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MYASTHENIA GRAVIS PATIENT ON VENTILATORY SUPPORT : A CASE REPORT.

Puneet Verma* MD, Anaesthesiologist. District Hospital, Mandi, Himachal Pradesh. *Corresponding Author

ABSTRACT

Anticholinesterase medications are the mainstay of treatment for myasthenia gravis patients. Severe episodes may be caused by insufficient medication (myasthenic crisis) or excessive medication (cholinergic crisis). The distinction between "cholinergic" and "myasthenic" crisis becomes inconsequential. I present a rare case of myasthenia gravis with recurrent exacerbations due to myasthenia/ cholinergic crisis requiring ventiltory support.

INTRODUCTION

Myasthenia gravis is an autoimmune disorder characterized by weakness and easy fatigability of skeletal muscle. The prevalence - 50–200 per million population. The incidence is highest in women during their third decade, and men exhibit two peaks, one in the third decade and another in the sixth decade. Fundamental defect is decrease in available Ach Receptors at postsynaptic muscle membrane. 75% of patients with MG have some degree of thymus abnormality (eg, hyperplasia in 85% of cases, thymoma in 15% of cases). Associated with other disorders of autoimmune origin such as thyroid hypofunction, rheumatoid arthritis and systemic lupus erythematosus. Both myasthenia and cholinergic crisis can present as respiratory failure but then are clinically indistinguishable.

CASE REPORT

A 42 years old female. Admitted with chief complaints of generalised weakness since 2 days and difficulty in breathing since 2 days.

History of present illness:

- K/C/O Mysthenia Gravis- admitted for one week in medicine department IGMC Shimla for generalised weakness and discharged one month back.
- Patient was taking Tablet pyridostigmine 60 mg tds and Tablet Omnacortil 40 mg OD.
- Presented now for generalised weakness.
- Difficulty in breathing for 2 days.
- Insidious onset, gradually progressive.
- No H/O cough, fever, orthopnea, PND.
- No H/O chest pain, palpitation, syncope.

(Day 1):

- In casualty- patient had respiratory distress
- HR- 100/min, BP- 116/86 mm Hg, RR- 50/min , use of accessory muscle present, SpO2- 75-85% with oxygen by ventimask
- ICU call was sent for respiratory distress- patient was intubated immediately at 9:35 am with CoETT 7.5 mm I/D and fixed at 21 cm.
- No bed was available in ICU at that time, so patient was put on ambu bag ventilation @ 10-12 bpm with oxygen attached.
- Neurology opinion was taken- diagnosis of mysthenia crisis made.

Treatment Plan Was Made:

- Plasmapharesis
- Tab. Pyridostigmine 60 mg tds
- Inj. Ceftriaxone 1gm i.v. BD
- Inj. Azee 500 mg i.v. OD
- Tab Omnacortil 40 mg OD

But Patient Could Not Afford Plasmapharesis

- tab. Omnacortil omitted and Inj. Methylprednisolone 1gm in 100cc NS i.v. for 3 days started.

Management in ICU:

- Patient was shifted to ICU at 5pm and put on ventilatory support; mode SIMV, Vt 450, rate 12, PEEP 5, FiO2 – 50%.
- Patient was catheterised with folley's catheter
- Ryle tube feeding was started

(Day 3):

- Patient was on ventilatory support; mode SIMV.
- Inj LMWH 0.4 ml s/c OD was started.
- inj methylprednisolone omitted and tab. Omnacortil 40mg OD started.
- Tab pyridostigmine was witheld as patient was on ventilatory support.[1]

(Day 5):

- Patient was on spontaneous mode of ventilation
- Psupport 10, PEEP 5, FiO2 40%
- Vitals were- HR- 90/m, BP- 138/80, SpO2- 96%
- Tab pyridostigmine 60 mg tds was started
- Patient was extubated and put on ventimask FiO2 40%

(Day 6):

- Patient was shifted to medicine ward on room air
- Shifting vitals- HR 84/m, BP- 140/84 mmHg, SpO2- 96%, RR-15/min.
- Inj Immunoglobulin 5gm i.v. 5 times a day was started in the ward

(Day 8):

- At 5:10 PM patient had shortness of breath with RR 25/m, SpO2- 90-95%
- Propped up position was given and O2 was started with ventimask.

