ISSN: 1607-8322, e-ISSN: 2220-5799 Anaesthesia, Pain & Intensive Care
Vol 27(6); December 2023 DOI: 10.35975/apic.v27i6.2351

ORIGINAL RESEARCH PERIOPERATIVE MEDICINE

Hypotension as a first presentation of myasthenia gravis in an elderly patient scheduled for cervical disc surgery: a case report

Sameh Hamdy Seyam[1], Mohamed Abdelhamid Ali[2], Hassan Razein[3], Islam Elkousy[4], Shyam Babu Schandran[5]

Author affiliations:

1. Sameh Hamdy Seyam, Assistant professor of anesthesia, critical care, and pain management, Al-Azhar University, Faculty of Medicine, Cairo, Egypt; ORCID: {0000-0002-9347-2239} E-mail: sameh_icu1@yahoo.com
2. Mohamed Abdelhamid Ali, Specialist Critical Care, Zulekha Hospital, Dubai, United Arab Emirates; E-mail: drrashwan77@yahoo.com
3. Hassan Razein, Specialist Pulmonologist, Zulekha Hospital, Dubai, United Arab Emirates; E-mail: assanrazein@outlook.com
4. Islam Elkousy, Specialist Critical Care, Zulekha Hospital, Sharjah, United Arab Emirates; E-mail: islamelkousy1@hotmail.com
5. Shyam Babu Schandran, Consultant Neurologist, Zulekha Hospital, Dubai, United Arab Emirates; E-mail: shyambabuc@gmail.com

Correspondence: Sameh Hamdy Seyam, **E-mail:** sameh_icu1@yahoo.com; **Phone:** +971 54 304 4156

ABSTRACT

Myasthenia gravis presents with progressive skeletal muscle weakness. The disease can affect any group of muscles, along with the proximal limb and neck musculature. Elderly patients, especially those over 60 y of age, frequently face extensive delays in diagnosis, or are misdiagnosed, as the symptoms may be erroneously attributed to different other causes frequently seen in this age group, such as degenerative joint disease, disk herniation, osteoporotic vertebral fractures, or metastases. We present a case of a 61-year-old male patient with significant medical records of hypertension, who presented with neck pain for a few months, posted for cervical disc surgery at C6-7 with cage fixation. Two attempts for general anesthesia induction were complicated by severe hypotension. He was evaluated with low-frequency repetitive stimulation, Acetyl-choline receptor antibody, and P/Q calcium channel antibodies. The low-frequency Repetitive Nerve Stimulation (RNS) showed a severe reduced response that was corrected after rest, which was consistent with the diagnosis of myasthenia gravis.

Key words: Myasthenia; Pain, Neck; Hypotension; Weakness

Citation: Seyam SH, Ali MA, Razein H, Elkousy I, Schandran SB. Hypotension as a first presentation of myasthenia gravis in an elderly patient scheduled for cervical disc surgery: a case report. Anaesth. pain intensive care 2023;27(6):763−767; **DOI:** 10.35975/apic.v27i6.2351

Received: July 08, 2023; **Revised:** September 16, 2023; **Accepted:** October 09, 2023

1. INTRODUCTION

Myasthenia gravis is a disease of skeletal muscles, and it may present at any age. The patient suffers from extreme weakness and inability to perform day to day work, especially that involved use of muscle strength. This clinical case describes an uncommon presentation of myasthenia gravis in crisis without any previous history. Myasthenia gravis is an established autoimmune sickness primarily affecting middle-aged females.

In myasthenia gravis, a weak point occurs from impaired action potential propagation to post-synaptic acetylcholine receptors, thereby preventing muscle depolarization.[1] The annual incidence of myasthenia gravis is incredibly low, generally around 10-20 newly reported instances per million individuals.[2]

Classically, myasthenia gravis presents with changing and fatigable skeletal muscle weakness, predominantly affecting the extraocular muscles, with the muscle groups of mastication being stressed to a lesser extent. However, myasthenia can affect any group of muscles, along with the proximal limb and neck musculature.[1] Elderly patients, especially those over 60 years old,

frequently face extensive delays in prognosis or conventional misdiagnoses, as the symptoms may also be erroneously attributed to different causes frequently viewed in this age group, such as degenerative joint disease, disk herniation, osteoporotic vertebral fractures, or metastases.[3] We present a case of a male patient with unusual presentation of severe hypotension during attempted induction of general anesthesia.

2. CASE REPORT

A 61-year-old male patient with significant medical history of hypertension, on a combination of telmisartan 80 mg plus hydrochlorothiazide 12.5 mg, presented to the neurosurgery clinic in April 2023 with neck pain for two years. He was posted for cervical disc surgery at C6-7 level, with cage fixation. The patient had undergone surgical fixation of C6-7 in his home country but the screws and plate were unstable (by CT and MRI findings). The neck ache had no identifiable triggers such as trauma, infection, or stress. The ache worsened at some point in the day, barring any precise exacerbating or alleviating factors. MRI of the neck revealed an anteriorly displaced disc implant slightly tilted to the left side and embedded within the C7 vertebral body with an erosion of the superior endplate associated with marrow edema (Figure 1).

Before the surgery, his echocardiography was normal with EF 70%. His preoperative blood pressure was between 140/97 and 150/100 mmHg. Given the need for neuromonitoring during the surgery, thus, TIVA (Total Intra-Venous Anesthesia) was used for the anesthesia using propofol and remifentanil infusions.

On the day of the procedure and immediately after the loading dose of propofol for induction, the patient

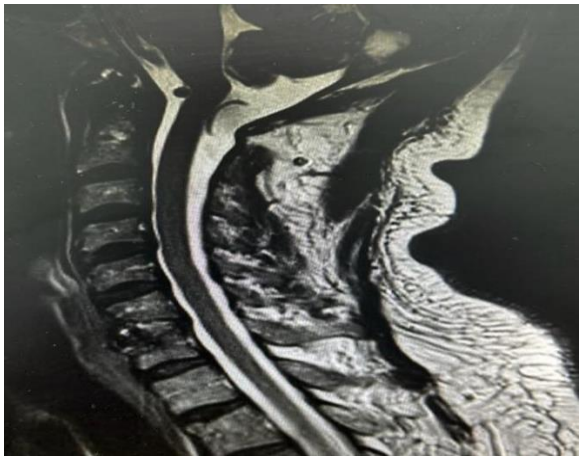

Figure 1: Neck MRI demonstrating an anteriorly displaced disc implant embedded within the C7 vertebral body with an erosion of the superior endplate associated with marrow edema

developed severe profound hypotension which was treated by incremental repeated doses of ephedrine, phenylephrine, and later on by dopamine infusion and rapid crystalloid infusions. The blood pressure slightly improved but the patient remained unstable so the surgery was decided to be postponed. The patient was woken up and uneventfully extubated.

After two days, he was rescheduled for surgery, the morning dose of antihypertensive was omitted, and his preoperative blood pressure was 150/79 mmHg. Anesthesia was induced with a loading dose of propofol (total of 160 mg) and fentanyl 80 mg. Again, his blood pressure dropped immediately and resuscitation was started with incremental repeated doses of ephedrine, and phenylephrine, and later on by dopamine infusion and rapid crystalloid infusions. The blood pressure was fluctuating and the patient remained unstable. He was shifted to the intensive care unit (ICU) for further management.

In the ICU his physical examination revealed signs of generalized fatigue and weakness of his extraocular muscles, bulbar, and limb muscles. The absence of preoperative symptoms like diplopia, ptosis, dysphagia, dysarthria, dysphonia, regurgitation, neck stiffness, photophobia, fever/chills, or respiratory problems further made the diagnosis more difficult. In ICU, the patient was conscious and oriented but did not meet the weaning criteria, with pupils 2 mm reactive to light, restricted eye movements laterally plus ptosis ++, neck flexion power +, upper limbs grade 1/5 proximal differential weakness and elbow + extension more than flexion. For the lower limb, hip flexion power was 3/5, and knee power was 5/5 (for both flexion and extension). He was evaluated with low-frequency repetitive nerve stimulation (RNS), acetyl-choline receptor antibody, and P/Q calcium channel antibodies. The low-frequency RNS showed a severe decreased response that got corrected after rest (Figures 2, 3, and 4).

His acetyl-choline receptor antibodies came 20 times higher than normal (Normal: less than 0.05 nmol/L).[1] The preliminary differential diagnosis was made of myasthenia crisis vs Lambert Eaton syndrome.

He was initiated on immunoglobulins 2 gm/kg IV for 7 days. Remarkably, the patient experienced a dramatic improvement in signs and symptoms following the initiation of intravenous immunoglobulins. The patient was gradually weaned off from the mechanical ventilator and then shifted to the ward. His quantitative myasthenia score was 38. Steroids were initiated at low dosages and gradually escalated.

Before discharge from the hospital, spirometry/ flow-volume studies were done (Figure 5). A restrictive ventilatory pattern was established as forced expiratory

volume in 1sec/ forced vital capacity (FEV1/FVC) ≥ 0.70, and a predicted EVC < 80%, in conformity with updated American Thoracic Society/Europian Respiratory Society guidelines.

The disorder development was not aggressive, and the patient remained stable on his ICU course. Also, no unfavorable consequences of the remedy were reported, enabling the patient to return to his preceding baseline lifestyle, free from difficulties with vision, swallowing, speech, or gait.

Lastly, this case study was approved by the hospital ethical committee No: ZH-07-2023, and patient informed consent was taken. All updated guidelines were followed during the management of the patient in the ICU.

3. DISCUSSION

The traditional management for stable myasthenia gravis includes indirect-acting anticholinesterases, such as pyridostigmine or neostigmine, and immunosuppressants.

Thymectomy, the surgical elimination of the thymus gland, is a viable alternative for symptom control in younger patients. However, it is commonly not liked for elderly patients, specifically those above 60 years old.[4] Elderly patients have commonly exhibited favorable responses to therapeutic interventions. As the principal muscle accountable for breathing, the diaphragm performs a fundamental function in inhalation and exhalation.[5] In myasthenia gravis, the immune-mediated assault on the post-synaptic acetylcholine receptors at the neuromuscular junction can extend to involve the diaphragmatic muscle fibers, impairing their contractility and inflicting weakness. However, the symptoms of diaphragmatic paralysis may be

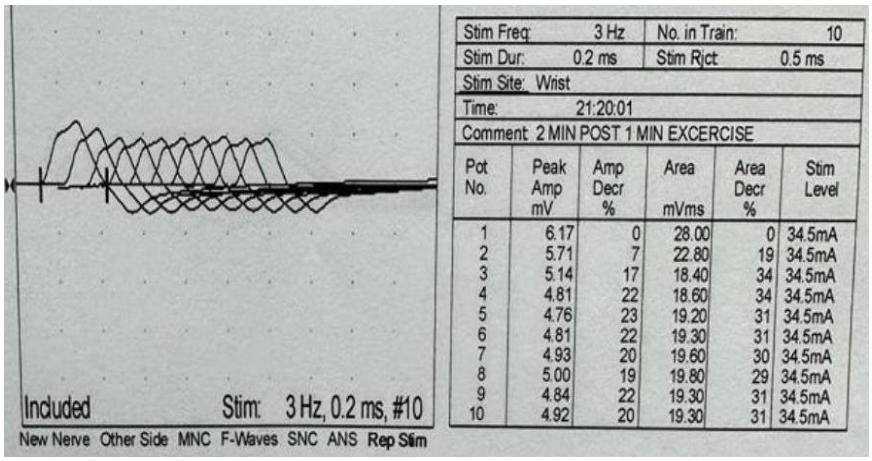

Figure 2: RNS study at 3HZ from left abductor digiti minimi and trapezius muscles showing significant decremental responses.

