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Myasthenia Gravis- a Disguised Cause of Recurrent Respiratory Failure: A Rare Case Treated Successfully at National Hospital Sri Lanka

M. N. S. K. Perera [a++*], D. H. H. L. Sathischandra [a#], I. A. Maharambe [a†] and N. M. Ellawala [a‡]

[a] National Hospital of Sri Lanka, Sri Lanka.

Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Myasthenia gravis is an autoimmune disease, due to the presence of antibodies against the proteins in the post-synaptic muscle membrane, which leads to localized or generalized muscle weakness.
Presentation of myasthenia gravis could vary from patient to patient, while ocular muscle weakness is the most common form.
Here, a case of a young male who presented with recurrent respiratory failure is reported.
He had respiratory failure as the only clinical manifestation of myasthenia gravis. It describes respiratory weakness as a rare isolated initial presentation of this disorder. This case highlights the importance of prompt diagnosis of this condition, considering the variable presentations, to achieve a successful therapeutic outcome.

[++] MD(Medicine), MBBS, Senior Registrar in Internal Medicine;
[#] FRCP, MD, MBBS-Specialist in Internal Medicine;
[†] MBBS, MD(Medicine), Senior Registrar in Medicine;
[‡] MBBS, Registrar in Medicine;
**Corresponding author: E-mail: nishadisaumya19@gmail.com;*

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1. INTRODUCTION

Myasthenia gravis is a neuromuscular disorder, which occurs due to autoantibodies against presynaptic neuronal proteins [1]. The majority of patients with myasthenia gravis present with ocular manifestations and, the minority with other muscle paralysis [2].

The case highlighted here is of a young male with isolated respiratory muscle weakness, which was initially a diagnostic challenge. Diagnosis of myasthenia gravis was made on clinical grounds, which was later confirmed by electromyography and serology. It highlights the importance of considering divergent clinical presentations for this disorder.

2. CASE PRESENTATION

A 27-year-old male, presented to the acute medical ward with worsening shortness of breath for 2 days.

His history was significant for a hospital admission 3 months before the current presentation, with severe acute respiratory syndrome coronavirus (SARS-CoV-2) infection complicated with pneumonia. During that admission, the patient progressed to type 2 respiratory failure necessitating ventilator support. During the Intensive Care Unit (ICU) stay, attempted endotracheal extubation failed twice but the third attempt was successful.

Evaluations were made to look for the cause of the failed extubation during that admission, and the investigations had been normal except for diaphragmatic Electromyography (EMG) showing some myopathic changes. He remained well following weaning off from respiratory supports, and was discharged home with a plan to review with a repeat EMG in 6 months or earlier if symptomatic. He remained well during the post-discharge period.

During the current admission, he complained of worsening difficulty in breathing for two days, without fever, cough, or sputum production. Further questioning did not reveal any probable exposure to pesticides or other toxins.

He demonstrated profound hypoxia with type 2 respiratory failure soon after admission. His lungs had normal vesicular breath sounds, without added sounds, on auscultation. He didn't have bulbar, limb, or neck muscle weakness. The patient was in acute respiratory distress and was electively intubated.

His arterial blood gas analysis, immediately prior intubation showed the following findings:

pH-7.15

HCO3-23.4 mmol/L

pCO2-72 mmHg

pO2-59 mmHg

SpO2-82%

He was managed in the Intensive Care Unit (ICU) for ventilatory support, while the exact reason for the respiratory failure was being evaluated. Due to repeated episodes of failed extubation, a tracheotomy was performed, and non-invasive ventilator care was given, through the tracheostomy tube.

There was no family history of myopathy, neuromuscular disorder or any other form of neurological disorders. His past history too didn't reveal any fatigable muscle weakness.

Complete blood count, serum electrolytes, liver and renal panels were unremarkable, and the urine toxicology screening was negative. Thyroid profile was normal and the retroviral screening was negative. Creatinine phosphokinase (CPK)values were also within normal range.

Lung parenchymal and vascular imaging with High-Resolution CT (HRCT) of the chest and CT pulmonary angiogram(CTPA) did not reveal any abnormality. Magnetic resonance Imaging(MRI) of the brain and electroencephalography (EEG) were unremarkable, so central causes for respiratory failure were excluded. At this point, electromyography(EMG) accompanied by Nerve conduction studies(NCS) revealed a significant decremental pattern with repetitive nerve stimulation studies in the ulnar nerve, suggesting a possible neuro-muscular junction disorder. Further assessment with neck imaging with Contrast-Enhanced CT(CECT) confirmed the presence of thymic hyperplasia He was started on intravenous steroids, and pyridostigmine, pending the serological studies, as myasthenia

gravis was likely based on the electrophysiological studies.

The patient showed a remarkable improvement, with the commencement of management, and the non-invasive ventilator supports were weaned off.

He was discharged with oral steroids and azathioprine for immune suppression, in addition to pyridostigmine. He was reviewed at the clinic frequently, and by the first clinic visit, his Acetylcholine-esterase antibody serology results were available as a significantly positive value, further confirming the diagnosis of myasthenia gravis. He did not have a recurrence of symptoms and has been well up to date.

3. DISCUSSION

Myasthenia gravis (MG) is an autoimmune disease, due to the presence of antibodies against the acetylcholine receptor (AChR), muscle-specific kinase (MuSK) or other AChR-related proteins in the postsynaptic muscle membrane, which leads to localized or general muscle weakness [1]. Therefore, the diagnosis is based on clinical findings, electrophysiology findings, and serology [3,4].

It commonly presents with ocular muscle weakness, progressing to limb involvement, which is seen in 85% of patients [5,6]. Clinical examination would reveal more than one extraocular muscle involvement, with pupillary sparing. 60% of patients with the disease would demonstrate bulbar muscle weakness, which can present with fatigable chewing and difficulty in deglutition [7]. Respiratory muscle involvement is rarely seen during the first two years of onset of the illness [8]. It occurs in intercostal and diaphragmatic muscle involvement, and if present, leads to myasthenic crisis, which is a life-threatening consequence, often requiring mechanical ventilation. Several precipitating factors for myasthenic crisis, such as medications and intercurrent infections have been identified in the literature [9].

The existing literature is scarce, when it comes to cases of myasthenia gravis presenting with respiratory failure as the first presentation [10]. The very few cases reported in the literature with such respiratory failure, have been usually associated with ocular and bulbar involvement [11,12].

The above-described patient demonstrated recurrent unexplained isolated respiratory failure. He didn't have ocular muscle involvement, bulbar muscle involvement, or any extremity skeletal muscle involvement in each of the episodes. All other possible aetiologies for respiratory muscle weakness have been excluded through thorough evaluation, while the electrophysiological studies later confirmed the diagnosis, as myasthenia gravis. However, the diagnosis was a challenge, due to the uncommon nature of its presentation as a sole respiratory manifestation.

4. CONCLUSIONS

In conclusion, we report a case of a young male who presented with recurrent respiratory failure as the first manifestation of myasthenia gravis. Though rare, respiratory weakness can be the isolated initial presentation of this disorder. This case highlights the importance of considering myasthenia gravis, in patients presenting with isolated, unexplained respiratory weakness.

CONSENT

Informed written consent was obtained from the patient.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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