Case report on Lambert Eaton syndrome; *a rare autoimmune musculoskeletal disorder*

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***Abstract*- Lambert-Eaton syndrome (LES) or Lambert-Eaton myasthenic syndrome (LEMS), is a rare autoimmune disorder characterized by muscle weakness and impaired neuromuscular transmission. The case report being presented is of a 56-year-old female patient who complained of progressive muscle weakness, and autonomic dysfunction. The patient's symptoms posed a diagnostic challenge due to their overlapping features with other neuromuscular {myasthenia gravis} and autoimmune conditions. However, thorough clinical evaluation, electromyography (EMG), and serological testing, including the detection of P/Q-type voltage-gated calcium channel antibodies led to a confirmed diagnosis of LES.**

Management of this patient involved a multidisciplinary approach, involving neurologists and oncologists. Treatment plan focused on symptomatic relief and immunomodulatory therapy. The patient underwent physical therapy, respiratory assistance, and pharmacological interventions to manage symptoms associated with muscle weakness and autonomic dysfunction. Additionally, immunotherapy with intravenous immunoglobulins (IVIG) and potassium channel blockers, such as 3,4-diaminopyridine (3,4-DAP), was administered.

Despite initial challenges in the diagnosis and management of LES, the patient demonstrated a favorable response to treatment, with gradual improvement in muscle strength and autonomic symptoms over time. Regular follow-up visits, along with adjustment to the treatment regimen, are essential to maintain disease stability and optimize the patient's quality of life.

This case report highlights the importance of early recognition and accurate diagnosis of LES, particularly in patients with a history of malignancy. It emphasizes the significance of a comprehensive evaluation, including clinical assessment, electrophysiological studies, and serological testing, to differentiate LES from other neuromuscular disorders. Timely initiation of appropriate treatment strategies, encompassing both symptomatic relief and immunomodulatory therapy, can significantly improve patient outcomes and enhance their overall well-being. Further research and clinical studies are warranted to refine diagnostic criteria and optimize treatment protocols for Lambert-Eaton syndrome.

INTRODUCTION:

LES is an immune mediated attack against P/Q type voltage gated calcium channels on presynaptic cholinergic nerve terminals at neuromuscular junction and autonomic ganglia. Occurrences can be seen in two forms. In patients with lung cancer {small cell in 80% patients} (CA LES) or autoimmune (NCA LES).

Patients usually report with gradual onset of lower extremity weakness, dry mouth is a common symptom of autonomic dysfunction. Other features include erectile dysfunction, constipation, postural hypotension, and dry eyes. Diagnostic findings include CMAPs with low amplitude, which increase after brief maximum voluntary muscle activation. new antibodies (SOX -1) analogous to CA LES have been discovered.

Mechanism is probably from cross-linkage between VGCC and VGCC-antibodies down regulating VGCC expression. Taylor therapy to individual using IVIG and 3,4 DAP improve muscle strength and CMAP amplitude.

Patient Information:

Age: 56 years
Gender: Female

CHIEF COMPLAINS:
Generalized muscle weakness and ptosis since past one year.

HISTORY OF PRESENTING ILLNESS:
Patient presented to the neurology clinic with progressive muscle weakness and fatigue for past few months. She reported experiencing weakness predominantly in her proximal muscles, such as the hips, shoulders, and thighs; weakness has progressively worsened - affecting her ability to perform daily activities.
Associated complaints – ataxia, dizziness and giddiness, dyspnea, amnesia, paresthesia of skin

MEDICAL HISTORY:
There were no significant medical condition or illnesses reported.

PHYSICAL EXAMINATION:
General/constitutional: well; cooperative;
Eyes: conjunctivae: normal; pupils: equal, round and reactive; normal; Optic disc: normal; sclera: white; cornea: normal; normal; lens: normal; Extraocular muscles: intact; retina: red reflex present; visual acuity: normal;
Neck: appearance: normal; palpation: supple; thyroid: smooth and non-tender; Range of Motion: Normal;
Respiratory: auscultation: clear.
Cardio/vascular: auscultation: rhythm - regular. no murmurs, rubs or gallops;
GI: abdomen: *auscultation*: normal; palpation: non-tender;
Extremities: peripheral edema: none.

