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Treatment Patterns and Outcomes of Anti-MuSK Antibody-Positive Myasthenia Gravis: A Retrospective Single-Center Study

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ABSTRACT

Background: There is limited knowledge regarding the real-world treatment status of muscle-specific tyrosine kinase antibody-positive myasthenia gravis (MuSK-MG) patients in clinical practice. The aim of this study was to analyze the overall treatment patterns and outcomes of MuSK-MG in Korea.
Methods: In this single-center cohort study, we retrospectively included patients diagnosed with MuSK-MG. We analyzed their demographic features, clinical characteristics, treatment patterns focusing on corticosteroids (CSs), non-steroidal immunosuppressants (NSISs), and rituximab, and treatment outcomes. Specifically, we investigated the achievement of minimal manifestation status with a prednisolone dose 5 mg/day or below (MM-5).
Results: The study included 30 patients with MuSK-MG, predominantly female (93.3%). The average age at time of diagnosis was 43.9 ± 16.0 years, with the onset age of 42.3 ± 16.2 years. All patients initially received CSs, and 24 patients (80%) were treated with NSISs. Tacrolimus was the most frequently prescribed NSIS, used in 70.8% of cases, followed by azathioprine (62.5%). All 10 patients who received rituximab therapy showed clinical improvement, with 5 patients (50%) achieving MM-5.
Conclusion: This study summarized the treatment patterns and outcomes of patients with MuSK-MG in real-world clinical settings. As patients who received rituximab demonstrated a favorable prognosis, early administration of rituximab may be an effective treatment strategy for MuSK-MG, potentially reducing the adverse effects associated with long-term use of CSs and NSISs. Rituximab should be considered early in MuSK-MG patients who show insufficient response to initial immunosuppressive treatment.

Keywords: Muscle-Specific Tyrosine Kinase Antibody-Positive Myasthenia Gravis; Immunosuppressive Agents; Rituximab

INTRODUCTION

Since the discovery of autoantibodies against muscle-specific tyrosine kinase (MuSK-Ab) in acetylcholine receptor antibody (AChR-Ab) negative myasthenia gravis (MG) patients in 2001, it has been recognized that muscle-specific tyrosine kinase antibody-positive

Disclosure

The authors have no potential conflicts of interest to disclose.

Author Contributions

Conceptualization: Seo D, Chung HY, Kim SW, Shin HY. Data curation: Seo D, Kim SW, Shin HY. Formal analysis: Seo D, Kim SW, Shin HY. Funding acquisition: Shin HY. Investigation: Seo D. Methodology: Seo D, Shin HY. Project administration: Shin HY. Resources: Kim SW, Shin HY. Supervision: Kim KH, Kim SW, Shin HY. Visualization: Seo D. Writing - original draft: Seo D, Shin HY. Writing - review & editing: Kim KH, Kim SW, Shin HY.

myasthenia gravis (MuSK-MG) presents with characteristic clinical features and treatment responses distinct from acetylcholine receptor antibody-positive myasthenia gravis (AChR-MG).[1-3] MuSK-Ab are detected in approximately 10% of MG patients and up to 70% of AChR-Ab negative generalized MG patients.[4-6] Patients with MuSK-MG are likely to have an acute or subacute onset with rapid progression, early ocular involvement, and common involvement of bulbar or neck muscles which are often severely affected throughout the disease course.[7-12] Additionally, in electrophysiological tests, impaired neuromuscular transmission is more commonly observed in the facial muscles than in the limb muscles.[13] In MuSK-MG, myasthenic crises are frequent and tend to occur early in the disease.[9,10,14] A significant portion of MuSK-MG patients are refractory to conventional therapeutic agents, including acetylcholinesterase inhibitors (AChEIs), corticosteroids (CSs), and non-steroidal immunosuppressant (NSIS) agents, and require a prolonged period of high-dose immunosuppressive therapy, leading to a substantial burden of long-term adverse effects.[7,15,16] Moreover, quality of life (QoL) is reduced in MuSK-MG patients and may require more social support than AChR-MG, as they present more severe phenotypes during the disease course.[17]

Rituximab, a chimeric monoclonal antibody targeting CD20 and inducing B-cell depletion, has been shown to be effective in treating MuSK-MG and to significantly reduce MuSK-Ab titers.[18-20] Guidelines recommend that rituximab should be considered as an early treatment option for MuSK-MG patients with an unsatisfactory response to initial immunotherapy.[21-24] However, in Korea, rituximab is prescribed off-label for MuSK-MG patients on a limited basis in certain circumstances since there is no regulatory approval for rituximab use in MuSK-MG. In addition to rituximab, novel therapeutic modalities, including neonatal Fc receptor inhibitors and MuSK chimeric autoantibody receptor T cells, have shown promising evidence of effectiveness in MuSK-MG patients.[25-28] These new therapies are expected to be available in Korea soon and improve MuSK-MG patients' outcomes and QoL. Therefore, understanding current MuSK-MG treatment patterns is essential for the successful implementation of upcoming therapies. However, there is limited knowledge regarding the real-world treatment status of MuSK-MG patients in clinical practice in Korea.

To explore the overall treatment patterns and outcomes of MuSK-MG, we retrospectively analyzed the clinical data of patients with MuSK-MG at a Korean MG center.

