J Neurol Disord
Volume 10:7, 2022

Journal of Neurological Disorders

Short Communication Open Access

Autonomic Dysfunction and Suspicious Small Fiber Involvement in Lambert-Eaton Myasthenic Syndrome

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Description

Lambert-Eaton Myasthenic Syndrome (LEMS) is a type of paraneoplastic syndrome that may initially manifest itself with proximal weakness and gait abnormalities. Approximately up to 50%of LEMS patients have a primary autonomic dysfunction. We report a case of 75-year-old male with symmetric proximal muscle weakness, dry mouth and constipation. The cutaneous response to scratch and upright tilt-table testing were positive. Repetitive nerve stimulation test showed decremental response of Compound Muscle Action Potential (CMAP) amplitude at 3 Hz and incremental response at 20 Hz. The test of Skin Sympathetic Response (SSR) detected a lower amplitude for the left hand while a lower amplitude and longer response duration was found in the right palm, but it evoked no response in the sole. The presence of antibodies against Voltage-Gated Calcium Channels (VGCC) confirmed the diagnosis. This is the first case, to our knowledge, in which Intraepidermal Nerve Fibers Density (IENFD) test *via* skin punch biopsy has been performed for a patient suffering from LEMS with autonomic disorder and it confirms that pathologically, small fiber can be affected by LEMS.

A 75-year-old male with no special medical history was inflicted with dry mouth and constipation for two months, and symmetric proximal muscle weakness for one month. The muscle strength of the both proximal limbs was 4/5, and that of the distal limbs 5/5. There was no sign of tendon reflexes in all the limbs while the reflex of the right knee increased to 1+ after a sustained 30-second contraction. Notably, the cutaneous response to scratch was positive. Orthostatic hypotension was found by upright tilt-table testing. Chest computed tomography, brain magnetic resonance imaging, ultrasound scan with abdomen and lymph nodes and serum tumor markers showed absolutely no evidence of tumor. Electromyography showed no neurogenic or myogenic damage, and the conduction velocity of the sensory nerve and motor nerve was normal. Repetitive nerve stimulation test showed decremental response of Compound Muscle Action Potential (CMAP) amplitude ranging from 13% to 27% at 3 Hz and incremental response ranging from 45% to 112% at 20 Hz in left median nerve, left ulnar nerve and bilateral common peroneus nerve. The test of Skin Sympathetic Response (SSR) detected a lower amplitude for the left hand while a lower amplitude and longer response duration was found in the right palm, but it evoked no response in the sole (Figure 1). Voltage-Gated Calcium Channels (VGCC) antibody was positive while anti-SRY-Related HMG-Box Gene 1(*SOX1*) antibodies was negative. All considered, the patient was diagnosed with Lambert-Eaton syndrome. Because of the prominent and early symptom of autonomic disorder, the patient further underwent Intraepidermal Nerve Fibers Density (IENFD) test *via* skin biopsy in distal leg (10 cm above lateral malleolus) so as to identify the substantial pathological evidence. His IENFD is 7.2(mm)$^{-1}$ while the normative value for clinical use among 70 years to 79 years old is 7.7(mm)$^{-1}$ [1]. According to international recommendations [2], the IENFD lower than 7.6(mm)$^{-1}$ at the distal leg has a diagnostic value with specificity of 90% and sensitivity of 82.8%. After a five-day treatment by intravenous immunoglobulins, the patient's clinical symptoms improved (Figure 2).

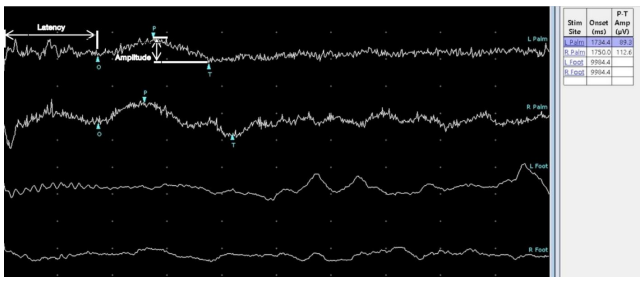

Figure 1. Sympethetic Skin Response (SSR) recorded from bilateral palms and feet detected a lower amplitude for the left hand, a lower amplitude and longer response duration, in the right palm, and no response was evoked in both soles.

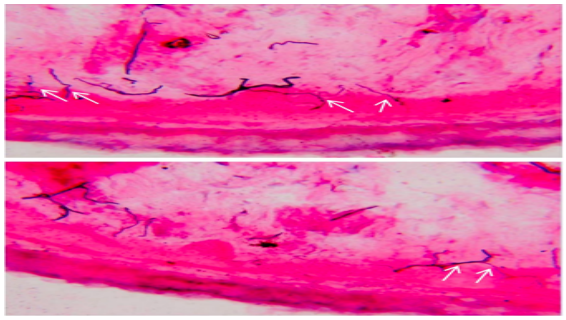

Figure 2. PGP 9.5 immunostaining of skin biopsy sections shows that intraepidermal nerve fiber density is 7.2(mm)$^{-1}$ while the normative value for clinical use in his age is 7.7(mm)$^{-1}$.

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Received: 18-Aug-2022, **Manuscript No.** JND-22-001-Preqc-22; **Editor assigned:** 22-Aug-2022, **PreQC No:** JND-22-001-Preqc-22 (PQ); **Reviewed:** 06-Sep-2022, **QC No:** JND-22-001-Preqc-22; **Revised:** 12-Sep-2022, **Manuscript No:** JND-22-001-Preqc-22 (R), **Published:** 20-Sep-2022; DOI: 10.4172/2329-6895.10.7.502

Lambert-Eaton Myasthenic Syndrome (LEMS), first reported by Edward Lambert and Lee Eaton in 1957 [3], is a type of paraneoplastic syndrome that may initially manifest itself with proximal weakness and gait abnormalities. Approximately up to 50% of LEMS patients have a primary autonomic dysfunction including dry mouth, constipation, loss of sweating, orthostatic hypotension, pupillary abnormalities etc [4-7]. In recent years, skin biopsies are making great inroads into the study of autonomic nerve disorders, In the case presented above, a history of obvious dry mouth and constipation at an early stage with abnormalities in SSR, tilt-table testing and cutaneous response to scratch is a compelling reminder of autonomic dysfunction.

Skin punch biopsy, a minimally invasive test that can be performed easily and quickly, serves as a window into autonomic disorders. Despite the recent advances, as far as many diseases of the autonomic nervous system are concerned, there is a lack of detailed descriptions of the autonomic innervation derived from skin biopsies. This is the first case, to our knowledge, in which skin punch biopsy along with SSR has been performed for a patient suffering from LEMS with autonomic disorder and comfirms that the small fiber can be affected in LEMS.

Conflict of Interest

There are no conflicts of interest to declare.

Ethical Publication Statement

The patient has provided consent for the use of the clinical information. This study has been approved by the Chinese PLA General Hospital Ethical Review Committee and has therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments.

Funding

This work was supported by National Natural Science Foundation of China (No. 81771358).

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How to cite this article: Zhang, Jinghong, Xusheng Huang and Qiang Shi. "Autonomic Dysfunction and Suspicious Small Fiber Involvement in Lambert-Eaton Myasthenic Syndrome." *J Neurol Disord* 10 (2022) : 502.