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Lance Adams Syndrome In a Probable ALS Patient: A Case Report

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Introduction

- **Lance adams syndrome(LAS)** is a rare complication that can occur after hypoxic brain injury or following cardiac arrest, which is typically characterized by **involuntary jerking movements, also known as myoclonus.**
- We present a rare case of a patient who was diagnosed with **LAS** due to **repetitive events of hypoxemic brain injury,** which was attributed to newly detected **amyotrophic lateral sclerosis (ALS).**

Case presentation (70/F)

Chief complaints	General weakness, severe myoclonus, repetitive hypoxemia
Onset	3 months before admission
Vector	After cardiac arrest with CPR of 5 minutes
Associated symptoms	Gait disturbance, dysarthria, dysphagia, and cognitive impairment
Past history	14-year history of schizophrenia, 2-year of progressive bulbar palsy with no known cause.
Previous medication	Madopar HBS 1C BID, Trihexin 2mg 1T, levetiracetam 500mg BID, propranolol 5mg BID
Neurologic examination	✓ Alert mental status ✓ MMSE 20(7-4-1-8) ✓ MRC grade 4 in all extremities ✓ Positive in both upper motor neuron signs(bulbar symptoms, ankle clonus) and lower motor neuron signs(muscle wasting in all extremities and lumbosacral region) (Fig 1-A)
Brain images and electrophysiological studies	✓ Brain MRI: 1. Multiple old microbleeds in both cerebral hemisphere 2. Mild leukoaraiosis in both cerebral white matter (Fig 1-B) ✓ Brain FP-CIT PET: Normal DAT binding pattern => No evidence of Parkinson's disease. ✓ Electrodiagnostic study (Fig 1-C) 1. Widespread denervation in bulbar, cervical and thoracic segments, reinnervation in cervical segment, and 2. sensorimotor polyneuropathy in the bilateral arms and legs.

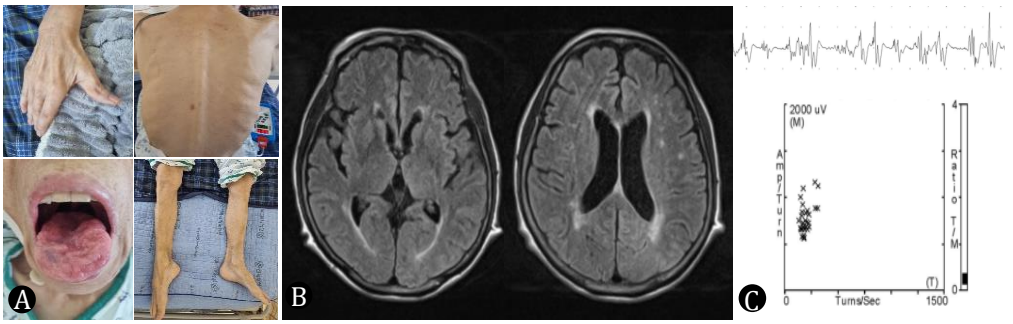

Fig.1 (A) Muscle atrophy in right hand, both legs, and axial muscles and tongue atrophy, (B) Brain MRI, (C) Electrodiagnostic study showing long duration and polyphasic MUAP in deltoid, and neuropathic pattern in turns-amplitude analysis

Treatment

- We prescribed aerobic exercise, which monitored the patient's subjective fatigue score, and **step-by-step functional and active daily living (ADL) training.** Also, we treated with levetiracetam 500mg BID, clonazepam 0.25mg TID and valproic acid 125mg TID, propranolol 5mg BID for the patient's myoclonus.
- **Bi-level positive airway pressure therapy** was applied for hypercapnia and hypoxemia during nighttime.

Outcome

- The patient's **myoclonus improved**, and her caregiver assistance in functional ability decreased. The hypercapnia and hypoxemia due to chronic respiratory failure were ameliorated, and there were **no more repetitive respiratory arrest events.**

Conclusion

- It is essential to consider **chronic respiratory failure** and any **neurological disease** in patients with mild hypoxic brain injury accompanying LAS, even in the absence of underlying heart disease. Accurate underlying diagnosis in patients with LAS can ultimately lead to appropriate rehabilitation goals, treatment, and monitoring, which can be tailored to the patient.