METHODS

Patient enrollment and data collection

This study is a retrospective, single-center study. We enrolled patients with MuSK-MG who visited the Department of Neurology at Severance Hospital, Seoul, Republic of Korea (South Korea), between January 1, 2005, and December 31, 2024, and had been followed for over one year. The diagnosis of MuSK-MG was based on clinical symptoms, electro-neurophysiological study results, and a positivity for anti-MuSK antibody in the patient's serum. The anti-MuSK-Ab was analyzed using a commercially available radioimmunoassay (anti-MuSK-Ab, Seoul Clinical Laboratories, Seoul, Korea), with the result considered negative when the Ab level was ≤ 0.02 nmol/L. Patients were excluded if 1) their medical records were missing key information, such as prescribed medications or data on therapeutic response, or 2) they were participating in clinical trials.

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For data analysis, we collected clinical information, including sex, age at onset and diagnosis, as well as clinical characteristics related to symptoms. The Myasthenia Gravis Foundation of America (MGFA) clinical classification was used to assess the clinical severity of MG at onset and the point of the worst disease severity. The MGFA clinical classification ranges from class I (ocular weakness only) to class V (state of intubation).

To identify treatment patterns, we obtained prescription data of CSs, NSISs, and rituximab. Azathioprine, tacrolimus, cyclosporine, and mycophenolate mofetil were classified as NSISs. In this study, rituximab was considered a treatment option for patients with MG who did not have a satisfactory response to initial immunotherapy, including CSs and immunosuppressants, or for whom such therapy could not be used due to adverse effects. Rituximab was administered weekly at a dose of 375 mg/m^2 body surface area or 500 mg for four consecutive weeks initially. After the rituximab therapy, peripheral blood CD19-positive B-cell levels were monitored. Re-treatment with rituximab was decided based on clinical evaluation and peripheral blood CD19-positive B-cell level.

To evaluate treatment response and prognosis, we gathered Myasthenia Gravis Activities of Daily Living (MG-ADL) scores and side effect reports. Additionally, other MG-related treatments, including pyridostigmine bromide, steroid pulse therapy, intravenous immunoglobulin, and plasma exchange therapy were assessed.

Study design

Treatment patterns of immunotherapies, specifically prednisolone, NSISs, and rituximab, were primarily analyzed. The sequence of applied treatment regimens was evaluated until the patient achieved minimal manifestation (MM)-5 status, which is defined as a state of MM or better according to the Myasthenia Gravis Foundation of America Post-Intervention Status (MGFA-PIS) criteria and maintaining this status with prednisolone at a dose of 5 mg/day or below for more than six months.[29] The treatment sequences of individual patients were visualized using a Sankey diagram.

Statistical analysis

Quantitative variables with a normal distribution were presented as the mean and standard deviation. Otherwise, quantitative data were expressed as the median and interquartile range. Categorical variables were summarized as numbers and percentages. All statistical analyses were performed using Python version 3.12.3 (Python Software Foundation, Wilmington, DE, USA) and visualized with the Plotly package.

Ethics statement

The Severance Hospital Institutional Review Board approved this study (approval No. 4-2025-0127), and the study has been conducted in accordance with the Declaration of Helsinki. The requirement for informed consent was waived due to the retrospective nature of the study and the use of anonymized data.

RESULTS

Demographic features of MuSK-MG patients

A total of 78 MuSK-MG patients were enrolled, and 30 patients met the inclusion criteria (**Fig. 1**).

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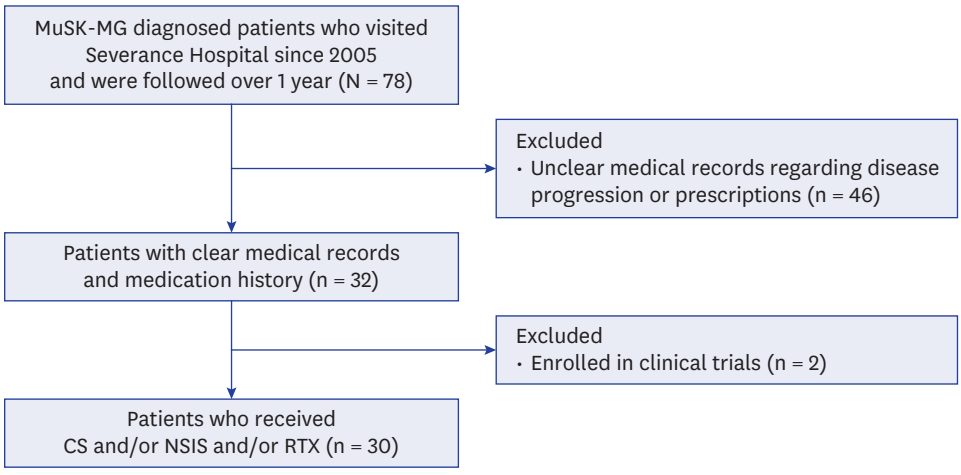

Fig. 1. Inclusion and exclusion criteria of the study population.
MuSK-MG = muscle-specific tyrosine kinase antibody-positive myasthenia gravis, CS = corticosteroid, NSIS = non-steroidal immunosuppressants, RTX = rituximab.

