Patient Portrayal

PATIENT CASE STUDIES

IS YOUR PATIENT'S gMG UNDER CONTROL?

The following pages include hypothetical case studies that show how consistent MG-ADL score monitoring and patient symptom logging may help identify characteristics of uncontrolled generalized myasthenia gravis (gMG).

MG-ADL=Myasthenia Gravis Activities of Daily Living.

MICHELLE

44-year-old married mother of 3 children; works as a nurse

Summary of medical history

- MGFA Class IIIa at diagnosis (AChR–Ab+)
- Multiple changes to drug therapy to manage gMG; currently managed with pyridostigmine (120 mg TID) and cyclosporine (5 mg/kg/day)
- Other medical history: leukopenia and recurrent respiratory infections; comorbid for depression and anxiety managed with sertraline (50 mg QD)

Three factors to consider in this case:

- Consistently high MG–ADL scores
- Debilitating symptoms that continually impact professional life and physical health
- Ongoing exacerbations

2017

Clinical assessments/findings

Presenting symptoms upon referral to neurology

Progressive limb weakness, fatigable bilateral ptosis, dysphagia, and exertional dyspnea

Recently quit volunteering at an animal shelter due to progressive weakness upon exertion

Diagnosis

gMG (MGFA Class IIIa)
MG–ADL: 12

Laboratory/radiological findings

Findings

AChR–Ab+

Therapeutic management/changes

Initial management

Pyridostigmine (60 mg TID) and prednisone (10 mg QD)

Referred for surgical resection of the thymus gland via open thymectomy

Clinical assessments/findings

Neurology visit 1 month later

Mild improvements to symptoms, but patient still presents with limb weakness (MMT3 bilateral), dysphagia, and exertional dyspnea

MG–ADL: 11

Therapeutic management/changes

Treatment change

Pyridostigmine titrated to maximum dose (120 mg TID); prednisone titrated to 30 mg QD

Thymectomy conducted

AChR–Ab+=acetylcholine receptor antibody–positive; BID=two times a day; MGFA=Myasthenia Gravis Foundation of America; MMT3=Manual Muscle Testing Grade 3; QD=one time a day; TID=three times a day.

2018

Clinical assessments/findings

PCP routine visit

General fatigue at work (floor nurse) required patient to transition to a stationary occupation

Patient reported feeling down and less social, related to increased weight (+3.6 kg since diagnosis)

MG-ADL: not recorded

Laboratory/radiological findings

DEXA scan conducted due to risk of osteoporosis under chronic high-dose prednisone therapy

(T-score: -2.4)

Therapeutic management/changes

Treatment change

Initiated azathioprine (50 mg QD, then titrated to BID) based on continuing symptoms; prednisone titrated down and discontinued due to risk of AEs (osteoporosis and weight gain present)

2019

Clinical assessments/findings

Exacerbation

Patient presented to the ED due to worsening dysphagia, dyspnea, and cough, and was admitted to the hospital

Febrile (38.4 °C/101.1 °F)

MG-ADL: not recorded

Laboratory/radiological findings

Workup for suspected chest infection

Chest CT and microbial testing confirmed community-acquired pneumonia

Therapeutic management/changes

Treatment

IVIg + prednisone (60 mg IV QD) for management of impending myasthenic crisis (2 g/kg over 3 days); background therapy was maintained

Amoxicillin (1 g TID) + azithromycin (500 mg first day, then 250 mg daily) for community-acquired pneumonia

2020

Clinical assessments/findings

Neurology visit

Patient noted she had recurrent lower respiratory tract infections that were treated empirically over 24 months; patient discussed recent anxiety and depressive thoughts. Patient also noted missing 1-2 days of work every month

MG-ADL: 9
MGFA Class IIIa

Laboratory/radiological findings

Findings

Leukopenia

Therapeutic management/changes

Treatment change

Transition from azathioprine to cyclosporine (2.5 mg/kg/day titrated to 5 mg/kg/day) using an IVIg bridge, due to leukopenia and recurrent infections suspected to be related to azathioprine

AEs=adverse events; CT=computed tomography; DEXA=dual-energy X-ray absorptiometry; IVIg=intravenous immunoglobulin.

