Internal Medicine
The Japanese Society of Internal Medicine
doi: 10.2169/internalmedicine.6180-20
Intern Med 60: 1485, 2021
http://internmed.jp

【 LETTERS TO THE EDITOR 】

Myopathy or Myasthenia Positive for Antibodies Against MuSK, Lrp4, and Titin

Key words: MuSK, Lrp4, myasthenia, repetitive nerve stimulation, myopathy

(Intern Med 60: 1485, 2021)
(DOI: 10.2169/internalmedicine.6180-20)

To the Editor We read with interest the article by Yamashita et al. about a 62-year-old woman with myasthenia due to antibodies directed against MuSK, Lrp4, and titin (1). It was concluded that in myasthenia patients, double- or triple-positive for antibodies against MuSK and Lrp4, respectively MuSK. Lrp4, and titin, not only the neuromuscular junction but also the muscle can be a target of the autoimmune response (1). We have several comments and concerns regarding these conclusions.

We would like to know why it took 10 years to diagnose myasthenia. The patient's 10-year history of ptosis and double vision, even if intermittent, should have prompted the treating physicians to consider myasthenia as a differential diagnosis earlier.

Unfortunately, no reference limits were provided for the blood gas analysis. We would like to know if the lactate value of 41 mEq/L was normal or not. If elevated, an explanation for lactic acidosis should be provided. There was obviously acidosis, but the elevated values of partial pressure of CO_2 suggest respiratory acidosis rather than metabolic acidosis.

The patient underwent needle electromyography (EMG), but whether the mean duration of motor-unit action potentials was increased or decreased is not mentioned. It was also not mentioned if the interference pattern was already dense at submaximal voluntary contraction. Since a muscle biopsy indicated muscle damage and nerve conduction studies were normal, we expect a myogenic rather than a neurogenic EMG pattern.

We would like to know if the results of repetitive nerve stimulation were normal because the patient was already under treatment with choline-esterase inhibitors when the investigation was carried out. Supposing the patient was not under anti-myasthenic treatment at the time of repetitive nerve stimulation, normal repetitive nerve stimulation should be explained.

Since the diagnosis of myasthenia was supported only by elevated antibodies and the clinical presentation, we would like to know if single-fibre EMG was carried out and if the jitter and number of blockings were increased. During myasthenic crisis, single-fibre EMG should indicate a transmission defect.

The fact that the patient underwent a muscle biopsy is unusual. Since nicotinamide adenine dinucleotide (NADH) staining revealed a disorganised intermyofribrillar network and one COX-negative fibre was detected, we would like to know the results of the Gomori-trichrome staining and the ultrastructural investigations conducted by electron microscopy. We would also like to know if the authors considered a metabolic myopathy rather than myasthenia. We are curious if the elevated antibody titres normalised with clinical recovery and under treatment with steroids, tacrolimus, cyclosporine, and pyridostigmine.

Overall, this interesting report has shortcomings as outlined, which should be addressed before drawing final conclusions.

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

Josef Finsterer[1] and Marlies Frank[2]

Reference

1. Yamashita R, Shimizu M, Baba K, et al. Anti-MuSK positive myasthenia gravis with anti-Lrp4 and anti-titin antibodies. Intern Med **60**: 137-140, 2021.

[1]Municipal Hospital Landstrasse, Messerli Institute, Austria and [2]First Medical Department, Municipal Hospital Landstrasse, Austria
Received: August 26, 2020; Accepted: October 18, 2020; Advance Publication by J-STAGE: December 7, 2020
Correspondence to Dr. Josef Finsterer, fifigs1@yahoo.de
