

Case Report

Use of non invasive ventilation to avoid Re-Intubation in Myasthenia Gravis; a case report and review of literature

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Abstract

Myasthenia Gravis (MG) in the elderly is an uncommon finding, especially when it is not related to thymoma. A case is presented with late onset Myasthenia Gravis treated with steroids, immunosuppressives and mechanical ventilation.

This 61 year Asian hypertensive lady clinically diagnosed as MG presented to emergency room with difficulty in swallowing liquid, diplopia, drooping of eyelids and generalized weakness. Within 24 hours in the ward developed respiratory distress and CO₂ narcosis, for which she was immediately intubated and shifted to ICU and managed by invasive ventilation and inotropic support. After stabilization and extubation BiPAP was applied successfully.

BiPAP use is an established, non invasive ventilation technique for Myasthenia Gravis. Its application to avoid re-intubation has not been thoroughly investigated. We intend to highlight this area for further research as it may change the total length of ICU and hospital stay and more importantly the outcome for this subset of patients.

Keywords: Myasthenia Gravis, BiPAP, Non-invasive ventilation.

Introduction

MG is a relatively uncommon disease, although prevalence has increased over time with recent estimates approaching 20 per 100 000 in the US population.¹ Incidence varies widely from 1.7 to 10.4 per million, depending on the location of study,² and has been reported to be as high as 21 per million in Barcelona, Spain.³ After 50 years of age, incidence is higher in men.⁴ The age peak for late-onset MG (onset after the age of 50 years) is now the same for both sexes.⁵

Use of noninvasive ventilation techniques to avoid re-intubation in myasthenia crisis is well established but relatively uncommon practice. In literature much of the focus is for avoiding Intubation and invasive ventilation immediately for myasthenic crises.⁶

Noninvasive ventilation using BiPAP should be considered in selected patients with MC who have respiratory compromise (those without hypercapnia and with the ability to synchronize with the machine) as the initial method of ventilatory support.⁶

BiPAP can be tried first in selected patients with acute respiratory failure especially due to myasthenic crisis and possibly other causes, while awaiting improvement from other specific therapies. However, it should be stressed, that there is a need for closer cardiac monitoring, as patients with myasthenic crisis are prone for cardiac arrhythmias.

Also, repeated clinical assessment and arterial blood gas monitoring must be done every 6-8 hours to judge the adequacy of NIV ventilation.^{7,8}

Literature review enhances the importance of NIV as an attempt to avoid invasive ventilation, but, for patients with MC (already extubated) at the brink of re-intubation the use of BiPAP as a rescue modality is relatively new concept in the presence of high PaCO₂ levels.

BIPAP Treatment Changes:

Early non-invasive ventilation after extubation reduces the risk of respiratory failure and lowered 90-day mortality in patients with hypercapnia.⁹ Routine implementation of this strategy for management of mechanically ventilated patients with chronic respiratory disorders is advisable.^{9,10}

Noninvasive ventilation may be effective in avoiding respiratory failure in selected patient categories like obesity, if applied during the first 48 hours of post-extubation.

Extubation failure is associated with high morbidity and mortality, and NIV has been suggested as a way to avoid re-intubation and improve outcomes.¹¹

In selected patients with chronic hypercarbia of various etiologies, early application of noninvasive ventilation proves to be of survival benefit.¹⁰⁻¹⁴

In non-COPD patients with persistent acute respiratory failure post extubation, NIV improved pulmonary gas exchange and breathing pattern, decreased intrapulmonary shunt and also the work of breathing.^{14,15}

Although the beneficial effect of noninvasive ventilation in improving, survival of hypercapnic patients with chronic respiratory disorders can not be emphasized more but this modality needs to be investigated further through prospective clinical trials.

Case Presentation

A 61 year female presented to the accident and

emergency with complaints of difficulty in swallowing liquids. It started gradually with solids but then gradually progressed to difficulty in swallowing liquids. There was no pain or swelling around the neck.

