

An undiagnosed myasthenia gravis presenting as isolated recurrent acute respiratory failure

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ABSTRACT

Acute respiratory failure is an uncommon initial presentation of myasthenia gravis (MG). In our case a 22-year-old woman of unrecognized MG presented to the emergency department with isolated respiratory failure as the first presenting symptom. Initially she presented with dysphonia and was managed by speech therapist and ENT surgeons for 3 months. Subsequently, she presented with signs and symptoms of sepsis and went into acute respiratory failure. This case highlights the need to consider MG in the differential diagnosis of an otherwise unexplained respiratory failure in the critical care setting.

Key words: Myasthenia gravis, respiratory failure, MuSK

Introduction

Myasthenia gravis (MG) is an autoimmune disorder resulting from the production of antibodies against acetylcholine receptors leading to the destruction of the postsynaptic membrane at the neuromuscular junction, which results in variable muscle weakness made worse by exercise.^[1]

MG is often complicated by respiratory failure, known as myasthenic crisis. However, most of the patients who develop respiratory symptoms do so during the late course of disease and already have other neurological signs and symptoms. However, in some patients respiratory failure is the initial presenting symptom. We present a case of previously undiagnosed MG who presented with recurrent acute respiratory failure.

Case Report

A 22-year-old woman presented to the emergency with

dyspnea for 3 days. She denied cough, sputum, chest pain, or other respiratory symptoms. She also denied symptoms of upper and lower extremity weakness, blurred vision, or swallowing difficulty. Further history revealed that she had speech problem (nasal speech) treated by ENT surgeons and speech therapist. There had no history of substance abuse or recent drug intake.

On the physical examination, she had labored breathing with respiratory rate of 40 breaths/minute. The patient was confused and SaO₂ was 85%. The blood pressure was 146/98 mm Hg, pulse was 104/minute and regular and body temperature was 36.7°C. Pedal edema and neck vein engorgement were absent. On the chest auscultation, there were mild crackles at both lung fields. The arterial blood gas analysis on an O₂ mask with 10 L was pH7.314, PaCO₂ 56.7 mm Hg, PaO₂ 92.2 mmHg and oxygen saturation 90%. The chest X-ray revealed no definite infiltration but the lung volume was reduced. Other laboratory evaluation showed a white blood cell count of 6980 µL, hemoglobin of 12.5 g/dL and platelet count of 345 000 µL, blood glucose 94 mg/dL, BUN/Cr 12/0.8 mg/dL, AST/ALT 24/25 units/L. The chest computed tomography (CT) scan showed no definite evidence of pulmonary thromboembolism. ECG showed sinus tachycardia and echocardiography revealed normal systolic and diastolic heart function. As respiratory failure was imminent, she was promptly given supplemental oxygen and transferred to the intensive care unit and mechanically ventilated under

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10.4103/0976-3147.91961

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rapid sequence induction (IV midazolam 3 mg and succinylcholine 70 mg). Her SaO₂ rapidly improved to 100% after intubation. However, she was unable to breathe without mechanical support even after the effect of muscle relaxant had worn off.

After admission, assessment by ENT surgeon did not reveal any upper airway obstruction. We considered other causes of respiratory failure such as neuromuscular disorders, Guillain-Barre syndrome or MG. The physical and neurological examination and cerebrospinal fluid analysis were normal. A diagnosis of MG was made on the neurophysiological studies. The electromyography showed a decremental response to repetitive nerve stimulation in various muscles. The pharmacological Jolly test revealed incremental responses of tidal volume of ventilation. The acetylcholine receptor antibody titer was 12.4 nmol/L. Her condition gradually improved with pyridostigmine bromide 720 mg/day and prednisolone 30 mg/day and intravenous gamma-globulin 400 mg/kg/day for 5 days were administered. Weaning from ventilator failed over the next 2 weeks; therefore, a tracheostomy was performed. However, the patient was eventually successfully weaned from the ventilator and 2 months later, she was discharged. She was advised to do thymectomy but she developed again respiratory distress and subsequent respiratory arrest with signs and symptoms of sepsis. She was successfully intubated and resuscitated. The prednisolone and pyridostigmine were stepped up along with antibiotics, fluid, and intravenous gamma-globulin 400 mg/kg/day for 5 days she received. She was finally discharged on day 24. The section of excised thymus showed follicular hyperplasia only. Currently she was still followed up in our hospital and responded well with maintenance steroid and pyridostigmine.

