Coexisting Cerebellar Ataxia and Lambert-Eaton Myasthenic Without Malignancy: Insights from a Case and Systematic Literature

Prachi Mohapatra, Shikha Priya, Rajesh K. Singh, Deepti Vibha, A. Elavarasi, Jasmine Parihar, Manjari Tripathi
Department of Neurology, AIIMS New Delhi

Introduction

- LEMS: Rare autoimmune disorder, impaired acetylcholine release via P/Q-type VGCC antibodies.
- 50–60% associated with malignancy.
- Coexistence of cerebellar ataxia: Uncommon (≈ 9%); almost always paraneoplastic.
- LEMS + cerebellar ataxia without existing malignancy is extremely rare.

Case Report

- 75/M, Right-Handed, Farmer, No prior known comorbidities.
- Chronic smoker x 30 years, Exposure- 7.5 pack years
- C/C
- 1. Imbalance while walking with swaying to either and wide-based gait x 3 years
- 2. Tremulousness of both upper limbs x 3 years
- 3. Urinary Disturbances in form of urgency, frequency, urge incontinence x 3 years
- 4. Double vision and drooping of both eyelids x 1
- Also had h/o dry mouth and erectile dysfunction.

References- 1. Titulaer MJ, Lang B, Verschuuren JJ. Lambert-Eaton myasthenic syndrome: from clinical characteristics to therapeutic strategies. *Lancet Neurol*. 2011;10(12):1098-1107.
2. Wada S, Kamei M, Uehara N, Tsuzaki K, Hamano T. Paraneoplastic cerebellar degeneration and Lambert-Eaton myasthenic syndrome with SOX-1 antibodies. *Intern Med*. 2021;60(10):1607-1610.

Case Report

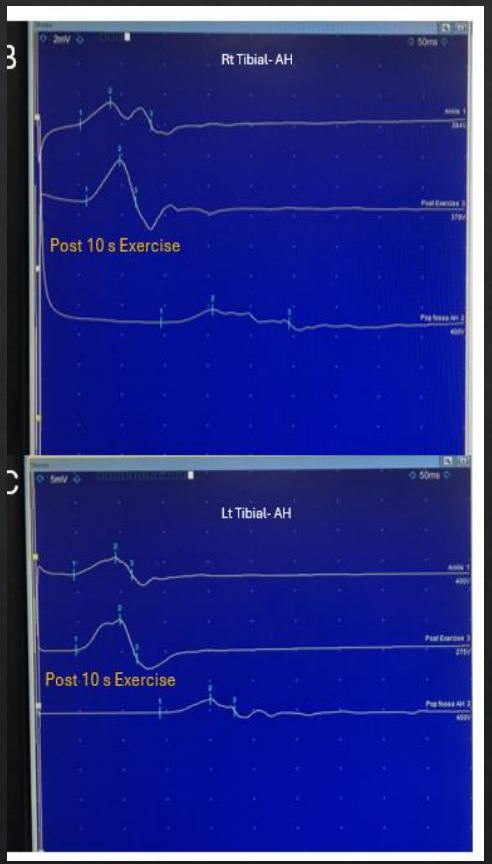
- O/E-
- Near-complete bilateral ptosis and complete ophthalmoplegia in all directions.
- Cerebellar features: Dysarthria, symmetrical bilateral appendicular ataxia, and severe gait ataxia
- Motor findings: Proximal lower limb weakness (power 4/5 at hips)
- Diminished reflexes, absent ankle jerks bilaterally.

Investigations

- Routine labs: normal, including autoimmune panels; AChR and MuSK antibodies negative.
- CSF: mild lymphocytic pleocytosis (10 cells/µL) and elevated protein (163 mg/dL).

Investigations

- Autoimmune encephalitis & paraneoplastic panels- negative.
- MRI brain: mild cerebellar atrophy.
- NCS: Reduced motor amplitudes in tibial nerves. 80% post-exercise facilitation after 10s of maximal contraction
- 3 Hz RNS- Low-amplitude CMAPs (<2 mV) in facial nerves. No significant decrement on 3 Hz RNS (facial, median, spinal accessory).
- High-frequency stimulation not feasible (poor cooperation).
- Serology: P/Q-type VGCC antibodies positive (158 pg/mL; cutoff >140).
- Malignancy screen: Whole-body 18F-FDG PET-CT, thyroid/testicular ultrasound, and serum tumor markers — no evidence of malignancy.



Treatment and Outcomes

- IVMP (1 g × 5d) + plasmapheresis (×5) → marked improvement in ptosis, ophthalmoparesis, proximal strength; mild gait benefit; ataxia unchanged.
- Pyridostigmine(60 mg TDS + Azathioprine (2 mg/kg/day) started.
- After 3-months: near-complete ocular recovery, improved ambulation (short distances independent, stick for long distances), moderate ataxia improvement.

Literature Review

- **66** published cases of coexistent LEMS + cerebellar ataxia reviewed (1991–present).

Conclusion

- Coexistence of LEMS and cerebellar ataxia without malignancy is rare, suggesting a potential autoimmune pathogenesis.
- Long-term cancer surveillance and timely immunotherapy are essential in such cases.

Variable	Value (n=66)
Age in years (Mean ± SD)	60.3 ± 6.8
Females, n (%)	25 (37.9%)
Males, n (%)	41 (62.1%)
Onset from symptoms to cancer diagnosis (months) (Median, IQR)/Range(min-max)	3.0 (0.5–6.0) / –60 to 39
Simultaneous onset of LEMS + cerebellar ataxia, n (%)	23 (34.8%)
LEMS preceding cerebellar ataxia, n (%)	18 (27.3%)
Cerebellar ataxia preceding LEMS, n (%)	22 (33.3%)
Anti-VGCC, n (%)	55 (83.3%)
Anti SOX1, n (%)	5 (7.6%)
Anti Hu, n (%)	2 (3.0%)
Negative Antibody, n (%)	4 (6.1%)
Malignancy Detected, n (%)	47 (71.2%)
Small Cell Lung Carcinoma (SCLC)	34 (51.5%)
Hilar Mass	1 (1.5%)
Non-Hodgkin Lymphoma (NHL)	1 (1.5%)
Prostate Adenocarcinoma	1 (1.5%)
Merkel Cell Carcinoma	2 (3%)
Squamous Cell Lung Cancer	1 (1.5%)
Neuroendocrine Tumour	2 (3%)
Primary Double Lung Cancer	1 (1.5%)
Non-Small Cell Lung Cancer	2 (3%)
Breast Cancer (Invasive Ductal Carcinoma)	1 (1.5%)
Nasopharyngeal Carcinoma	1 (1.5%)
No Malignancy Detected, n (%)	19 (28.8%)
Improvement in LEMS, n (%)	33 (50.0%)
Improvement in ataxia, n (%)	29 (43.9%)
Death, n (%)	14 (21.2%)
Death with associated malignancy	13 (19.6%)
Death without associated malignancy	1 (1.5%)