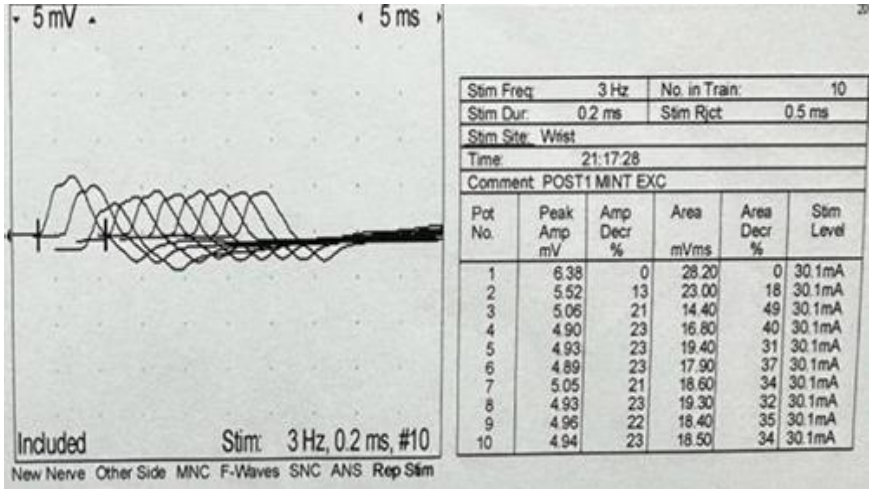

Figure 3: RNS study at 3HZ from left abductor digiti minimi and trapezius muscles showed significant decremental responses

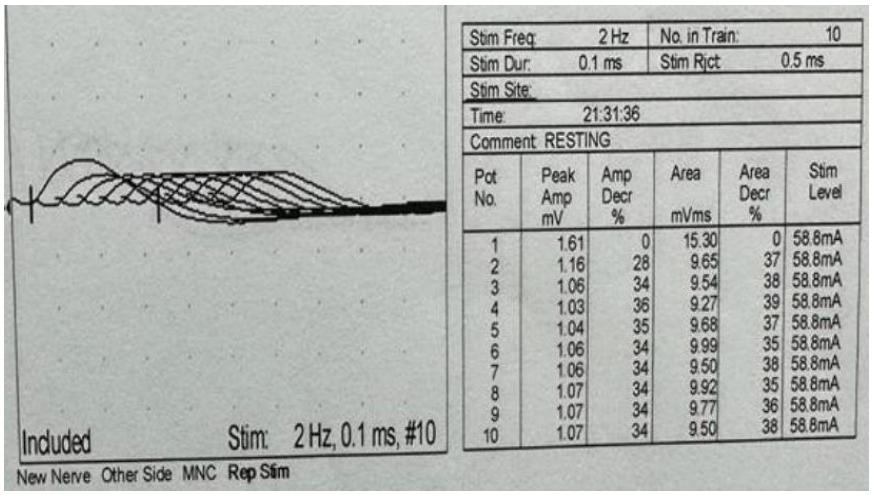

Figure 4: RNS study at 3HZ from left abductor digiti minimi and trapezius muscles showed significant decremental responses.

