Case report:

Lambert-Eaton myasthenic syndrome: Case report

Dr. Abhishek Bansal, Dr. Amit Kothari, Dr. Nilesh Jagdale, Dr. Tanvi Batra, Dr. A.L. Kakrani, Dr. V.S. Gokhale

Background:
Lambert-Eaton myasthenic syndrome (LEMS) is a presynaptic disorder of the neuromuscular transmission characterised by impaired quantal release of acetylcholine that causes proximal muscle weakness, depressed tendon reflexes and post tetanic potentiation; additionally, autonomic changes are present. Here with we presented a case of A 55 years old male patient presented with progressive weakness in both the lower limbs, difficulty in rising from a chair and climbing stairs, followed by difficulty in lifting heavy weights and hand grip weakness since 4-5 months. Repetitive nerve stimulation test (RNST) was also performed on Rt & Lt median & ulnar nerves which were suggestive of decremental response at low frequency (3 Hz) & an incremental response at high frequency (30 Hz). These findings were suggestive of a pre synaptic neuromuscular junction disorder and the possibility of Lambert Eaton Myasthenic Syndrome (LEMS) was considered. Final diagnosis of a pre-synaptic neuromuscular junction, Lambert Eaton Myasthenic Syndrome (LEMS) with small cell carcinoma of Lung was established.

Keywords: Lambert-Eaton myasthenic syndrome, Repetitive nerve stimulation test

Case Report
A 55 years old male patient presented with progressive weakness in both the lower limbs, difficulty in rising from a chair and climbing stairs, followed by difficulty in lifting heavy weights and hand grip weakness since 4-5 months. He also had difficulty in chewing and swallowing food. There were no h/o sensory disturbances, gait imbalance, incoordination, ptosis, dysarthria or fluctuation in symptoms. There was no h/o fever, joint pain & rash, no bowel and bladder incontinence. There was no h/o drug/toxin exposure. All the vitals were stable on admission. Pulse was 84/min and blood pressure was 114/72 mm Hg. Based on the history myopathy, myasthenia gravis and chronic inflammatory demyelinating polyneuropathy were the initial differentials. Routine serological investigations were normal. Total Creatinine Phosphokinase (CPK Nac) was within normal limits which ruled out myopathy. As there were no sensory abnormalities CIDP was less likely; also absence of ptosis & no fluctuation in symptoms were against...

www.ijhbr.com  ISSN: 2319-7072
The diagnosis of myasthenia Gravis. General examination was unremarkable. Neurological examination was suggestive of proximal & distal muscle weakness (power grade 4/5) in both the upper and lower limbs. Deep tendon reflexes were normal. Further, patient also revealed two additional interesting symptoms of dryness of mouth & impotence since 6 months which were s/o autonomic dysfunction. In view of areflexia & muscle weakness, nerve conduction study (NCS) was performed which revealed low amplitude compound muscle action potentials (CMAPs) in Rt & Lt. median, ulnar, common peroneal and tibial nerves with normal conduction velocities. Repetitive nerve stimulation test (RNST) was also performed on Rt & Lt median & ulnar nerves which were suggestive of decremental response at low frequency (3 Hz) & an incremental response at high frequency (30 Hz). These findings were suggestive of a pre synaptic neuromuscular junction disorder and the possibility of Lambert Eaton Myasthenic Syndrome (LEMS) was considered.

As LEMS is often a non-metastatic manifestation of malignancy, a search for malignancy anywhere in the body was carried out. Chest radiography was s/o segmental collapse or consolidatory changes in Rt upper lobe. HRCT thorax confirmed the X ray findings along with small calcified lymph nodes s/o infective etiology. Bronchoscopy was done and broncho-alveolar lavage (BAL) and lung mass biopsy was taken. Histopathological examination of the lung mass was s/o Small Cell Carcinoma.

Final diagnosis of a pre-synaptic neuromuscular junction, Lambert Eaton Myasthenic Syndrome (LEMS) with small cell carcinoma of Lung was established.

**Discussion**

About 60% of patients have a small cell lung cancer associated with recorded or observed (SCLC) \(^3\). Evidence that it is an autoimmune disease mediated by antibodies to voltage gated calcium channels (VGCCs) at motor nerve terminals includes the clinical response to plasma exchange \(^4\). It was first described as a paraneoplastic syndrome in patients with lung cancer but we now know about half of the patients with LEMS do not have cancer. When tumour occurs it is usually SCLC \(^5,6\). It is known that electrophysiological findings of LEMS may occasionally overlap with those seen in myasthenia gravis; a decremental response at low stimulation rate, normal CMAP amplitudes and absent facilitation at high stimulation rate. Furthermore, facilitation up to 50% can also be seen in myasthenia gravis \(^7\); complicating electrophysiological diagnosis of the two myasthenic syndromes.

(a) Upper curve shows low amplitude compound muscle action potential (CMAP) & a decremental response at 3 Hz stimulation.

(b) Lower curve shows an incremental response at 30 Hz stimulation.

Treatment: for LEMS with underlying Small Cell Carcinoma of Lung: chemotherapy for the tumor, takes care of LEMS as well

For symptoms of LEMS Acetylcholine inhibitors, guanidine, aminopyridines

To deplete serum autoantibodies: plasmapheresis

And immunosuppression can be carried out. Our patient was started on chemotherapy, but succumbed to pneumonia with septicaemia during the course of chemotherapy.
Clinical implications: LEMS though rare has to be thought of in all patients presenting with new onset progressive muscle weakness, diagnosis should be confirmed by Nerve Conduction Velocity and a thorough search has to be made for underlying malignancy.

References: