REVIEW ARTICLES

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Lambert-Eaton myasthenic syndrome – a misdiagnosed condition

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Abstract

Background: Lambert-Eaton myasthenic syndrome (LEMS) is a rare disorder of the neuromuscular junction. Clinical features include proximal muscle weakness, markedly in the lower limbs, reduced deep tendon reflexes that can increase after exercise, and autonomic disturbances. The clinical picture as well as knowledge of the laboratory test that accompany LEMS will permit early recognition of the disease, that is crucial because it is often associated with malignancy, especially small cell lung cancer (SCLC). In this article we present a patient with proximal muscle weakness and typical changes on repetitive nerve stimulation, as well as a short literature review on the topic.

Conclusions: The diagnosis of LEMS is usually made on clinical grounds. The diagnosis is confirmed by electrophysiological testing, main features including decrement response on slow repetitive nerves stimulation (3Hz), and an increment of more than 100% in CMAP amplitude after brief exercise, or high frequency repetitive stimulation (30-50 Hz). Immunological panel assay with positive P/Q-type VGCC antibody is strongly suggestive of LEMS. While symptomatic treatment with 3,4 – diaminopyridine is available, one of the main priorities is evaluation for underlying malignancies in these patients, the most common being SCLC. Evaluation of patients with LEMS and no known cancer should start with CT of the chest, abdomen and pelvis. Brain imaging is recommended if focal neurological signs are present. If the initial evaluation of the patient is negative, repeated screening for malignancy after 6 months and up to two years is recommended.

Key words: Lambert-Eaton myasthenic syndrome, cancer, weakness, increment.