The basic characteristics of MuSK-MG patients are presented in **Table 1.** The mean age at diagnosis was 43.9 ± 16.0 years, and the mean age at onset was 42.3 ± 16.2 years. The study population showed a female predominance (28/30, 93.3%). Patients initially presented with symptoms of muscle weakness affecting the ocular (20/30, 66.7%), bulbar (11/30, 36.7%), and limb (4/30, 13.3%) muscles. At the time of diagnosis, 16 patients (46.7%) were classified as MGFA grade IIb, followed by grade IIa (5 patients, 16.6%). At nadir, 6 patients (20.0%) experienced a myasthenic crisis (MGFA grade V), 1 patient (3.3%) was classified as MGFA grade IV, and 10 patients (33.3%) were classified as MGFA grade III. The median follow-up period was 5.2 years, ranging from 1.0 to 19.0 years.

Table 1. General characteristics of muscle-specific tyrosine kinase antibody-positive myasthenia gravis patients

Variables	Total (N = 30)
Diagnosis age, yr	43.9 ± 16.0
Sex	
Male	2 (6.7)
Female	28 (93.3)
Onset age, yr	42.3 ± 16.2
Onset symptom	
Ocular	20 (66.7)
Bulbar	11 (36.7)
Limb	4 (1.3)
MGFA clinical classification at diagnosis	
I	6 (20.0)
IIa/IIb	5 (16.7)/14 (46.7)
IIIa/IIIb	1 (3.3)/4 (13.3)
IV	0 (0.0)
V	0 (0.0)
MGFA clinical classification at nadir	
I	0 (0.0)
IIa/IIb	0 (0.0)/13 (43.3)
IIIa/IIIb	2 (6.7)/8 (26.7)
IV	1 (3.3)
V	6 (20.0)
Thymectomy	1 (3.3)
Follow-up period, yr	5.2 (3.4–15.8)

Values are presented as number (%), mean ± standard deviation, or median (interquartile range).
MGFA = Myasthenia Gravis Foundation of America.

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Treatment status and regimen prescription patterns

Pyridostigmine bromide, an AChEI, was prescribed to 23 patients after MG diagnosis, and among them, adverse effects such as muscle twitching, cramps, and weakness were reported in 11 patients (47.8%). None of the patients continued to use AChEI.

The treatments of MuSK-MG patients with CSs, NSISs, and rituximab are presented in **Table 2**. All patients were initially treated with prednisolone. NSISs were prescribed to 24 (80.0%) patients. Tacrolimus (17/24, 70.8%) was the most prescribed regimen, followed by azathioprine (15/24, 62.5%), cyclosporine (5/24, 20.8%), and mycophenolate mofetil (2/24, 8.3%). The maintenance dose for tacrolimus was 3 mg/day in most tacrolimus-treated patients (11/17, 64.7%), and the maintenance dose for azathioprine was 2–3 mg/kg/day in more than half of azathioprine-treated patients (9/15, 60.0%).

Ten patients (33.3%) received rituximab therapy. Eight patients initially received 375 mg/m^2 for induction, and two patients initially received 500 mg (**Table 2**, **Supplementary Fig. 1**). The re-treatment was performed 13 times in seven patients, with doses ranging from a single administration of 375 mg/m^2 to four weekly doses of 375 mg/m^2. The median interval between rituximab courses was 12 months, with a range of 3 to 75 months. Except for one three-month interval, all intervals were nine months or longer.

Numbers of NSISs prescribed for each treatment pattern are summarized in **Table 3**. Among patients who initiated rituximab therapy, 5 patients (50.0%) started after being treated with one type of NSISs, and the others started after two or more types of NSISs. The follow-up period after rituximab ranged from 12 weeks to 8 years.

Table 2. Treatments with CSs, NSISs, and RTX

Treatments	Total (N = 30)
CSs	30 (100.0)
NSISs	24 (80.0)
Tacrolimus	17
Maintenance dose	
2 mg/day	1
3 mg/day	11
4 mg/day	5
Azathioprine	15
Maintenance dose	
< 2 mg/kg/day	6
2–3 mg/kg/day	9
Cyclosporine	5
Maintenance dose	
< 3 mg/kg/day	0
3–6 mg/kg/day	5
Mycophenolate mofetil	
Maintenance dose	
1,000 mg bid	1
1,500 mg bid	1
RTX	10 (33.3)
Initial dosing	
375 mg/m^2 weekly, 4 times	8
500 mg weekly, 4 times	2
Repeated courses[a]	7
Interval, mon	12 (3–75)

Values are presented as number (%) or median (range).
CS = corticosteroid, NSIS = non-steroidal immunosuppressants, RTX = rituximab.
[a]Re-treatment with RTX was decided based on clinical evaluation and CD19 level.

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Table 3. Treatment patterns of muscle-specific tyrosine kinase antibody-positive myasthenia gravis patients

Treatment patterns	Total (N = 30)
Corticosteroid only	6 (20.0)
Corticosteroid + NSISs	14 (46.7)
Applied 1 NSIS	8
Applied 2 NSISs	5
Applied 3 NSISs	1
Applied 4 NSISs	0
Corticosteroid + NSISs + RTX	10 (33.3)
Applied 1 NSIS before RTX	5
Applied 2 NSISs before RTX	3
Applied 3 NSISs before RTX	1
Applied 4 NSISs before RTX	1

Values are presented as number (%).
NSISs = non-steroidal immunosuppressants, RTX = rituximab.

