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Under the aegis of Department of Neurology

IMS-BHU



“A dance of missteps: Echoes of the cerebellum in the neuromuscular cleft”

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Background:

- Lambert–Eaton Myasthenic Syndrome (LEMS) is a rare autoimmune disorder affecting presynaptic voltage-gated calcium channels (VGCCs) at the neuromuscular junction.
- Cerebellar ataxia is recognized in paraneoplastic LEMS (P-LEMS),
- Its occurrence in non-paraneoplastic LEMS (NP-LEMS) is exceedingly rare

Case description:

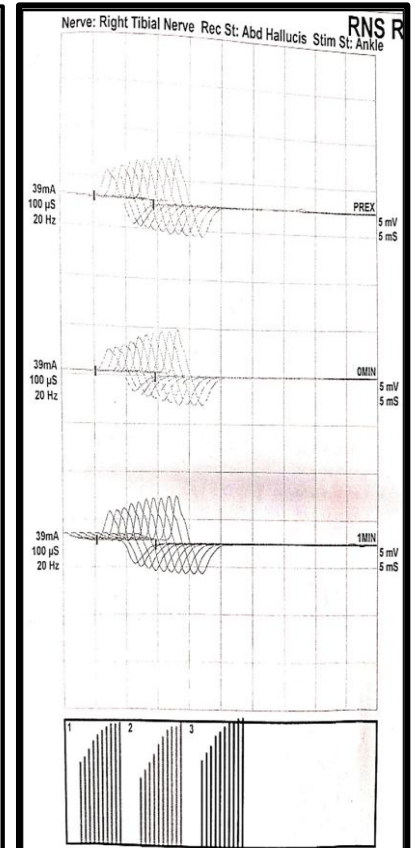
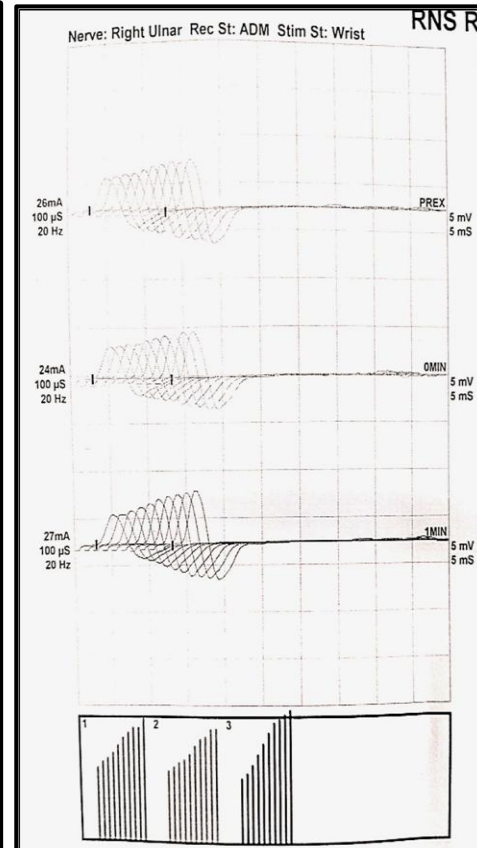
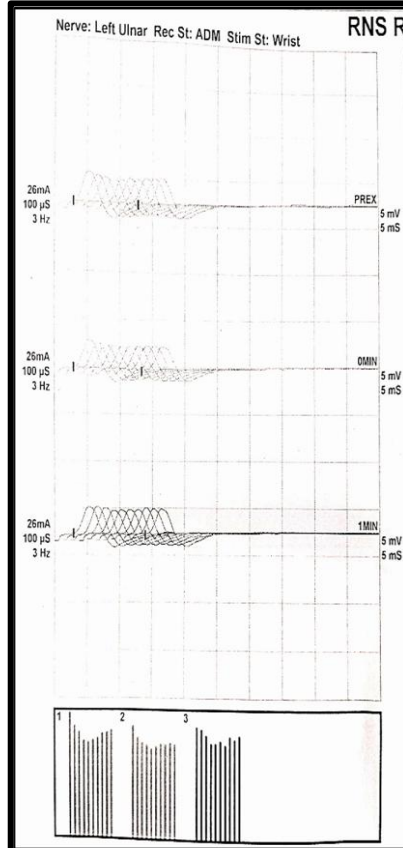
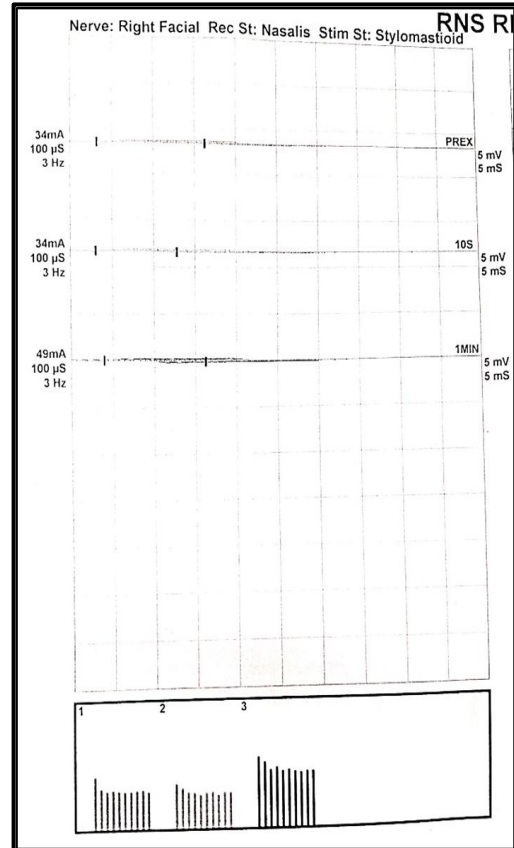
- A 63-year-old chronic smoker presented with progressive gait imbalance and recurrent falls over 4 months, worsened in low light and narrow spaces.
- Clinical examination revealed cerebellar signs (hypometric saccades, broken pursuits, mild limb incoordination, ataxic wide-based gait) and global hyporeflexia with preserved power and sensation.
- MRI brain and spine were unremarkable.
- Nerve conduction studies revealed axonal motor neuropathy.
- **Repetitive nerve stimulation test showed a significant decremental (>10%) and incremental (>100%) response.**
- Routine blood work, autoimmune panel, and CSF studies were normal.
- **Serum anti-VGCC antibodies were positive.**
- Imaging for malignancy (CECT NTAP) was negative.

