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【 LETTERS TO THE EDITOR 】

Reply to: Myopathy in Myasthenia Positive for Antibodies Against MuSK, Lrp4, and Titin

Key words: myasthenia gravis, anti-MuSK antibody, anti-Lrp4 antibody, anti-titin antibody

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The Authors Reply We thank Dr. Joseph Finsterer and Dr. Marlies Frank for their comments on our article, "Anti-MuSK positive myasthenia gravis with anti-Lrp4 and anti-titin antibodies." We reported a 62-year-old woman who showed positivity for anti-MuSK, anti-Lrp4, and anti-titin antibodies. She developed myasthenia gravis (MG) crisis following a 10-year history of intermittent double vision with ptosis. Needle electromyography (EMG) and a muscle biopsy revealed myopathic changes (1). We would like to respond to the questions by clarifying the details regarding the issues that were mentioned.

The patient had not visited any hospital because of ptosis or double vision before the sudden episode of unconsciousness. This is why it took 10 years to diagnose MG.

When the patient visited our hospital, her blood gas analysis showed elevated levels of both partial pressure of CO_2 and bicarbonate, indicating respiratory acidosis rather than metabolic acidosis. However, an elevated lactate level was also detected (41 mg/dL; misprinted as 41 Eq/dL). We assumed that the initial shock caused a transient increase in the lactate levels.

Regarding EMG, motor unit potential showed early recruitment with a normal duration. Polyphasic potential was observed only in the sternocleidomastoid muscle. Repetitive nerve stimulation tests of patients with muscle-specific kinase (MuSK)-MG often show normal results, especially in the limb muscle (2). Consistently, no apparent warning signs were noted in the right trapezius or abductor digiti minimi. Single-fiber EMG was not performed. Based on these findings, we initially suspected myopathy or MG. We performed a muscle biopsy to eliminate the possibility of myopathy.

Although the results of Gomori-trichrome staining were normal and we did not perform electron microscopy, the muscle biopsy showed myopathic changes without inflammatory cell infiltration. Consistent with MG, the level of anti-MuSK antibody decreased from 28.6 nmol/L to 4.6 nmol/L (normal, <0.02 nmol/L) after 2 months of intensive immunomodulatory therapies. Therefore, the diagnosis of MG was established, accompanied by biopsy-proven mild myopathic changes.

Myopathic changes are frequently detected on EMG in patients with MuSK-MG (3, 4). The present patient also showed elevated levels of anti-Lrp4 and anti-titin antibodies. These antibodies might have contributed to the augmentation of the disease severity of MG by accelerating autoimmune reactions against not only the neuromuscular junctions but also the muscle fibers.

The authors state that they have no Conflict of Interest (COI).

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