Treatment response and outcome

The treatment patterns and outcome of MuSK-MG patients are demonstrated in **Fig. 2**. Each flow represents the treatment course of individual MuSK-MG patients and their MM-5 achievement. After the initial treatment with prednisolone, 6 (20%) had achieved MM-5 and one was well controlled using prednisolone 5 mg/day but was followed-up shortly. NSIS were additionally prescribed for 23 patients for CS sparing. Of 6 patients who achieved MM-5, one patient experienced MG exacerbation. The patient was added to tacrolimus and rituximab therapy after the exacerbation.

The most common first NSIS was azathioprine (15/23, 65.2%), followed by tacrolimus (7/23, 30.4%) and cyclosporine (1/23, 3.3%). Using the first NSIS, 5 (5/23, 21.7%) patients achieved MM-5, and 3 patients were well controlled using ongoing regimen. The other 15 patients switched to another treatments. Second NSIS was prescribed to 11 patients, and rituximab was initiated to the other 4 patients. Tacrolimus (7/11, 63.6%) was the most frequently prescribed second NSIS, followed by cyclosporine (4/11, 36.4%). Using the second NSIS, 4 (4/11, 36.4%) patients achieved MM-5, and one patient was controlled with the ongoing regimen. Three patients switched to third NSIS, and another 3 were treated with rituximab. Using the third NSISs, none reached MM-5 and needed another treatment including rituximab and additional NSISs.

All the 10 patients treated with rituximab responded to the treatment and 5 (50%) patients reached MM-5. During rituximab therapy, no other therapies were added in all patients expect one patient who was unable to continue rituximab treatment for financial reasons. This patient was in MM-5 status for a year after the first rituximab therapy, but symptoms relapsed. Despite increasing CS dose and adding azathioprine to treatment, symptoms were not controlled. The patient regained MM-5 status after an additional course of rituximab therapy. Among five patients who initiated rituximab treatment after one NSIS, three (60%) achieved MM-5. Of the five patients who began rituximab treatment after two or more NSIS, two (40%) reached MM-5.

A total of 10 myasthenic crisis episodes occurred in 5 patients, with 7 crisis episodes occurred while treated with prednisolone alone and 3 crisis episodes occurred while receiving combination of prednisolone and tacrolimus. No myasthenic crisis occurred after initiating rituximab therapy.

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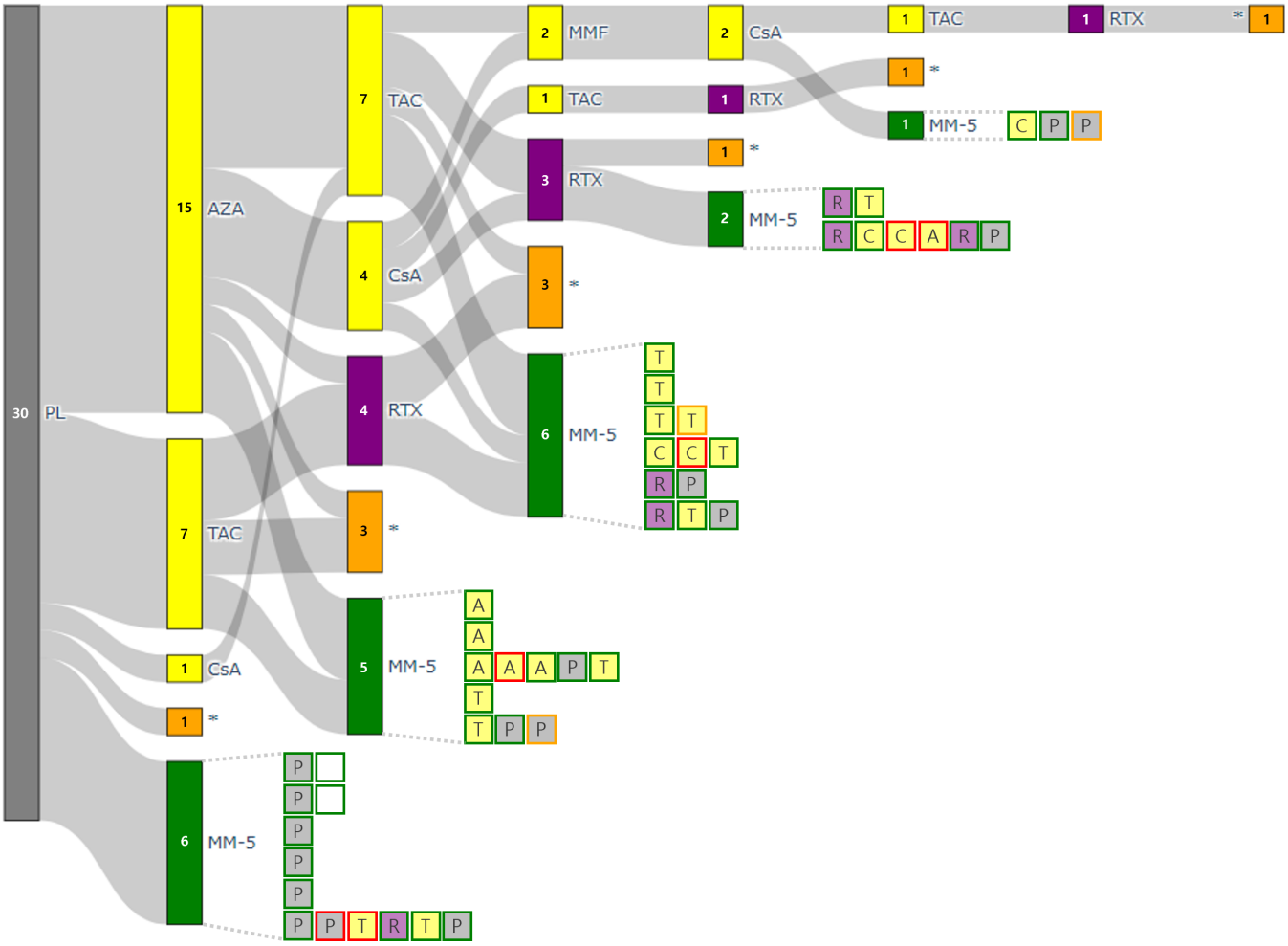