(Day 9):

- At 6 AM ICU call was sent for respiratory distress with RR of 50-60/m with use of acessory muscles, SpO2 was 75-80%
- Patient was intubated with CoETT 7.5 mm and shifted to ICU at 11 am.
- Diagnosis of myasthenia crisis was made due to inappropriate immunosupression.
- Tab omnacortil was increased to 50 mg OD.
- Patient was put on ventilatory support.
- Tab pyridostigmine was omitted.[2]

(Day 13):

- Since extubation was planned: Tab pyridostigmine 60 mg QID was started Steroids were tapered- tab. Omnacortil dose was reduced to 40 mg OD

(Day 15):

- Patient was put on spontaneous mode of ventilation with Psupport 10, PEEP 5, FiO2- 35%.
- Patient suddenly become unresponsive with no

respiratory effort and ventilator mode was changed to SIMV.
- On examination both pupils were constricted.
- Tab omnacortil was increased to 50 mg OD in consultation with the neurologist.
- Tab pyridostigmine were omitted.
- Patient improved after 4-5 hours with improvement in consciousness.

(Day 22):
- Inj methylprednisolone 1gm i.v. OD in 100 cc NS given over 45-60 minutes for 3 days

(Day 25):
- Patient was put on spontaneous mode of ventilation with Psupport 10, PEEP 5, FiO2- 35%.
- Tab pyridostigmine 60 mg QID was restarted.
- Patient was extubated and oxygen was given by ventimask FiO2 40%.
- Vitals were HR 98/m, BP- 138/90, SpO2 94%, RR-17/min.

(Day 26):
- Dose of Tab Pyridostigmine was increased to 60 mg + 120mg +60 mg + 120 mg

(Day 27):
- At 10:45 am patient had shortness of breath with RR= 45/min, SpO2 = 76-86% .
- On examination pupils were constricted at that time.
- Patient was given non invasive ventilation with interface. Psupport- 15 cm H2O, PEEP= 5, FiO2= 50%.
- Patient had 2 episodes of diarrohea.
- Next 2 doses of tablet pyridostigmine were withheld.
- Patient improved ater 3-4 hours and tablet pyridostigmine 60 mg tds was started on next day.

(Day 29):
- NIV removed and patient was given O2 by ventimask with FiO2 40%.
- HR= 87/M, BP= 152/98 mmHg, SpO2= 95%.

(Day 32):
- Patient shifted to medicine ward on room air.
- Shifting vitals; HR=92/min, BP= 152/96 mmHg, RR= 16/min, SpO2= 94% on room air
- Patient was discharged from the ward on day 34.

DISCUSSION

Triggers for myasthenia crisis include disease exacerbations, noncompliance with cholinesterase inhibitor medication, adverse effects of other medications, fever, and emotional stress. Cholinergic crises develop secondarily to an overdose of cholinesterase inhibitor medication. Respiratory failure may be present without other cholinergic symptoms.[3] Myasthenic crisis should be treated with a cautious administration of appropriate longer acting cholinesterase inhibitors. Treatment for cholinergic crisis is respiratory support, discontinuation of all anticholinesterase drugs.

How often does cholinergic crisis occur? Long back years ago, cholinergic crisis may occur was indisputable because of irreversible inhibitors of cholinesterase used in the treatment of myasthenia. It is not clear whether or how often it occurs in patients treated with the standard drugs now in use. Indeed, as crisis itself has become less frequent, the question has disappeared from the literature. There are still, however, occasional references to the use of edrophonium as a method to determine whether weakness is "myasthenic" or "cholinergic".

- no role for acetylcholinesterase inhibitors in the acute setting of exacerbation-

1) Although work quickly to improve neuromuscular transmission they may promote excessive secretions which can lead to diarrhoea or increasing pulmonary secretions.

2) Cardiac arrhythmias—especially common in patients with MGC—may be triggered by IV acetylcholinesterease inhibitors.

CONCLUSION

- First line treatment of myasthenia gravis patient is pyridostigmine.
- Its effect begins within 30 minutes, peaks at about 2 hours with a half-life of 4 hours.
- It should be slowly titrated to effect to avoid a cholinergic crisis.
- There is still no way to determine the optimal dosage of pyridostigmine therapy, except by a trial that depends upon subjective responses of patient and physician.
- However cholinergic crisis is insignificant in modern literature but the question arises is it still relavant in clinical practice?

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