Clinical assessments/findings

Psychiatry visit

Patient self-presented to a psychiatry clinic at the recommendation of her husband

Reported poor sleep, fatigue, depressive symptoms, and worry over managing her symptoms in the context of her busy life (managing 3 young teenagers, job change, and social activities). She also is strongly considering changing to a part-time job, which causes additional worry about loss of income

Therapeutic management/changes

Sertraline (50 mg QD) was indicated for depression and anxiety

Clinical assessments/findings

ED visit

Presented to the ED due to acute worsening dyspnea and productive cough

MG-ADL: not recorded

Laboratory/radiological findings

Findings

FVC: 59%
MIP: -32 cm H_2O

Therapeutic management/changes

Treatment

IVIg (1 g/kg over 2 days) for a myasthenic exacerbation + prednisone (60 mg IV QD) until resolution

Follow-up visit

Respiratory symptoms resolved; generalized extremity weakness still present

Continued stable cyclosporine regimen (5 mg/kg/day)

FVC=forced vital capacity; MIP=maximal inspiratory pressure.

BILL

56-year-old widowed father of 3 adult children; works in construction

Summary of medical history

- gMG MGFA Class IIa (AChR–Ab+, diagnosed 5 years prior); managed with prednisone (5 mg QD) until 2017; mycophenolate mofetil (1500 mg BID)
- Hypertension; managed with benazepril + amlodipine
- Type 2 diabetes mellitus (DMII); managed with metformin + repaglinide

Three factors to consider in this case:

- Frequent therapy changes and escalation indicating patient's gMG is poorly controlled
- Debilitating symptoms that continually impact professional life and physical health
- Ongoing challenges with comorbidity management

2015

Clinical assessments/findings

Presenting symptoms upon referral to neurology

Mild ptosis and fluctuating diplopia, dysphagia, hoarseness, and generalized weakness (arms and legs)

Patient had to leave work early on several occasions

Diagnosis

gMG
MG–ADL: 8

Laboratory/radiological findings

Findings

AChR–Ab+

Therapeutic management/changes

Initial management

Initiated pyridostigmine (60 mg TID) and soon titrated to 120 mg TID based on poor limited improvement of bulbar manifestations and extremity weakness

Clinical assessments/findings

Neurology follow–up to MG diagnosis

Changes in MG–ADL

Has to sometimes use arms to rise from chair

gMG (Class IIIa)
MG–ADL: 9

Therapeutic management/changes

Treatment change

Titrated prednisone (20 mg QD) due to persistent bulbar symptoms and extremity weakness

2016

Clinical assessments/findings

PCP clinic visit for routine bloodwork

Patient reported difficulty getting on and off construction equipment over the course of the day during the past month

Laboratory/radiological findings

BMI: 30.5 kg/m^2
Blood glucose (fasting): 210 mg/dL
HbA1c: 10.5%
Blood pressure: 145/90 mmHg

Therapeutic management/changes

Treatment change

Added metformin for DMII and benazepril + amlodipine for hypertension

Clinical assessments/findings

Neurology referral following PCP visit

Went from sometimes to always having to use arms when rising from a chair over the last several months

Shortness of breath with exertion

MG–ADL: 11

Therapeutic management/changes

Treatment change

Mycophenolate mofetil (1500 mg QD) in 2016 as MG symptoms persisted and due to concern about prednisone–related AEs (DMII and hypertension)

Prednisone reduced to 5 mg QD

2017 – 2019

Clinical assessments/findings

Routine visits to PCP and neurology

Routine visits to PCP were generally unremarkable, although the patient routinely reported dyspnea and generalized fatigue when pressed for symptoms. Diplopia was infrequent, and the patient expressed little interest in frequent neurology visits for MG–related symptoms.

Laboratory/radiological findings

Findings (2017)

HbA1c: 9.0%
Blood pressure: 135/80 mmHg

Therapeutic management/changes

Treatment change

Prednisone discontinued in 2017 due to difficulty managing DMII and hypertension; mycophenolate mofetil continued

BMI=body mass index; HbA1c=hemoglobin A1c.

2020

Clinical assessments/findings

Neurology visit

Referred to neurology from PCP after complaint of recent dyspnea on exertion and progressive fatigue

QMG: 18

Patient noted difficulty in doing construction work related to weakness in extremities

Patient reported recent weight gain (currently 33.2 kg) and foot pain while working

MG–ADL: not recorded

Laboratory/radiological findings

Mild pancytopenia
Blood glucose (fasting): 178 mg/dL
HbA1c: 8.1%

Therapeutic management/changes

Treatment change

Follow–up visit to PCP added repaglinide (1 mg TID) to manage DMII

QMG=Quantitative Myasthenia Gravis test.