Since two months the patient had been experiencing drooping of both eyelids, which also started gradually. Initially she could resist it but then she had to physically raise them. This was followed with double vision in both eyes, fatigue which she described as feeling weak on and off, having difficulty turning in the bed, difficulty in sitting and walking without support which was not there before, but all of these symptoms would last for a couple of hours only and then she would feel normal again. She had also on and off slurred speech and reduced bowel movements. Her physician clinically diagnosed her with Myasthenia Gravis and started oral Pyrostigmine 15 mg QID and Prednisolone 20mg OD.

She was a known hypertensive for over 40 years on Losartan 50mg daily and bilateral knee osteoarthritis on Calcium supplementation and analgesics. She had a history of cataract and varicose vein operation in 1998 and 2000 respectively. There was no family history of any medical illness and she was not a smoker or alcoholic.

On arrival to A&E, she was looking weak, had drooping of both the eyelids, talking but getting exhausted very soon. She couldn't keep her eyes open or keep her hands straight for more than 60 seconds. Her muscle tone and power was generally reduced with normal muscle bulk. Chest examination revealed bilateral equal air entry with clear breath sounds. Heart sounds were normal on auscultation. Abdomen was bulky but soft, not tender with no organomegaly and positive bowel sounds. Complete blood count showed white blood count of 15000/cm³, Hb= 14.1g/dl, normal renal functions, liver functions and thyroid functions. CT scan of thorax was done as thymoma was suspected but the result was normal. Blood samples for DS DNA, Ach antibodies and ANuclear factor were sent to a neighboring hospital due to lack of the facility at our hospital.

She was conservatively managed in the ward with oral Pyrostigmine 15mg qid, prednisolone 20mg od, Iv ceftriaxone and Intravenous fluids.

Just after 24 hours, suddenly she became unresponsive to speech and pain, BP started to drop with declining oxygen saturation. Arterial blood gas revealed pH=7.01, pCO₂=147.3, pO₂=77.2, HCO₃⁻= 37.1 and O₂ saturation=85% confirming severe respiratory acidosis and CO₂ retention. She was immediately intubated, kept on ventilator support and transferred to Intensive Care Unit. After intubation she was started on dopamine infusion, intravenous fluids, increased Pyrostigmine to 30mg qid, increased prednisolone to 25mg od and azathioprine was started at 200mg once daily. Plasmapheresis and Tensolin

test were considered but due to lack of facility at out hospital, they were not done.

By the 6th day on ventilator, she was off ionotropes, subsequent ABGs had been normal with minimal ventilator support and rapid shallow breathing index of <100 so she was successfully extubated and remained stable.

After 2 days post extubation, she started becoming drowsy, disoriented but was talking, ABGs showed increased pCO₂.

She was put on BiPAP intermittently for 4 hours and resting in between. BiPAP settings were IPAP 16 and EPAP 6-8 cmH₂O, with 4 hours ON and 2 hours rest.

After 24 hours of the therapy, she became more alert, fully conscious, oriented and communicating well. BIPAP was discontinued after 24 hours and ABGs were evaluated everyday. Regular physiotherapy was given for the chest and limbs and intensive spirometry everyday. She started improving and eventually on the 17th day she was discharged with oral Pyrostigmine 30mg thrice daily, azathioprine 200mg once daily and tapering prednisolone.

Discussion

The successful management of this patient not only involved the pharmacological aspect but a major part was mechanical ventilation. Although widely used for normocarbic patients, the BiPAP application for hypercapnic patient in order to avoid re-intubation is promising and probes to conduct randomized controlled trials on the technique before concluding the overall beneficial outcome.

When applied properly it not only reduces ICU and hospital stay but also in hospital mortality and ventilator associated infections.

Conclusion

We conclude that non invasive ventilation for MC prevents re intubation in selected patients. We recommend a randomized controlled trial on the subject before applying this promising technique to general population.

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