Fig. 2. Sankey diagram representing the order of treatment regimens in muscle-specific tyrosine kinase antibody-positive myasthenia gravis patients. The flow from left to right represents an individual patient receiving a specific regimen and the subsequent regimen. Flows that end in orange blocks (*) indicate that the patient is responding to the treatment but has not yet achieved MM-5. Flows that end in green blocks (MM-5) indicate that the patient has achieved MM-5 status. Smaller blocks after green blocks (MM-5) indicate the treatment regimen at the time of achieving MM-5 and thereafter. The blank block indicates that the patient is not receiving any treatment. Green-outlined smaller blocks indicate that the patient is maintaining or has regained MM-5 status with the regimen. Orange-outlined smaller blocks indicate that the patient is responding to the treatment but has not yet achieved MM-5. Red-outlined smaller blocks indicate that symptoms aggravated or relapsed while being treated with the regimen. When patients are being treated with non-steroidal immunosuppressants, they are being treated in combination with steroids.
MM = minimal manifestation, PL = prednisolone, AZA = azathioprine, TAC = tacrolimus, CsA = cyclosporine, MMF = mycophenolate, RTX = rituximab, P = prednisolone, T = tacrolimus, R = rituximab, A = azathioprine, C = cyclosporine.

The MGFA-PIS classification at last visit and treatment regimens are summarized in **Table 4**. On the last visit, one patient (3.3%) was in complete stable remission, 8 patients (26.7%) were in pharmacologic remission, and 14 patients (46.7%) were in MM status.

DISCUSSION

In this single-center retrospective study, patient with MuSK-MG had female predominance and significant portion of patients presenting ocular or bulbar muscle weakness at onset, which aligns with the characteristic features of MuSK-MG.[30] All patients were initially treated with prednisolone, and one-fifth of them reached MM-5. Tacrolimus was the most common NSIS, and azathioprine was the most common first NSIS. Without rituximab therapy, 39.1%

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Table 4. MG-post-intervention status classification and treatment regimen at last visit

Treatment regimens	Total (N = 30)
CSR	1 (3.3)
PR	8 (26.7)
None	1
PL only	1
PL + RTX	2
PL + TAC	2
PL + AZA	2
MMs	14 (46.7)
PL only	4
PL + RTX	3
PL + TAC	4
PL + TAC + RTX	3
Others	7 (23.3)
PL + TAC	3
PL + AZA	2
PL + TAC + RTX	1
PL + AZA + RTX	1
Death	0 (0.0)

Values are presented as number (%).
"Others" category refers to patients who exhibited functional limitations related to MG, but did not meet the criteria for CSR, PR, MM, or death.
MG = myasthenia gravis, CSR = complete stable remission, PR = pharmacologic remission, PL = prednisolone, RTX = rituximab, TAC = tacrolimus, AZA = azathioprine, MM = minimal manifestation.

of patients who did not achieve MM-5 with CS were able to reach MM-5 with one or two NSISs. Rituximab was administered to one-third of the enrolled patients; half of patients under rituximab therapy achieved MM-5, and the other also showed favorable response. After initiating rituximab therapy, there were no myasthenic crisis episodes reported. On the last visit, 76.7% of the enrolled patients were in MM status or better.

Several retrospective studies have demonstrated the long-term effects of rituximab in MG, with even greater benefits in MuSK-MG.[19,31-33] According to a recent meta-analysis study, rituximab therapy has been shown to contribute to MuSK-MG patients achieving MM or better status.[34] A randomized clinical trial among generalized MG patients who were mostly positive for AChR-Ab reported that the patient group which received a single dose of 500 mg rituximab had a greater portion of minimal MG manifestations.[35] Similarly, MuSK-MG patients who received 500 mg of rituximab every 6 months had decreased prednisolone dose and improved MG-ADL and Quantitative Myasthenia Gravis (QMG) score.[36] In our study, all patients who initiated rituximab therapy showed stable symptom control with reduced CS doses. Among 5 patients who did not achieve MM-5, 3 patients were stably reducing steroid doses, but did not reach MM-5 due to a short follow-up period of less than one year since starting rituximab. These 3 patients might have reached MM-5 if the follow-up period was longer. Given the high frequency of myasthenic crisis in patients with MuSK-MG, early initiation of rituximab therapy should be considered among patients who do not respond to treatment with CS and/or NSIS. In this study, nearly half of the patients did not reach MM-5 with CS and NSIS, and these patients could have benefited from early rituximab therapy.

