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Letter to the Editor

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Lambert-Eaton Myasthenic Syndrome with A Twenty-Three-Year Delay in Diagnosis

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Dear Editor,

Lambert-Eaton myasthenic syndrome (LEMS) is a rare neuromuscular junction disorder resulting from the development of auto-antibodies against voltage-gated calcium channels (VGCCs) in pre-synaptic terminals (1). It has tumoral and non-tumoral forms; the former is associated with small cell lung cancer. Clinically, it is characterized by proximal weakness, autonomic symptoms, and loss/reduction of deep tendon reflexes (2). Muscular weakness, frequently occurring in the lower extremities, almost always begins symmetrically in the proximal muscles, progressing to involve the distal muscles over time (3). It may be confused with myopathic disorders due to the presence of symmetrical muscular weakness involving the proximal muscles. Herein we present the case of a non-tumoral LEMS patient who was diagnosed as having myopathy due to weakness that started in the legs nearly 23 years ago. Written consent was taken from the patient.

A 51-year-old female recalled having difficulty in climbing up the stairs approximately 23 years ago. At that time, she had had a slowly progressing clinical course. Electromyography (EMG) and muscle biopsy were first performed 2 years after the initial onset of her symptoms, and she had diagnosed as myopathy. After that, she never received medical treatment, although she had ptosis in the past 10 years and was unable to walk without assistance in the past 1 year. She had no dryness of the mouth, constipation, or sweating. Her general physical examination was unremarkable, and she had no orthostatic hypotension. In her neurological examination, she was found to have ptosis bilaterally, with minimal limitation in outward eye movements in both eyes. She had normal speech and normal lower cranial nerve examination results. Her muscle power in the upper extremities was 4/5 proximally, while her muscle power in the lower extremities was 2/5 and 4/5 proximally and distally, respectively. Except for a Creatinine Kinase (CK) level of 302 U/L, she had normal blood biochemistry values. EMG showed a low motor response amplitude in nerve conduction studies. Needle EMG showed myogenic MUP changes in extremity muscles. In the repetitive stimulation of the abductor digiti minimi, a continuous pathological decrement was noted with a stimulation frequency of 3 Hz, while a 300% increase in amplitude was found at 50 Hz for 4 s (Figure 1). In a single-fiber EMG examination, a single jitter was detected in five potential outputs recorded in the right EDC muscle. The VGCC antibody level was elevated (321 pmol/L), and work-up for paraneoplastic conditions was negative. Intravenous immunoglobulin, pyridostigmine, 3.4-diaminopyridine, and azathioprine were administered on the basis of a diagnosis of non-tumoral LEMS with partial improvement.

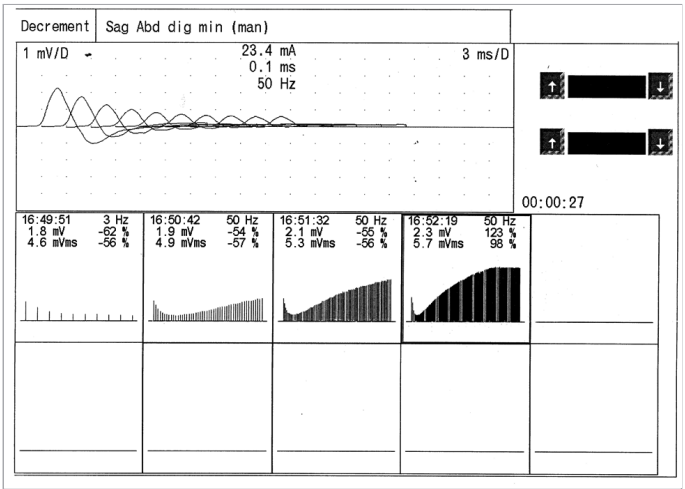

Figure 1. A repetitive neurological study with ADM recording: Continuous pathological decrement with a 3-Hz stimulation frequency and 300% increase in amplitude with a 50-Hz stimulation of 4 s

As LEMS is a condition that affects the neuromuscular junction, myasthenia gravis represents the entity that is most frequently considered in the differential diagnosis. However, proximal weakness may also lead to misdiagnosis as myopathy, particularly in case of mild weakness and slow

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progression in the non-tumoral type as well as in the presence of electrophysiological myogenic changes, similar to our case (4). The reported time to diagnosis from the onset of symptoms in LEMS varies between 2 months and 25 years (5). The diagnostic delay in our patient encompassed a period of 23 years. Although absent in our patient, autonomic symptoms, which are generally mild with no significant effects on functions, are seen in many cases (6). Thus, it should be remembered that these symptoms may be easily overlooked unless actively questioned.

With this case presentation, we would like to emphasize that in patients with proximal muscular weakness, autonomic symptoms, loss/decrease in DTR, and low motor amplitudes despite normal sensory conduction in electrophysiological studies, a diagnosis of LEMS should be kept in mind. Even in the presence of myogenic changes on needle EMG, repetitive nerve stimulation should be performed in these subjects.

Informed Consent: Written informed consent was obtained from patient who participated in this study.

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REFERENCES

1. Titulaer MJ, Lang B, Verschuuren JJ. Lambert-Eaton myasthenic syndrome: from clinical characteristics to therapeutic strategies. Lancet Neurol 2011; 10:1098-1107. [CrossRef]
2. O'Neill JH, Murray NM, Newsom-Davis J. The Lambert-Eaton myasthenic syndrome. A review of 50 cases. Brain 1988; 111:577-596. [CrossRef]
3. Titulaer MJ, Wirtz PW, Kuks JB, Schelhaas HJ, van der Kooi AJ, Faber CG, et al. The Lambert-Eaton myasthenic syndrome 1988-2008: a clinical picture in 97 patients. J Neuroimmunol 2008; 201-202:153-158. [CrossRef]
4. Crone C, Christiansen I, Vissing J. Myopathic EMG findings and type II muscle fiber atrophy in patients with Lambert-Eaton myasthenic syndrome. Clin Neurophysiol 2013; 124:1889-1892. [CrossRef]
5. Pellkofer HL, Armbruster L, Linke R, Schumm F, Voltz R. Managing non-paraneoplastic Lambert-Eaton myasthenic syndrome: clinical characteristics in 25 German patients. J Neuroimmunol 2009; 217:90-94. [CrossRef]
6. Gilhus NE. Lambert-eaton myasthenic syndrome; pathogenesis, diagnosis, and therapy. Autoimmune Dis 2011; 2011:973808. [CrossRef]