Neurological: Mental Status: *alert*; head & neck: *Flexion strength & tone*: 5+; *Extension strength & tone*: 5+; language: intact; cranial nerves: *II-XII*: intact; *I*: intact; *II*: intact; *III*: intact; *IV*: intact; *IX*: intact; *V*: intact; *VI*: intact; *VII*: Left intact, Right intact; *VIII*: intact; *X*: intact; *XI*: intact; *XII*: intact; R arm: *strength & tone*: Deltoid 5+, bicep 5+, triceps 5+, wrist flexion 5+, wrist extension 5+, finger flexion 5+, finger extension 5+, finger abduction (FDI) 5+, finger abduction (ADM) 5+, thumb abduction 5+; L arm: *strength & tone*: Deltoid 5+, bicep 5+, triceps 5+, wrist flexion 5+, wrist extension 5+, finger flexion 5+, finger extension 5+, finger abduction (FDI) 5+, finger abduction (ADM) 5+, thumb abduction 5+; R leg: *strength & tone*: hip flexion 5+, hip extension 5+, hip abduction 5+, hip adduction 5+, knee flexion 5+, knee extension 5+, ankle dorsiflexion 5+, ankle plantarflexion 5+, ankle eversion 5+, ankle inversion 5+, toe flexion 5+, toe extension 5+; L leg: *strength & tone*: hip flexion 5+, hip extension 5+, hip abduction 5+, hip adduction 5+, knee flexion 5+, knee extension 5+, ankle dorsiflexion 5+, ankle plantarflexion 5+, ankle eversion 5+, ankle inversion 5+, toe flexion 5+, toe extension 5+; sensation of pain (sharp): *head & neck*: intact; *trunk*: intact; L arm: intact; R arm: intact; L leg: intact; R leg: intact; sensation of touch (dull): *head & neck*: intact; *trunk*: intact; L arm: intact; *R arm*: intact; L leg: intact; R leg: intact; proprioception & vibration: *Proprioception*: Normal; *Vibration Sense*: Normal; deep tendon reflexes: *bicep(s)*: 2+; *triceps'*: 2+; *patellar*: 2+; *Achilles*: 2+; *plantar/Babinski*: Left down going toes, Right down going toes; coordination: *heel-to-shin*: intact; *finger-to-nose*: intact; *Rapid Alternating Movement(s)*: intact; *Rhomberg*: negative; gait and station: intact.

DIAGNOSIS:
Blood tests: Routine blood investigations, were all within normal limits.
Electromyography (EMG): revealed low compound muscle action potentials (CMAPs) amplitudes, incremental response on repetitive nerve stimulation, and post-exercise facilitation.
Autoantibody testing: elevated levels of VGCC antibodies.
Imaging: No significant findings were observed.

TREATMENT:
Firdapse 10mg, Gammaplex 10% inj, pyridostigmine 180mg, gabapentin 400mg, topiramate 50mg. Physical therapy was also recommended to maintain muscle strength and function.

The patient was scheduled for regular follow-up visits with both the neurology and oncology departments to monitor the response to treatment, adjust medication dosages, and assess cancer status.

DISCUSSION:

Lambert-Eaton Syndrome (LES) is a rare autoimmune disorder characterized by muscle weakness and impaired neuromuscular transmission. It is often associated with small cell lung cancer (SCLC) and is thought to be immune-mediated, antibodies against VGCCs impair release of acetylcholine at the neuromuscular junction.

Diagnosis of LES involves clinical assessment, electromyography findings, and the presence of VGCC antibodies. Identifying an underlying malignancy, particularly small cell lung cancer, is crucial.

Treatment of LES focuses on managing symptoms and improving neuromuscular transmission. Medications such as 3,4-diaminopyridine (DAP) and pyridostigmine can be used to enhance acetylcholine release or inhibit its breakdown. Additionally, physical therapy and close monitoring of underlying malignancy are essential for comprehensive patient care.

CONCLUSION:

This case report emphasizes the importance of considering Lambert-Eaton Syndrome in patients presenting with proximal muscle weakness, particularly in the context of small cell lung cancer. Early diagnosis and a multidisciplinary approach involving neurology and oncology specialties are crucial in providing appropriate management and improving patient outcomes.

REFERENCES:

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