Rituximab has also been shown to resolve symptoms and allow steroid discontinuation in refractory patients.[37,38] One patient from this study, who was severely refractory with multiple episodes of crisis and requires plasma exchange as maintenance therapy, has not experienced MG crisis episodes since receiving rituximab for 4.6 years. After initiating

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therapy, this patient's symptoms are stably controlled which implies that rituximab may also benefit refractory patients and stabilize the disease course.

Azathioprine is generally recommended as the first-line NSIS for MG patients.[21-23] However, a retrospective cohort study reported that 3 out of 5 MuSK-MG patients under tacrolimus achieved a QMG score of 0, while all 5 MuSK-MG patients treated with azathioprine failed to achieve satisfactory outcomes.[39] In addition, another study demonstrated that MuSK-MG patients had reduced steroid dose and improvement in QMG and MG-ADL scores after receiving 4 weeks of tacrolimus.[31] According to the Sankey diagram, the portion of patients who switched to different NSIS or started rituximab therapy was higher in azathioprine (11/15, 73.3%) than tacrolimus (9/17, 52.9%). Tacrolimus is also preferred over cyclosporine due to its lower nephrotoxicity, and mycophenolate mofetil and methotrexate are scarcely prescribed due to teratogenicity risks since many patients were women of reproductive age.[40,41] Therefore, tacrolimus may be more suitable for MuSK-MG treatment than other NSIS regimens.

In our study population, initial ocular manifestations were reported more frequently than in previous reports.[7] This may be due to the small sample size or the influence of the excluded patients. The proportion of patients presenting with ocular symptoms was 66.7% in the study subjects and 48.9% in the excluded patients (data not shown), although the difference was not statistically significant. However, some studies have reported a relatively higher prevalence of ocular symptoms in MuSK-MG. A retrospective study of 82 MuSK-MG patients found that 96.4% experienced ocular symptoms, and 58.5% presented with them.[42] Another case series study also reported a notably high prevalence of ocular symptoms at onset.[43] These findings suggest that the prevalence of ocular muscle involvement in MuSK-MG may be higher than traditionally thought and warrants further investigation.

The main limitations of this study are that this is a retrospective study, and the study population was small. However, to the best of our knowledge, the sample size is relatively large when compared with other MuSK-MG studies. Since the treatment patterns were highly diverse, it was difficult to directly compare between different treatment patterns and determine the optimal treatment strategy for MuSK-MG patients. Multi-center or clinical trial studies regarding the effectiveness of early initiation of rituximab therapy among MuSK-MG patients will be necessary. Although the median follow-up period for all study participants was 5.2 years, the follow-up period after rituximab ranged from 12 weeks to 8 years. Some patients treated with rituximab had a short follow-up period, which may have led to an underestimation of its therapeutic effect. Longitudinal studies with extended follow-up durations are necessary to more comprehensively assess the long-term efficacy of rituximab.

In conclusion, this study summarized the characteristics and overall treatment patterns of MuSK-MG patients in a large single center. Approximately, half of the patients achieved MM-5 without receiving rituximab therapy. One-third of the patients were undergoing rituximab therapy and all of them showed favorable response. Patients undergoing rituximab therapy had stable symptom control without the need to change or add to their ongoing immunosuppression treatment. Therefore, early initiation of rituximab therapy should be recommended to patients who do not respond to initial CS and NSIS treatment in terms of reducing the steroid burden.

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SUPPLEMENTARY MATERIAL

Supplementary Fig. 1

Rituximab dosing, number of administrations, and treatment schedule in 10 patients.