Diagnosis: Cerebellar ataxia (CA) in NP-LEMS (Immune-mediated)

Treatment and follow up:

- The patient was treated with IVIg followed by oral corticosteroids

| Motor Nerve Conduction Study | | | | | | | | | |
|------------------------------|--------|--------|---------|----------|-----------------------|----------|----------|-------|---------|
| Site | Lat. | Dur. | Amp. | Area | Segment | Distance | Interval | NCV | CCV |
| Median Left | | | | | | | | | |
| Wrist | 4.9ms | 6.0ms | 1.2mV | 4.1mVms | *Wrist | | | 4.9ms | |
| Elbow | 9.1ms | 7.9ms | 940.0uV | 4.4mVms | Wrist-Elbow | 230mm | | 4.3ms | 54.1m/s |
| Elbow | 16.0ms | 7.8ms | 720.0uV | 3.4mVms | Elbow-Erb's | | | 6.9ms | |
| Median Right | | | | | | | | | |
| Wrist | 4.5ms | 5.8ms | 1.9mV | 6.2mVms | *Wrist | | | 4.5ms | |
| Elbow | 8.9ms | 6.8ms | 1.7mV | 5.4mVms | Wrist-Elbow | 220mm | | 4.4ms | 52.9m/s |
| Elbow | 16.5ms | 6.7ms | 1.1mV | 3.7mVms | Elbow-Erb's | | | 7.7ms | |
| Ulnar Left | | | | | | | | | |
| Wrist | 3.5ms | 6.7ms | 790.0uV | 2.3mVms | *Wrist | | | 3.5ms | |
| Elbow | 8.3ms | 6.9ms | 380.0uV | 1.3mVms | Wrist-Elbow | 250mm | | 4.8ms | 52.6m/s |
| Erb's | 14.6ms | 10.0ms | 250.0uV | 1.1mVms | Elbow-Erb's | | | 6.4ms | |
| Ulnar Right | | | | | | | | | |
| Wrist | 3.1ms | 4.4ms | 1.8mV | 3.1mVms | *Wrist | | | 3.1ms | |
| Elbow | 7.8ms | 5.8ms | 1.6mV | 4.1mVms | Wrist-Elbow | 250mm | | 4.7ms | 53.8m/s |
| Erb's | 14.7ms | 5.4ms | 1.3mV | 3.8mVms | Elbow-Erb's | | | 7.0ms | |
| Peroneal Left | | | | | | | | | |
| Ankle | 4.3ms | 4.9ms | 940.0uV | 2.4mVms | *Ankle | | | 4.3ms | |
| Head of fibul | 12.1ms | 3.7ms | 550.0uV | 1.2mVms | Ankle-Head of fibula | 380mm | | 7.8ms | 48.7m/s |
| | | | | | Head of fibula-Poplit | | | | |
| Peroneal Right | | | | | | | | | |
| Ankle | 4.4ms | 4.8ms | 550.0uV | 1.6mVms | *Ankle | | | 4.4ms | |
| Head of fibul | 13.0ms | 4.4ms | 460.0uV | 1.2mVms | Ankle-Head of fibula | 380mm | | 8.6ms | 44.4m/s |
| | | | | | Head of fibula-Poplit | | | | |
| Tibial Left | | | | | | | | | |
| Ankle | 4.4ms | 57.3ms | 1.9mV | 19.5mVms | *Ankle | | | 4.1ms | |
| Popliteal | 14.2ms | 5.8ms | 1.6mV | 4.1mVms | Ankle-Popliteal | 420mm | | 9.0ms | 42.9m/s |



Discussion:

- Cerebellar ataxia in NP-LEMS is infrequent and may obscure the diagnosis of LEMS.
- Anti-VGCC antibodies may cross-react with cerebellar structures, particularly Purkinje cells, contributing to ataxia.
- While LEMS symptoms often improve with immunotherapy

Conclusion:

NP-LEMS should be considered in patients with subacute cerebellar ataxia, especially when routine evaluations are inconclusive. Early detection and immunotherapy may improve neuromuscular symptoms and functional outcomes.