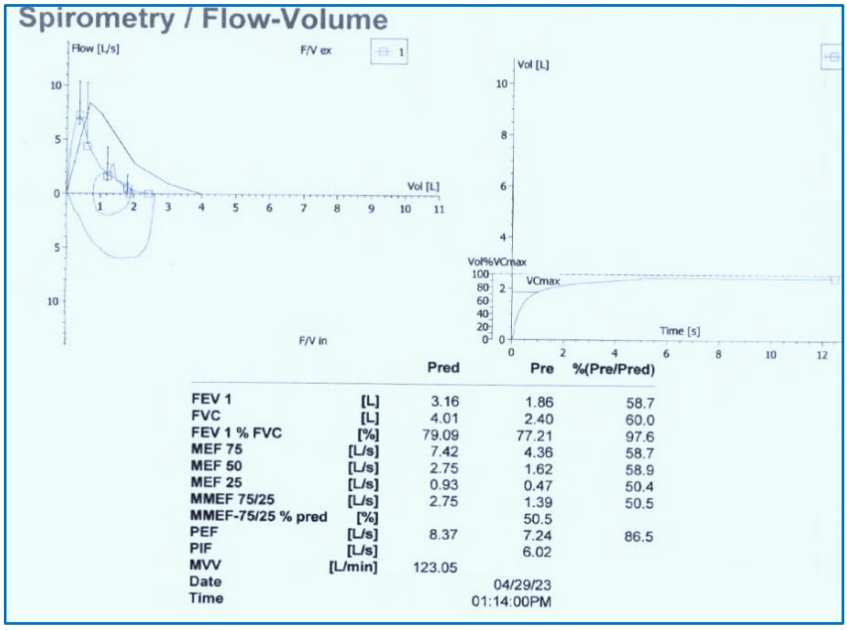

Figure 5: Spirometry/Flow-volume study prior to discharge from the hospital.

nonspecific or attributed to other causes, mainly to a lengthening in fantastic diagnosis.[6]

Cervical trauma or manipulation has been pronounced a possible trigger for diaphragmatic paralysis in positive cases. Injuries or undue stress to the cervical area can affect the phrenic nerve, which factors in motor innervation to the diaphragm. Consequently, injury to the phrenic nerve can result in unilateral or bilateral diaphragmatic dysfunction, impairing the regular respiratory mechanics and leading to respiratory compromise.[7]

The diagnostic assessment of diaphragmatic dysfunction might also contain a range of modalities. Pulmonary function tests, such as spirometry and size of forced vital capacity (FVC), can offer valuable information about lung volumes and airflow. Unfortunately, it could not be possible to do this before ICU admission as the patient was admitted directly from the operation theatre. Imaging studies, including chest X-rays and fluoroscopy, can help visualize the diaphragm's function and movement during respiration.[8]

Myasthenia gravis (MG) is usually troublesome by respiratory failure, commonly called a myasthenic crisis. Nonetheless, a lot of the patients who evolve pulmonary manifestation do so during the tardy course of the disease and have other central signs and symptoms. However, in some patients, respiratory failure is the early displaying symptom.[9]

Treatment strategies for diaphragmatic paralysis in the context of myasthenia gravis center on managing the underlying autoimmune disease.[10] This normally includes using immunosuppressive medications, such as corticosteroids or other immunomodulating agents, to minimize the immune response and forestall additional damage to the neuromuscular junction.[11] Concurrently, respiratory aid and management are crucial to make sure ample ventilation. In extreme cases, interventions such as noninvasive pressure ventilation (NIPPV) or, in severe cases, invasive mechanical air flow may also be required to perform a respiratory function.[12, 13]

Hypotension can occur in myasthenia crisis due to multiple causes. Myasthenia gravis can affect respiratory muscles contributing to normal breathing, leading to respiratory failure. The resulting deficient oxygenation can generate vasodilatation and hence hypotension. A malfunction of the autonomic nervous system can happen in a myasthenia crisis. Also, fluid depletion, fasting, diuretics, as well as antihypertensives can contribute to profound hypotension in a myasthenia crisis.[14]

This case highlights a notably unique presentation of an uncommon condition, emphasizing the importance of maintaining an excessive clinical index of suspicion for myasthenia gravis in aged patients exhibiting unexplained neck or bulbar weakness, even in the absence of classical ocular muscle weakness or previous history.