REFERENCES

1. Hoch W, McConville J, Helms S, Newsom-Davis J, Melms A, Vincent A. Auto-antibodies to the receptor tyrosine kinase MuSK in patients with myasthenia gravis without acetylcholine receptor antibodies. *Nat Med* 2001;7(3):365-8. PUBMED | CROSSREF
2. Gilhus NE, Tzartos S, Evoli A, Palace J, Burns TM, Verschuuren JJGM. Myasthenia gravis. *Nat Rev Dis Primers* 2019;5(1):30. PUBMED | CROSSREF
3. Gilhus NE. Myasthenia gravis. *N Engl J Med* 2016;375(26):2570-81. PUBMED | CROSSREF
4. Gilhus NE, Skeie GO, Romi F, Lazaridis K, Zisimopoulou P, Tzartos S. Myasthenia gravis - autoantibody characteristics and their implications for therapy. *Nat Rev Neurol* 2016;12(5):259-68. PUBMED | CROSSREF
5. Kim MJ, Kim SW, Kim M, Choi YC, Kim SM, Shin HY. Evaluating an in-house cell-based assay for detecting antibodies against muscle-specific tyrosine kinase in myasthenia gravis. *J Clin Neurol* 2021;17(3):400-8. PUBMED | CROSSREF
6. Zisimopoulou P, Brenner T, Trakas N, Tzartos SJ. Serological diagnostics in myasthenia gravis based on novel assays and recently identified antigens. *Autoimmun Rev* 2013;12(9):924-30. PUBMED | CROSSREF
7. Guptill JT, Sanders DB, Evoli A. Anti-MuSK antibody myasthenia gravis: clinical findings and response to treatment in two large cohorts. *Muscle Nerve* 2011;44(1):36-40. PUBMED | CROSSREF
8. Sanders DB, El-Salem K, Massey JM, McConville J, Vincent A. Clinical aspects of MuSK antibody positive seronegative MG. *Neurology* 2003;60(12):1978-80. PUBMED | CROSSREF
9. Evoli A, Tonali PA, Padua L, Monaco ML, Scuderi F, Batocchi AP, et al. Clinical correlates with anti-MuSK antibodies in generalized seronegative myasthenia gravis. *Brain* 2003;126(10):2304-11. PUBMED | CROSSREF
10. Pasnoor M, Wolfe GI, Nations S, Trivedi J, Barohn RJ, Herbelin L, et al. Clinical findings in MuSK-antibody positive myasthenia gravis: a U.S. experience. *Muscle Nerve* 2010;41(3):370-4. PUBMED | CROSSREF
11. Keritam O, Vincent A, Zimprich F, Cetin H. A clinical perspective on muscle specific kinase antibody positive myasthenia gravis. *Front Immunol* 2024;15:1502480. PUBMED | CROSSREF
12. Rodolico C, Bonanno C, Toscano A, Vita G. MuSK-associated myasthenia gravis: clinical features and management. *Front Neurol* 2020;11:660. PUBMED | CROSSREF
13. Kim SW, Sunwoo MK, Kim SM, Shin HY, Sunwoo IN. Repetitive nerve stimulation in MuSK-antibody-positive myasthenia gravis. *J Clin Neurol* 2017;13(3):287-92. PUBMED | CROSSREF
14. Zhou Y, Chen J, Li Z, Tan S, Yan C, Luo S, et al. Clinical features of myasthenia gravis with antibodies to MuSK based on age at onset: a multicenter retrospective study in China. *Front Neurol* 2022;13:879261. PUBMED | CROSSREF
15. Cortés-Vicente E, Álvarez-Velasco R, Pla-Junca F, Rojas-Garcia R, Paradas C, Sevilla T, et al. Drug-refractory myasthenia gravis: clinical characteristics, treatments, and outcome. *Ann Clin Transl Neurol* 2022;9(2):122-31. PUBMED | CROSSREF
16. Rath J, Brunner I, Tomschik M, Zulehner G, Hilger E, Krenn M, et al. Frequency and clinical features of treatment-refractory myasthenia gravis. *J Neurol* 2020;267(4):1004-11. PUBMED | CROSSREF
17. Stankovic M, Peric S, Stojiljkovic Tamas O, Stankovic T, Nikolic A, Lavrnic D, et al. Quality of life in patients with MuSK positive myasthenia gravis. *Acta Neurol Belg* 2018;118(3):423-7. PUBMED | CROSSREF
18. Yi JS, Guptill JT, Stathopoulos P, Nowak RJ, O'Connor KC. B cells in the pathophysiology of myasthenia gravis. *Muscle Nerve* 2018;57(2):172-84. PUBMED | CROSSREF
19. Díaz-Manera J, Martínez-Hernández E, Querol L, Klooster R, Rojas-García R, Suárez-Calvet X, et al. Long-lasting treatment effect of rituximab in MuSK myasthenia. *Neurology* 2012;78(3):189-93. PUBMED | CROSSREF
20. Iorio R, Damato V, Alboini PE, Evoli A. Efficacy and safety of rituximab for myasthenia gravis: a systematic review and meta-analysis. *J Neurol* 2015;262(5):1115-9. PUBMED | CROSSREF
21. Wiendl H, Abicht A, Chan A, Della Marina A, Hagenacker T, Hekmat K, et al. Guideline for the management of myasthenic syndromes. *Ther Adv Neurol Disord* 2023;16:17562864231213240. PUBMED | CROSSREF

