaning trials failed.

Case Discussion

Diaphragmatic Paralysis in CIDP

Introduction

Chronic inflammatory demyelinating polyneuropathy is a condition affecting the nervous system, marked by ongoing weakness and reduced sensation in the arms and legs. While primarily affecting the limbs, CIDP can also involve respiratory muscles, leading to diaphragmatic paralysis and respiratory compromise.

Clinical Presentation

Chronic inflammatory demyelinating polyneuropathy may present with diverse neurological symptoms, including

- Muscle weakness.
- Sensory deficits.
- Impaired reflexes.

In some cases, respiratory involvement manifests as

- Dyspnoea.
- Orthopnoea.
- Respiratory failure.

Diaphragmatic paralysis, although rare, can occur, posing significant challenges in management.[4]

Diagnostic Workup

Diagnosis of CIDP involves clinical evaluation, electromyography (EMG), nerve conduction studies, and cerebrospinal fluid analysis.[5] In cases of suspected respiratory involvement, PFT and imaging such as chest X-rays or ultrasound may be warranted. Confirmation of diaphragmatic paralysis often requires specialized imaging modalities like fluoroscopy or MRI.

Management

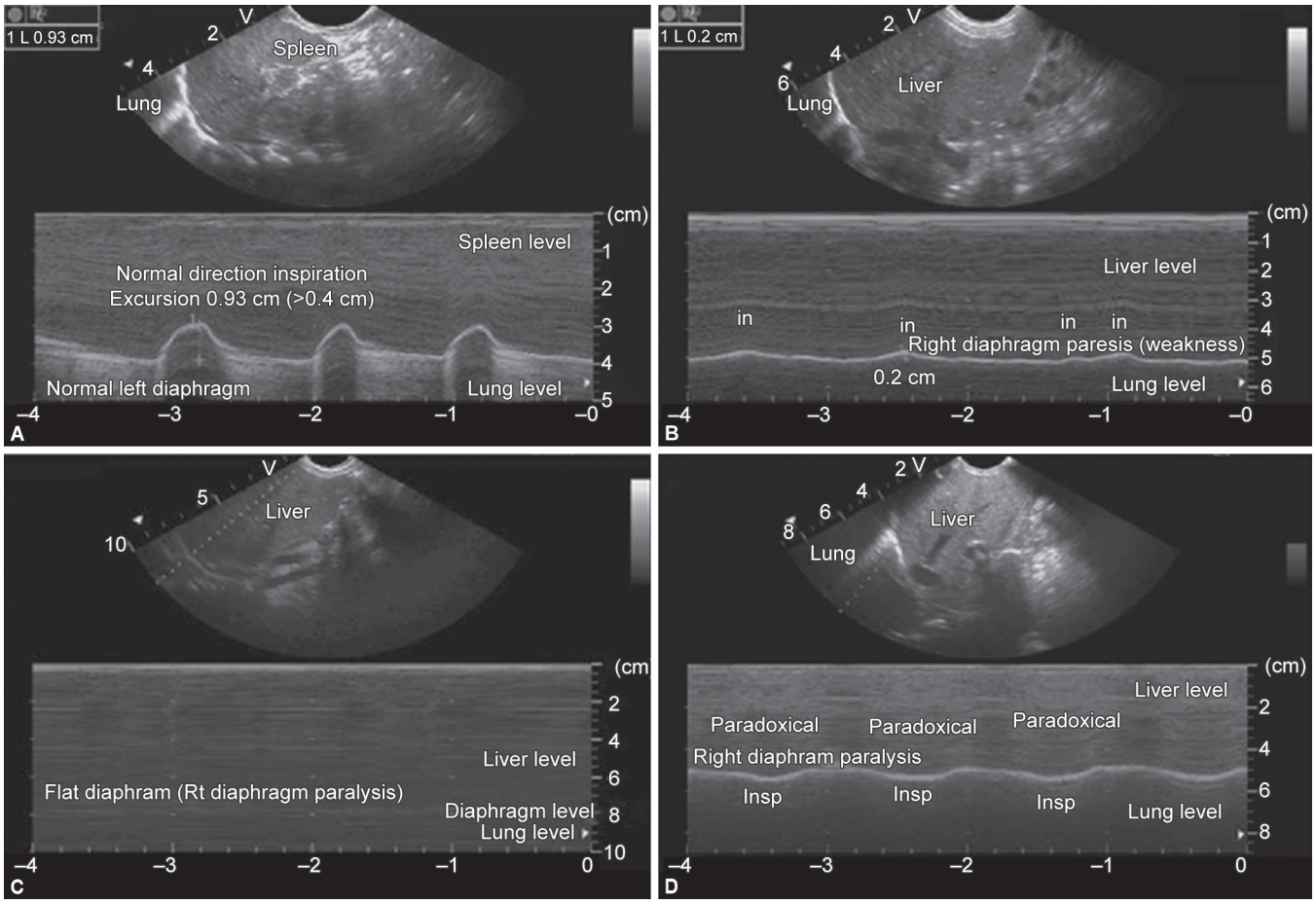
Management of diaphragmatic paralysis in CIDP revolves around immunomodulatory therapy to suppress autoimmune activity and prevent further nerve damage. High-dose steroids, IVIG or plasma exchange are commonly employed treatments. Additionally, respiratory support measures such as NIV or intubation may be necessary in severe cases of respiratory failure.[6]

Prognosis

The prognosis of diaphragmatic paralysis in CIDP varies depending on the extent of neurological involvement and response to treatment. Early recognition and intervention are crucial in preventing respiratory compromise and optimizing outcomes. Long-term management focuses on controlling disease activity, minimizing disability, and improving quality of life (Fig. 2).

Conclusion

Diaphragmatic paralysis in CIDP represents a rare but potentially life-threatening complication of this autoimmune disorder. Prompt recognition, thorough diagnostic evaluation, and multidisciplinary



Figs 2A to D: Lung US: M mode view, (A) Normal left diaphragm movement: It moves upward on the tracing during breathing in, with a height of more than 4 millimetres. We measure its excursion from the end of breathing out to the peak of breathing in; (B) Right diaphragm paresis: It still moves in the right direction, but the distance it moves is less than 4 millimetres; (C) Right diaphragm paralysis: There's no movement at all, showing up as a flat tracing; (D) Right diaphragm paralysis: It moves in the wrong direction during breathing in, which is called paradoxical motion

management involving neurologists, pulmonologists, and intensivists are essential for optimal patient care and outcomes. Further research is needed to elucidate the pathophysiology of respiratory involvement in CIDP and refine therapeutic approaches.[7,8]

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