4. CONCLUSION

Although there are not many recorded cases of myasthenia gravis displaying only neck pain among elderly patients. Elderly patients are at a big risk of being misdiagnosed due to their complexity. This case scenario put an important focus that detailed neurological examination and screening for myasthenia gravis should be carried out in elderly patients presenting with severe hypotension as a response to triggering factors for myasthenic crisis.

5. Conflicts of interest

No conflicts of interest declared by the authors.

6. Authors contribution

All authors contributed in the conduct of this case management and the preparation of this manuscript.

7. Ethical issues

Patients written consent was obtained to share this case report for educational purposes.

8. REFERENCES

1. Phillips LH 2nd. The epidemiology of myasthenia gravis. Ann N Y Acad Sci. 2003 Sep;998:407-12. [PubMed] DOI: 10.1196/annals.1254.053
2. Spillane J, Higham E, Kullmann DM. Myasthenia gravis. BMJ. 2012 Dec 21;345:e8497. [PubMed] DOI: 10.1136/bmj.e8497
3. 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 Jan 19;96(3):114-122. [PubMed] DOI: 10.1212/WNL.0000000000011124
4. Schon F, Drayson M, Thompson RA. Myasthenia gravis and elderly people. Age Ageing. 1996 Jan;25(1):56-8. [PubMed] DOI: 10.1093/ageing/25.1.56
5. Fujibayashi S, Shikata J, Yoshitomi H, Tanaka C, Nakamura K, Nakamura T. Bilateral phrenic nerve palsy as a complication of anterior decompression and fusion for cervical ossification of the posterior longitudinal ligament. Spine (Phila Pa 1976). 2001 Jun 15;26(12):E281-6. [PubMed] DOI: 10.1097/00007632-200106150-00029
6. Evoli A, Meacci E. An update on thymectomy in myasthenia gravis. Expert Rev Neurother. 2019 Sep;19(9):823-833. [PubMed] DOI: 10.1080/14737175.2019.1600404
7. Berrouschot J, Baumann I, Kalischewski P, Sterker M, Schneider D. Therapy of myasthenic crisis. Crit Care Med. 1997 Jul;25(7):1228-35. [PubMed] DOI: 10.1097/00003246-199707000-00027
8. Fregonezi GA, Resqueti VR, Güell R, Pradas J, Casan P. Effects of 8-week, interval-based inspiratory muscle training and breathing retraining in patients with generalized myasthenia gravis. Chest. 2005 Sep;128(3):1524-30. [PubMed] DOI: 10.1378/chest.128.3.1524
9. Gracey DR, Divertie MB, Howard FM Jr. Mechanical ventilation for respiratory failure in myasthenia gravis. Two-year experience with 22 patients. Mayo Clin Proc. 1983 Sep;58(9):597-602. [PubMed]
10. Mehta S. Neuromuscular disease causing acute respiratory failure. Respir Care. 2006 Sep;51(9):1016-21 [PubMed]
11. Conti-Fine BM, Milani M, Kaminski HJ. Myasthenia gravis: past, present, and future. J Clin Invest. 2006 Nov;116(11):2843-54. [PubMed] DOI: 10.1172/JCI29894
12. Thieben MJ, Blacker DJ, Liu PY, Harper CM Jr, Wijdicks EF. Pulmonary function tests and blood gases in worsening myasthenia gravis. Muscle Nerve. 2005 Nov;32(5):664-7. [PubMed] DOI: 10.1002/mus.20403
13. Dushay KM, Zibrak JD, Jensen WA. Myasthenia gravis presenting as isolated respiratory failure. Chest. 1990 Jan;97(1):232-4. [PubMed] DOI: 10.1378/chest.97.1.232
14. Vernino S, Cheshire WP, Lennon VA. Myasthenia gravis with autoimmune autonomic neuropathy. Auton Neurosci. 2001 May 14;88(3):187-92. [PubMed] DOI: 10.1016/S1566-0702(01)00239-9