Discussion

MG is a neuromuscular disorder of autoimmune origin. The hallmark of the disease is fluctuating weakness and increased by exertion. Weakness increases during the day and improves with rest. Presentation and progression vary.^[2] Majority of patients are female. The patients may present with diplopia, dysphagia, ptosis, and limb weakness. Respiratory failure can be a complication during the late course of MG in about 3% to 8% of cases, known as a myasthenic crisis.^[3] However, isolated respiratory failure as the presenting symptom, as in the present case, is very unusual and this patient in our knowledge is the first case reported in the literature.

Our case has demonstrated the rare but well-known manifestation of MG. Indeed our patient has already

presented with symptoms of dysphonia for 3 months to ENT department, which was compatible with MG. However, it was not diagnosed until the patient presented with acute respiratory failure. As the diagnosis was not known at our department, succinylcholine was used as muscle relaxant for rapid sequence intubation. It was well known that it may cause prolonged paralysis in myasthenic patients. However, our patient did not manifest any prolonged paralysis afterwards.^[4]

We tried to determine the cause of respiratory failure. We did not find any evidence of a hypoxemic respiratory failure; there were no definite infiltrations, edema, effusion, or pneumothorax on chest X-ray. There was no evidence of pulmonary thromboembolism on chest CT scan and no evidence of intracardiac shunting or congestive cardiac failure on echocardiography. Therefore, we suspected an acute ventilatory failure. Drugs or trauma that could cause ventilatory failure was ruled out. The electrolyte levels were within normal limits. Based on these results, we investigated the possibility of a neuromuscular disease, especially the Guillain-Barre Syndrome and MG, the most common and the second most common cause of neuromuscular disease, respectively. However, the cerebrospinal fluid was normal, and the neurophysiological studies showed evidence of MG.^[5] Acute respiratory failure can be due to fatigue of respiratory muscles. It can also be due to upper airway obstruction. They may present with stridor.^[6-8] The vocal cords of our patient was also involved by the disease as she already had a speech problem.

Generally patients with neuromuscular disease present with hypoxia and a normal D (A-a) O₂ due to hypoventilation, but in the present patient there was an increased D(A-a)O₂. Patients with MG occasionally have atelectasis which could result in this finding. It is difficult to consider a neuro muscular disorder, especially MG, unless there are other neurological symptoms such as ocular or bulbar symptoms.^[9] Myasthenic cases with positive anti-MuSK antibody tend to be young female subjects presenting with bulbar weakness, and often responding poorly to symptomatic treatment with a higher risk of respiratory failure.^[10] An obvious advantage of plasmapheresis over high doses of intravenous immunoglobulins has not been demonstrated; however, rapid effects of plasmapheresis support this therapeutic approach.^[11] Our patient presented only with respiratory symptoms, the patient had no other symptoms, generally associated with MG during her 6-month follow up visits.

MG is not a common entity that we encounter daily. Patients, on occasions, may present to the emergency department because of acute exacerbation. Though

most of them were known cases, we should be aware of some unrecognized cases and should consider MG as a differential diagnosis for patient with acute respiratory failure.

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How to cite this article: Sharma SR, Sharma N, Yeolekar ME. An undiagnosed myasthenia gravis presenting as isolated recurrent acute respiratory failure. *J Neurosci Rural Pract* 2012;3:80-2.

Source of Support: Nil. **Conflict of Interest:** None declared.