22. Sanders DB, Wolfe GI, Benatar M, Evoli A, Gilhus NE, Illa I, et al. International consensus guidance for management of myasthenia gravis: executive summary. *Neurology* 2016;87(4):419-25. PUBMED | CROSSREF
23. Evoli A, Antonini G, Antozzi C, DiMuzio A, Habetswallner F, Iani C, et al. Italian recommendations for the diagnosis and treatment of myasthenia gravis. *Neurol Sci* 2019;40(6):1111-24. PUBMED | CROSSREF
24. Narayanaswami P, Sanders DB, Wolfe G, Benatar M, Cea G, Evoli A, et al. International consensus guidance for management of myasthenia gravis: 2020 update. *Neurology* 2021;96(3):114-22. PUBMED | CROSSREF
25. Howard JF Jr, Bril V, Vu T, Karam C, Peric S, Margania T, et al. Safety, efficacy, and tolerability of efgartigimod in patients with generalised myasthenia gravis (ADAPT): a multicentre, randomised, placebo-controlled, phase 3 trial. *Lancet Neurol* 2021;20(7):526-36. PUBMED | CROSSREF
26. Bril V, Drużdż A, Grosskreutz J, Habib AA, Mantegazza R, Sacconi S, et al. Safety and efficacy of rozanolixizumab in patients with generalised myasthenia gravis (MycarinG): a randomised, double-blind, placebo-controlled, adaptive phase 3 study. *Lancet Neurol* 2023;22(5):383-94. PUBMED | CROSSREF
27. Antozzi C, Vu T, Ramchandren S, Nowak RJ, Farmakidis C, Bril V, et al. Safety and efficacy of nipocalimab in adults with generalised myasthenia gravis (Vivacity-MG3): a phase 3, randomised, double-blind, placebo-controlled study. *Lancet Neurol* 2025;24(2):105-16. PUBMED | CROSSREF
28. Oh S, Mao X, Manfredo-Vieira S, Lee J, Patel D, Choi EJ, et al. Precision targeting of autoantigen-specific B cells in muscle-specific tyrosine kinase myasthenia gravis with chimeric autoantibody receptor T cells. *Nat Biotechnol* 2023;41(9):1229-38. PUBMED | CROSSREF
29. Murai H, Utsugisawa K, Motomura M, Imai T, Uzawa A, Suzuki S. The Japanese clinical guidelines 2022 for myasthenia gravis and Lambert–Eaton myasthenic syndrome. *Clin Exp Neuroimmunol* 2023;14(1):19-27. CROSSREF
30. Borges LS, Richman DP. Muscle-specific kinase myasthenia gravis. *Front Immunol* 2020;11:707. PUBMED | CROSSREF
31. Bi Z, Li Y, Lin J, Gui M, Li Z, Bu B. Long-term efficacy and safety of tacrolimus in anti-MuSK antibody-positive myasthenia gravis: a retrospective single-center cohort study. *Neurol Sci* 2025;46(2):943-9. PUBMED | CROSSREF
32. Zhao C, Pu M, Chen D, Shi J, Li Z, Guo J, et al. Effectiveness and safety of rituximab for refractory myasthenia gravis: a systematic review and single-arm meta-analysis. *Front Neurol* 2021;12:736190. PUBMED | CROSSREF
33. Inan B, Orhan IG, Bekircan-Kurt CE, Erdem-Ozdamar S, Tan E. Clinical and laboratory remission with rituximab in anti-MuSK-positive myasthenia gravis. *Ir J Med Sci* 2024;193(6):2989-94. PUBMED | CROSSREF
34. Chayanopparat S, Banyatcharoen P, Jitprapaikulsan J, Uawithya E, Apiraksattayakul N, Viarasilpa V. Efficacy and safety of rituximab in anti-MuSK myasthenia gravis: a systematic review and meta-analysis. *Sci Rep* 2025;15(1):7219. PUBMED | CROSSREF
35. Piehl F, Eriksson-Dufva A, Budzianowska A, Feresiadou A, Hansson W, Hietala MA, et al. Efficacy and safety of rituximab for new-onset generalized myasthenia gravis: the RINOMAX Randomized Clinical Trial. *JAMA Neurol* 2022;79(11):1105-12. PUBMED | CROSSREF
36. Yang X, Zhang W, Chang X, Li Z, Du R, Guo J. Promising efficacy of low-dose rituximab in muscle specific kinase antibody positive myasthenia gravis. *Neurosci Lett* 2024;818:137561. PUBMED | CROSSREF
37. Heckmann JM. A single low-dose rituximab infusion in severe chronic refractory myasthenia gravis in resource-limited settings. *J Neurol Sci* 2022;442:120394. PUBMED | CROSSREF
38. Zebardast N, Patwa HS, Novella SP, Goldstein JM. Rituximab in the management of refractory myasthenia gravis. *Muscle Nerve* 2010;41(3):375-8. PUBMED | CROSSREF
39. Zhao S, Zhang K, Ren K, Lu J, Ma C, Zhao C, et al. Clinical features, treatment and prognosis of MuSK antibody-associated myasthenia gravis in Northwest China: a single-centre retrospective cohort study. *BMC Neurol* 2021;21(1):428. PUBMED | CROSSREF
40. Narayanaswami P, Sanders DB, Thomas L, Thibault D, Blevins J, Desai R, et al. Comparative effectiveness of azathioprine and mycophenolate mofetil for myasthenia gravis (PROMISE-MG): a prospective cohort study. *Lancet Neurol* 2024;23(3):267-76. PUBMED | CROSSREF
41. Nagane Y, Utsugisawa K, Obara D, Kondoh R, Terayama Y. Efficacy of low-dose FK506 in the treatment of myasthenia gravis - a randomized pilot study. *Eur Neurol* 2005;53(3):146-50. PUBMED | CROSSREF
42. Evoli A, Alboini PE, Iorio R, Damato V, Bartoccioni E. Pattern of ocular involvement in myasthenia gravis with MuSK antibodies. *J Neurol Neurosurg Psychiatry* 2017;88(9):761-3. PUBMED | CROSSREF
43. Skjei KL, Lennon VA, Kuntz NL. Muscle specific kinase autoimmune myasthenia gravis in children: a case series. *Neuromuscul Disord* 2013;23(11):874-82. PUBMED | CROSSREF