MARIA

38-year-old female of Mediterranean descent; works as a teacher

Summary of medical history

- MGFA Class IIb at diagnosis in 2017 (MuSK–Ab+)
- Multiple changes to drug therapy to manage refractory gMG
- Comorbidities: GERD, managed with lanzoprazole; hypertension managed with metroprolol; dyslipidemia managed with atorvastatin

Three factors to consider in this case:

- Distinct clinical presentation and treatments for poorly controlled MuSK–Ab+ gMG
- Debilitating symptoms that continually impact professional life and physical heath
- Ongoing exacerbations and crises

2017

Clinical assessments/findings

Presenting symptoms upon referral to neurology

Significant neck and respiratory weakness with intermittent GI symptoms

Patient had to change role at work because of slurred speech

Diagnosis

gMG
MG–ADL: 8

Class IIb

Laboratory/radiological findings

Findings

MuSK–Ab+

Repeated nerve stimulation normal

SFMEG positive (trapezius muscle)

Therapeutic management/changes

Initial management

Oral GCS QAD 10 mg taper

Azathioprine 150 mg PO QD

Clinical assessments/findings

Neurology follow–up to MG diagnosis

Presenting symptoms upon referral to neurology

Significant trouble with chewing and slurred speech with moderate weakness in arms and legs

Class IIIb

MG–ADL: 10

Chewing and slurred speech improved somewhat 1 month after starting rituximap (December)

MG–ADL: 7

Therapeutic management/changes

Treatment change

Rituximap

(2 infusions of 1 g separated by 2 weeks with planned 1–g maintenance infusions every 6 months)

2018

Clinical assessments/findings

Myasthenia crisis

Patient presented to the ED due severe restrictive pathology and difficulty swallowing in the context of a viral infection

Acute chronic respiratory failure requiring almost 24 hours of BiPAP daily

MG–ADL: 17

Class V

Follow–up visit

Infection cleared and respiratory function improved although BiPAP was occasionally needed in weeks following discharge. Bulbar symptoms persisted.

Laboratory/radiological findings

FVC: ~50% predicted

DLCO: 51% predicted

Febrile (38.4 °C/101.5 °F)

Microbial testing confirmed influenza

Therapeutic management/changes

Treatment change

IVIG⋆ (2g/kg over 2 days) + prednisone (60 mg IV QD) ⋆PLEX not available due to donor shortage

Baloxavir marboxil 40 mg for influenza

2019

Clinical assessments/findings

Unsatisfactory treatment response

Patient had to switch to a remote administrative part–time role

Patient has been seen in the office multiple times over the past 6 months due to respiratory distress and occasional choking while eating solid food

MG–ADL: 14

Class IIIB

Laboratory/radiological findings

Prominent dyspnea and use of accessory respiratory muscles; mildly slurred speech; vital capacity is low, and MIP is below 1/3 of normal (–25 cm H2O)

Therapeutic management/changes

Treatment change

4 courses of IVIG (1g/kg)

Clinical assessments/findings

Myasthenia crisis

Patient presented to the ED due severe restrictive pathology and difficulty swallowing

MG–ADL: 18

Laboratory/radiological findings

FEV1: 35% predicted

FVC: 33% predicted

DLCO: 51% predicted

Negative inspiratory force improved from –24 to –46 after PEX

Therapeutic management/changes

Treatment change

Admitted for 5 sessions of PLEX

After her second session, she was able to walk around in the hallways without BiPAP and discharged

2020

Clinical assessments/findings

Patient has a history of extended sick leaves from work due to symptoms and multiple hospitalizations of the past 2 years, with 3 admissions to the ICU

MG-ADL: ranged from 8 to 18

Therapeutic management/changes

Cycles of IVIG and the PLEX rescue therapies have provided only transient relief

Clinical assessments/findings

Patient experienced respiratory worsening needed her BiPAP 3–4 hours during the day

MG-ADL: 15

Therapeutic management/changes

Treatment change

Readmitted for another 5 sessions of PLEX

Monitor MG-ADL scores before, during, and after visits

Recording MG-ADL scores at every visit and encouraging patients to monitor their score between visits could help determine if your patient's gMG is no longer under control. Consider:

- **How are you currently assessing your gMG patients?**
  Are you using the MG-ADL questionnaire or other assessment measures?
- **Do your patients keep track of their condition between visits?**
  How do you advise they keep track?
- **Are your patients sharing the full burden of their disease?**
  Do you discuss changes in their personal and/or professional lives?

Have deeper disease-management discussions

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Help your patients track their day-to-day

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References

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